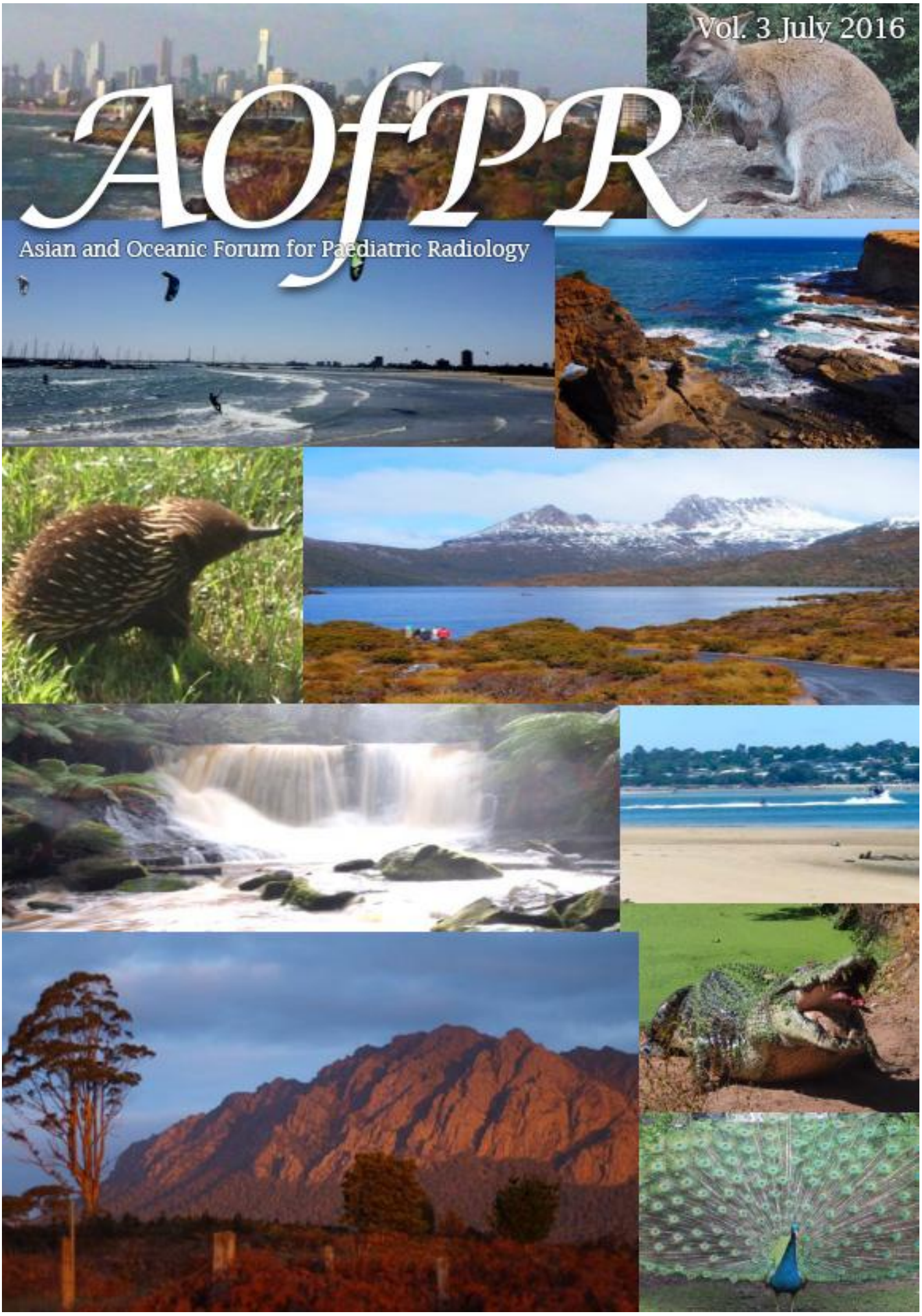
Vol. 3 July 2016
AOfPR
Asian and Oceanic Forum for Paediatric Radiology

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Asian and Oceanic Forum for Paediatric Radiology

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It is a great privilege for Australia to be asked to write the third issue of the Asian and Oceanic Forum for Pediatric Radiology and I am very pleased to be the guest editor.

Paediatric Radiology was formalized in Australia with the first stand-alone meeting of paediatric radiologists in 1979 at the Adelaide Children's Hospital. The following year, the Australasian Society of Paediatric Imaging (ASPI) was formed and a yearly scientific meeting has occurred since that time. In 2007 the society changed its name to the Australian and New Zealand Society of Paediatric Radiology (ANZSPR) to better reflect its membership. In 2012, for governance reasons and to improve its relevance to the general radiology community, it became a Special Interest Group of the Royal Australian and New Zealand College of Radiologists (RANZCR).

ASPI, and subsequently ANZSPR, has had ties with AOSPR since it's foundation. There have always been many joint members and in 2014 a very successful and enjoyable AOSPR/ANZSPR Conjoint Scientific Meeting was held in Queenscliff, Victoria. We look forward to many more years of successful collaboration.

I would like to thank all the contributors, the members of our Editorial Board and webmaster for their ongoing efforts to make this publication possible.

Magnetic Resonance Evaluation of Developmental Delay: Are contrast, Diffusion and Spectroscopy Sequences Necessary?

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ABSTRACT

INTRODUCTION: Developmental delay is common in the paediatric population and MRI is frequently used to evaluate the aetiology and guide management and prognosis. To minimize or exclude unnecessary sequences will reduce scan time and patient morbidity and increase the likelihood of obtaining a diagnostic study. This study aims to evaluate whether there is benefit of Gadolinium contrast, spectroscopy and DWI sequences for the routine evaluation of developmental delay. METHOD: A retrospective study at a tertiary paediatric institution was performed by a word search of the Radiology Information System for subjects less than 16 years who had an MRI performed between year 2005-11 with the clinical indication of "developmental delay". One hundred and fifteen patients were identified and included in the study. Patients were stratified by clinical indication for the MRI into four subgroups: isolated developmental delay 60/115 (52%), developmental delay and neurological findings 20/115 (17%), developmental delay and epilepsy 32/115 (28%) and developmental delay and suspicion of metabolic disorder 3/115 (3%). The usefulness of Gadolinium contrast, spectroscopy and DWI sequences was assessed as to whether they added any useful diagnostic information. RESULTS: There were similar rates of abnormal studies in all 4 study groups: isolated developmental delay (37%), developmental delay with neurological findings (35%), developmental delay and investigation for metabolic disorder (34%) and developmental delay with epilepsy (31%). In all of the cases performed for the investigation of developmental delay, the post contrast, DWI and spectroscopy sequences did not aid in establishing the diagnosis. CONCLUSION: The application of Gadolinium contrast, DWI and MR spectroscopy did not aid the evaluation of causes of developmental delay in our study. Their routine use in clinical practice may be of limited yield.

Keywords: Developmental delay, Diffusion weighted imaging, Post Contrast, Spectroscopy.

INTRODUCTION

Developmental delay is common in the paediatric and adolescent population with a prevalence of up to 2.2% [1]. Magnetic resonance imaging (MRI) is frequently used to evaluate the aetiology and therefore guide the management and prognosis of developmental delay.

It is challenging to obtain patient co-operation in this population. Minimizing unnecessary sequences will reduce scan time and not performing a post gadolinium sequence avoids unpleasant intravenous cannulation for young patients. The above measures reduce scan time and patient distress, enabling better patient co-operation and increase the likelihood of producing a diagnostic study.

This study aims to evaluate whether post contrast, magnetic resonance spectroscopy (MRS) and diffusion weighted imaging (DWI) sequences are useful for the routine evaluation of developmental delay.

METHODS

A retrospective audit was performed of MRIs with the primary clinical indication of developmental delay in patients under 16 years of age at a tertiary paediatric centre between 2005 and 2011. Relevant studies were identified by a search of the phrase "developmental delay" on an intranet based report search tool.

Eleven patients with a pre-existing diagnoses such as Pierre Robin, Prune belly syndrome, chromosomal abnormalities, Klippel Feil, Wilms tumour and Cowdens syndrome were excluded from the study, leaving a cohort of 115 patients (age range 9 months - 16 years, median age 4 years, 61/115 (53%) were male).

CLINICAL INDICATION	CONTRAST	DWI	SPECTROSCOPY	TOTAL
Developmental delay	14 (23%)	30 (50%)	7 (12%)	60 (52%)
Developmental delay and neurology	11 (55%)	9 (45%)	9 (45%)	20 (17%)
Developmental delay and metabolic disorder	3 (100%)	1 (33%)	3 (100%)	3 (3%)
Developmental delay and epilepsy	11 (34%)	21 (66%)	7 (22%)	32 (28%)
Total	39 (34%)	61 (53%)	26 (23%)	115 (100%)

Table 1. Clinical indication and sequences applied.

The scans were performed on a Siemens Symphony TIM (Erlangen, Germany) or Siemens AVANTO (Erlangen, Germany) 1.5 T MRI scanners. Routinely, T1, T2 and FLAIR sequences were obtained. In addition to this, 39/115 (34%) of patients had a post contrast sequence, 61/115 (53%) had DWI and 26/115 (23%) had MRS sequences (Figure 1).

Patients were stratified by clinical indication for the MRI into four subgroups: isolated developmental delay 60/115 (52%), developmental delay and neurological findings 20 (17%), developmental delay and epilepsy 32 (28%) and developmental delay with suspicion of metabolic disorder 3 (3%).

The images were reviewed by an experienced paediatric neuroradiologist and paediatric imaging fellow. The MRI diagnoses were classified into 7 descriptive categories: normal; signal abnormality with white matter loss; signal abnormality without white matter loss; no signal abnormality with white matter loss; ventricular dilation with white matter loss; ventricular dilatation without white matter loss and malformation.

The usefulness of each sequence was categorized into one of three categories: (1) not useful, (2) useful, and (3) essential in reaching the diagnosis.

RESULTS

The results of correlating the clinical indication with the sequences performed are summarized in Table 1 and Figure 1. Patients with developmental delay and suspicion of underlying metabolic disorder were all administered contrast and spectroscopy (100%). Those who had developmental delay and neurological findings had the next highest incidence of contrast administration (55%). Those who had developmental delay and epilepsy had the highest frequency of DWI studies (66%)

The results of correlating the clinical indication with MRI findings are summarized in Table 2. The percentage of abnormal results were similar in distribution in each of the patient categories; 37% in isolated developmental delay, 35% in developmental delay with neurological findings, 34% in developmental delay and investigation of metabolic disorder and 31% in developmental delay and epilepsy.

Results of correlating of the sequences performed versus the MRI findings are summarized in Table 3 and 4. There

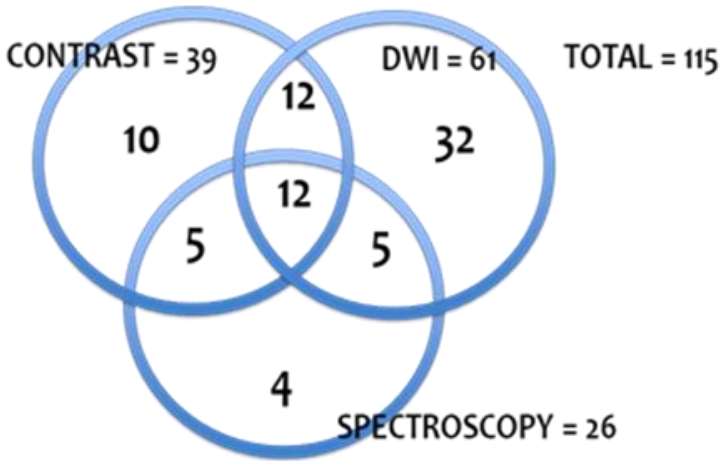

Figure 1. Summary of sequences applied.

CATEGORY	NORMAL	VENTRICULAR DILATION WITH NO WHITE MATTER LOSS	WHITE MATTER LOSS WITH VENTRICULAR DILATION	WHITE MATTER LOSS WITH SIGNAL ABNORMALITY	SIGNAL ABNORMALITY WITH NO WHITE MATTER LOSS	WHITE MATTER LOSS AND NO SIGNAL ABNORMALITY	MALFORMATION	TOTAL
Developmental delay	38 (63%)	3 (5%)	7 (12%)	5 (8%)	3 (5%)	1 (2%)	3 (5%)	60 (100%)
Developmental delay and neurological findings	13 (65%)	0 (0%)	0 (0%)	4 (20%)	0 (0%)	1 (5%)	2 (10%)	20 (100%)
Developmental delay and suspected metabolic disorder	2 (66%)	1 (33%)	0 (0%)	0 (0%)	0 (0%)	0 (0%)	0 (0%)	3 (100%)
Developmental delay and epilepsy	22 (69%)	1 (3%)	2 (6%)	3 (9%)	1 (3%)	0	3 (9%)	32 (100%)
Total	75 (65%)	5 (4%)	9 (8%)	12 (10%)	4 (3%)	2 (2%)	8 (7%)	115

Table 2. Clinical indication and MRI findings

was no correlate between the patient category or the sequences applied to the abnormalities detected. Of those who received contrast, 36% were abnormal however in no cases did the administration of contrast impact upon the diagnosis of pathology.

The MRI findings are summarised in Table 2. Most patients (65%) had a normal MRI scan. In a minority of patients (7%) a specific cerebral malformation was diagnosed. The remaining 28% had variable non specific white matter volume loss or gliosis and associated ventricular dilatation. Examples of pathology detected included malformations such as ectopic posterior pituitary (Figure 2), focal gliosis (Figure 3), migrational abnormality (Figure 4) and periventricular leukomalacia (white

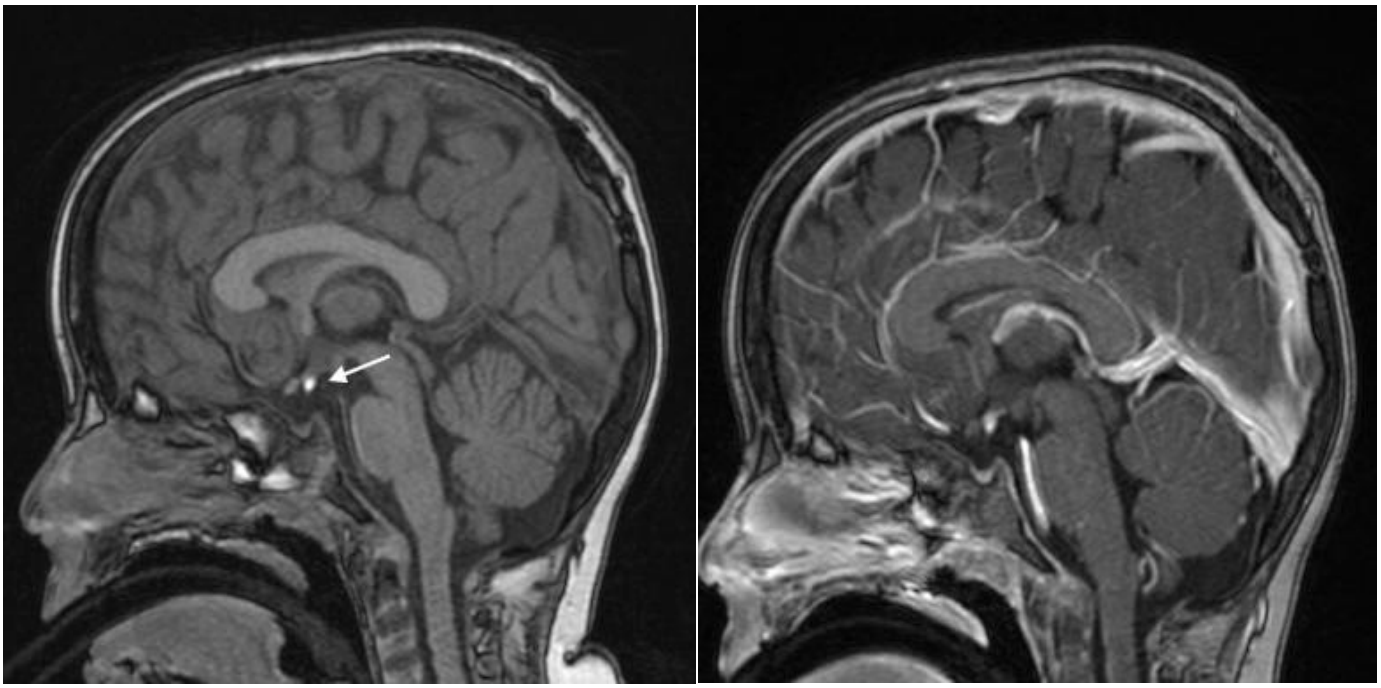

Figure 2. 5-year old male with global developmental delay. Non contrast sagittal T1 weighted image (a) demonstrates ectopic posterior pituitary located posterior to the optic chiasm (arrow) (a). Post contrast images (b) did not add diagnostic information.

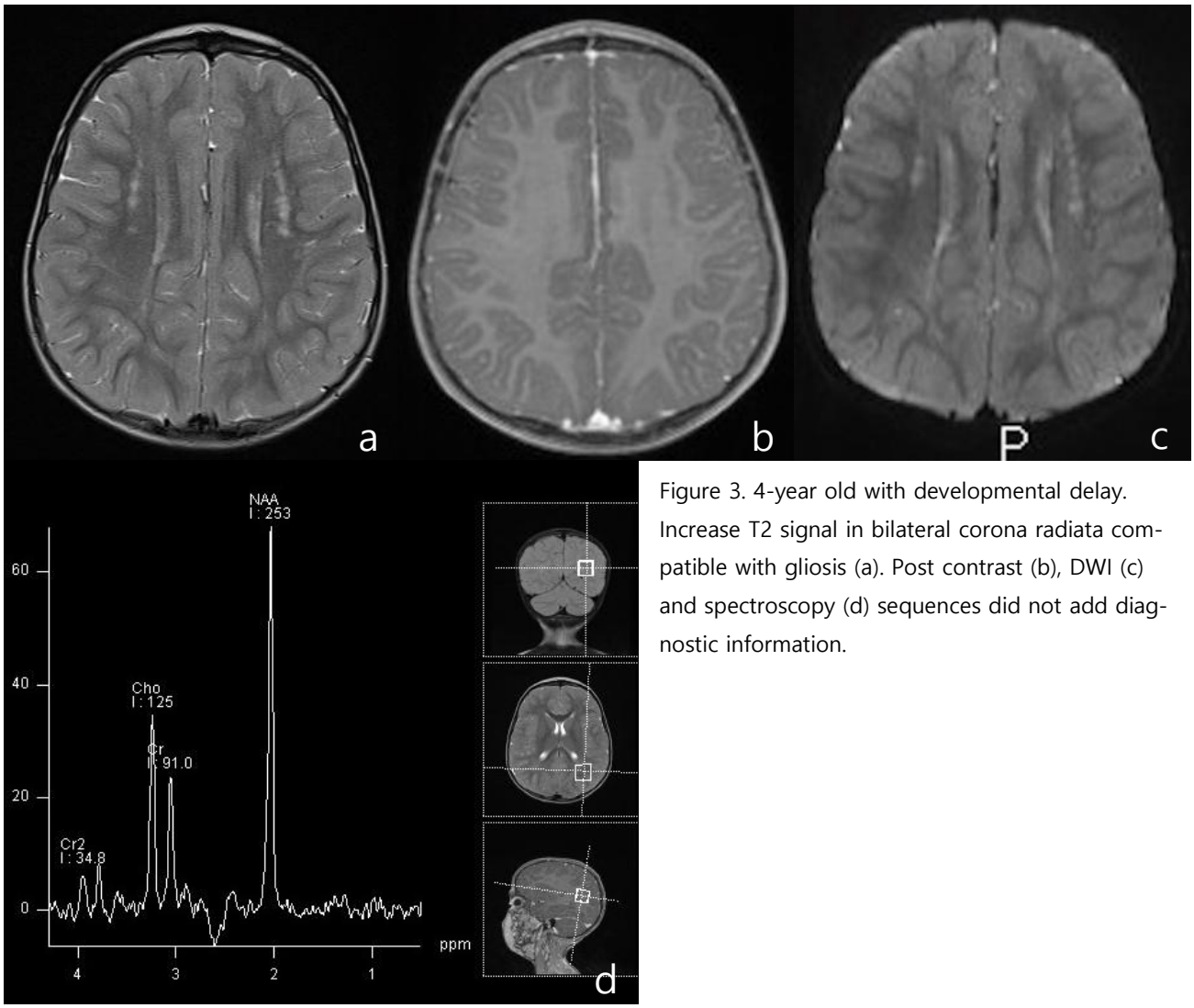

Figure 3. 4-year old with developmental delay. Increase T2 signal in bilateral corona radiata compatible with gliosis (a). Post contrast (b), DWI (c) and spectroscopy (d) sequences did not add diagnostic information.

matter injury of prematurity) (Figure 5). In none of these studies did the application of contrast, DWI or spectroscopy aid in the diagnosis.

DISCUSSION

The yield of MRI in evaluation of the underlying causes of developmental delay has been reported to be approximately 30%, with higher frequency if developmental delay is associated with positive neurological findings (41.2%) [2]. This was not replicated in this study which demonstrated similar rates of abnormal studies in patients with a history of isolated developmental delay (37%), developmental delay with neurology (35%), developmental delay and investigation of metabolic disorder (34%) and developmental delay and epilepsy (31%).

In all of the cases performed for the investigation of developmental delay, the post contrast, DWI and spectroscopy sequences did not aid in establishing the diagnosis. This is not surprising, as developmental delay, is a chronic non progressive condition and the underlying

SEQUENCE	NORMAL	ABNORMAL	USEFUL	TOTAL
Contrast	25 (64%)	14 (36%)	0 (0%)	39
DWI	44 (72%)	17 (28%)	0 (0%)	61
MRS	15 (58%)	11 (42%)	0 (0%)	26
Total	84 (67%)	42 (33%)	0 (0%)	126

Table 3. Sequence performed vs presence or absence of abnormality

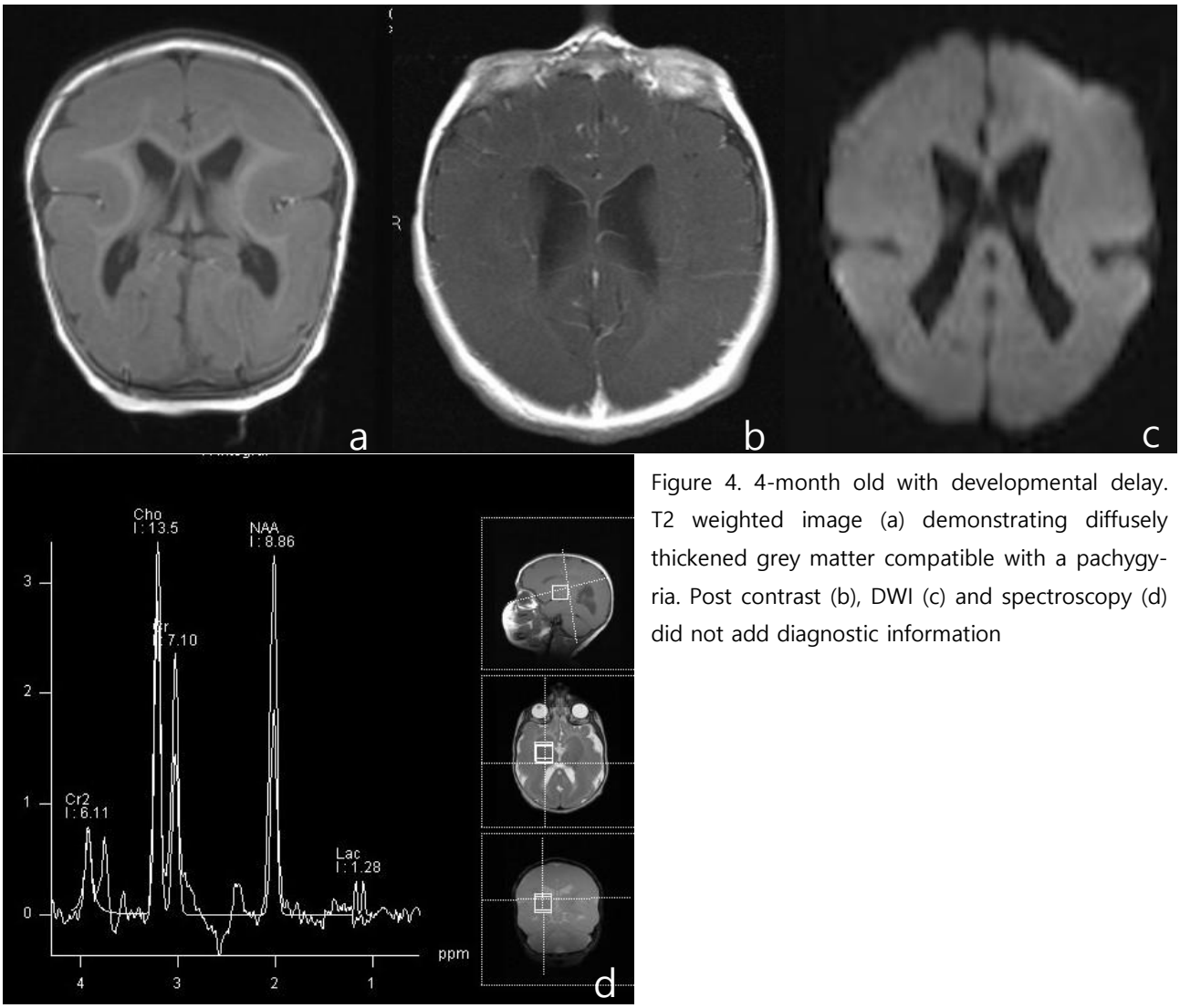

Figure 4. 4-month old with developmental delay. T2 weighted image (a) demonstrating diffusely thickened grey matter compatible with a pachygyria. Post contrast (b), DWI (c) and spectroscopy (d) did not add diagnostic information

pathologies such as a brain malformation or birth related injury will not demonstrate abnormal contrast enhancement, restricted diffusion or MRS abnormalities. If the clinical setting is progressive with evidence of regression, and not simple developmental delay, then active pathologies such as metabolic disorders, tumours and inflammatory conditions need to be excluded and these sequences then become useful.

Previous studies focusing on the use of administration of gadolinium contrast sequences in this population suggest a limited role for this indication [3, 4]. A study by Foerster et al. which evaluated the indiscriminatory use of gadolinium contrast in 170 patients under the age of 2 for evaluation of developmental delay concluded that post contrast imaging has extremely low yield unless there is a clinical concern regarding tumour or infection [4]. No studies evaluating the yield of contrast in older children have been published.

The use of DWI in neuro-imaging is usually reserved for detection of early cellular injury or necrosis such as in acute ischemic insult. DWI sequences did not add diagnostic value in this study, which was an expected finding as developmental delay is not a progressive condition.

The application of MR spectroscopy is not well documented in the literature. There are studies which support the use of MRS in patients with neurological signs however its role in patients with no neurological abnormality is very limited [5,6]. In our study spectroscopy was performed in 45% of patients with a history of neurology however in no cases did spectroscopy aid in diagnosing the underlying cause of developmental delay. Spectroscopy was applied to all patients investigated for a metabolic disorder however the sample size was only 3 and the results of these studies were all normal. The use of spectroscopy in diagnosing metabolic disorders has been well documented in the literature. In particular,

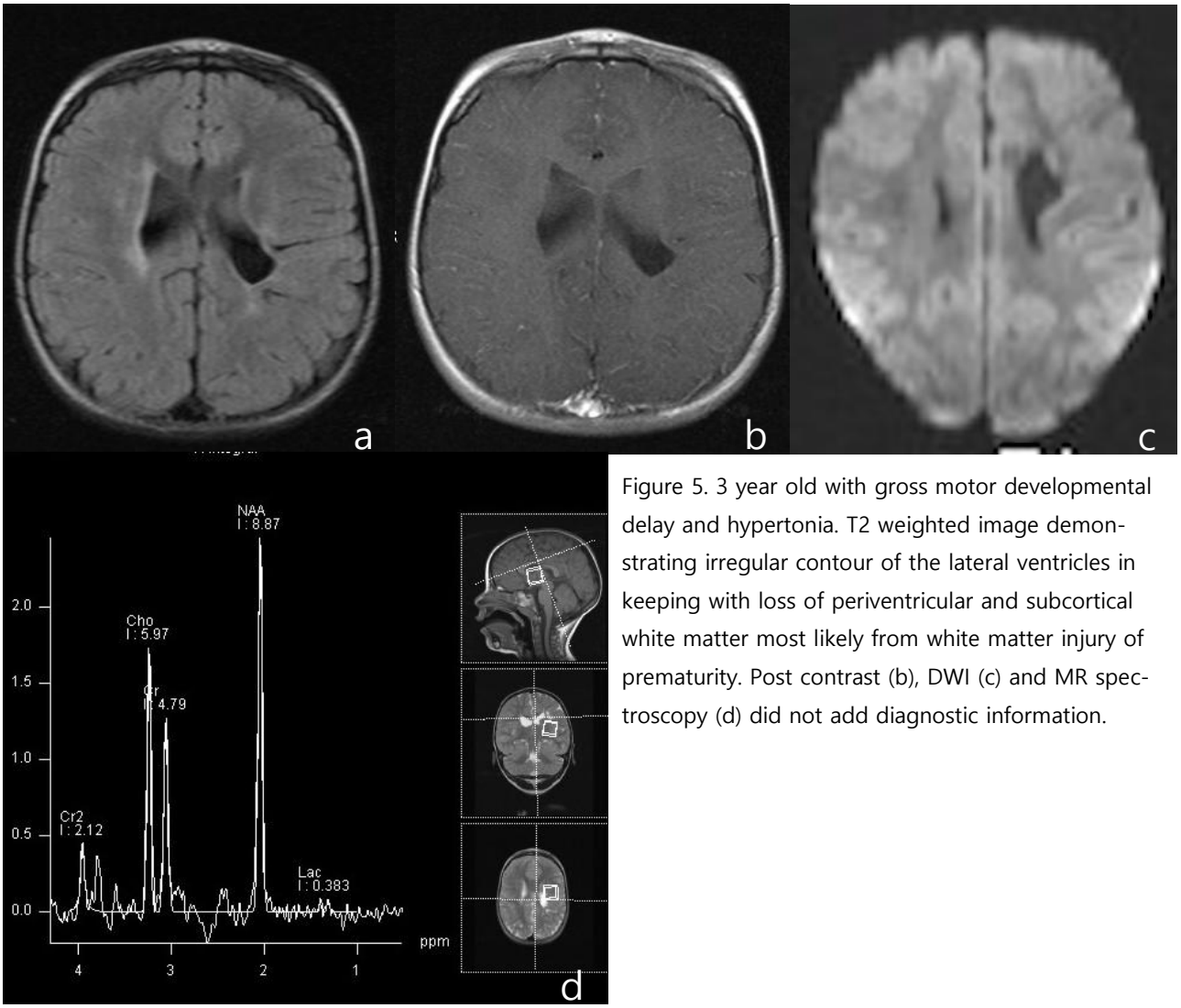

Figure 5. 3 year old with gross motor developmental delay and hypertonia. T2 weighted image demonstrating irregular contour of the lateral ventricles in keeping with loss of periventricular and subcortical white matter most likely from white matter injury of prematurity. Post contrast (b), DWI (c) and MR spectroscopy (d) did not add diagnostic information.

MRS has been shown to be useful in otherwise normal appearing brain tissue in the context of metabolic disorders such as creatine deficiency syndromes [7]. However, due to the rare incidence of these conditions and the usual clinical setting of a progressive condition and patient deterioration, the value of routine application of MRS is questionable.

CATEGORY	NORMAL	VENTRICULAR DILATION WITH NO WHITE MATTER LOSS	WHITE MATTER LOSS WITH VENTRICULAR DILATION	WHITE MATTER LOSS WITH SIGNAL ABNORMALITY	SIGNAL ABNORMALITY WITH NO WHITE MATTER LOSS	WHITE MATTER LOSS AND NO SIGNAL ABNORMALITY	MALFORMATION	TOTAL
Contrast	25(64%)	2(5%)	3(8%)	3(8%)	3 (8%)	0 (0%)	3 (8%)	39 (100%)
DWI	44(72%)	2(3%)	3(5%)	7(11%)	3 (5%)	0 (0%)	2 (3%)	61 (100%)
MRS	15(58%)	1(4%)	1(4%)	5(19%)	2 (8%)	0 (0%)	2 (8%)	26 (100%)
Total	84(67%)	5(4%)	7(6%)	15 (12%)	8 (6%)	0 (0%)	7 (6%)	

Table 4. Pathology related to sequences

CONCLUSION

The application of post contrast, DWI and MR spectroscopy did not aid the diagnosis of the underlying cause of developmental delay in our study. This suggests their routine use in clinical practice may be of limited yield.

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White Matter Injury of Prematurity: Slipping through the net.

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ABSTRACT

INTRODUCTION: White matter injury of prematurity (WMIP) is relatively common in premature patients and is a significant cause of developmental disabilities in childhood. WMIP is usually diagnosed by cranial ultrasound (US) performed in the neonatal period, however MRI at a term equivalent age is increasingly used because of its greater sensitivity. This study was performed to determine the frequency of WMIP following normal cranial US. METHODS: A retrospective study at a tertiary paediatric institution was performed by a word search of the Radiology Information System for subjects less than 18 years who had a post term equivalent age (TEA) MRI performed between 2006-14 with a diagnosis of "White Matter Injury of Prematurity", "WMIP", "Periventricular Leucomalacia" or "PVL". This identified 205 subjects. 133 of these were diagnosed with WMIP after review of the reports and imaging. 60 /133 (45%) subjects (male 47%, average gestational age 29 weeks) also had at least one neonatal cranial US performed and these represented the study group. The US reports and images were reviewed and abnormalities consistent with WMIP at 1-7 days, 8-14 days and more than 14 days of life were recorded. RESULTS: Neonatal US performed at 1-7 days, 8-14 days and more than 14 days of life in patients later diagnosed to have WMIP were normal in 16/45 (40%), 12/23 (52%) and 23/45 (51%) of cases respectively. Forty five percent (21/47) of US performed after 1 week were normal. CONCLUSION: Neonatal cranial US is frequently normal in patients later diagnosed with WMIP by MRI.

INTRODUCTION

Among the potential neurological events that can occur during the vulnerable preterm neonatal period, white matter injury of prematurity (WMIP), also known as periventricular leucomalacia (PVL), is the most common and most significant cause of long-term neurodevelopmental impairment and cerebral palsy with a reported incidence of between 3-20%. [1]

The white matter in premature infants (< 30 weeks) and very low birth weight patients (< 1500g) has been shown to be particularly vulnerable to ischaemia, infection and inflammation. This is due to the delicate interplay of the cerebral end arteries at the periventricular zone, immature cerebral autoregulation and the highly susceptible premyelinating oligodendrocytes, which are concentrated in the periventricular white matter [2].

WMIP encompasses a spectrum of findings that can be broadly categorised into cystic and non-cystic lesions. Due to advances in neonatal intensive care and preventative measures, the severe cystic and focally necrotic form is now less common, accounting for less than 10% of cases [3,4]. The non-cystic form is a more diffuse lesion, which results in global delay of myelination secondary to the loss of the early premyelinating oligodendrocytes and astrogliosis. Both the cystic and non-cystic WMIP can result in white matter volume loss and gliosis. [5].

The two main neuroimaging modalities for detection of WMIP are cranial ultrasound and MRI. The early US features are of periventricular echogenicities in the first two weeks (Figure 1), which may progress to cyst formation (Figure 2) and white matter volume loss. These echo-

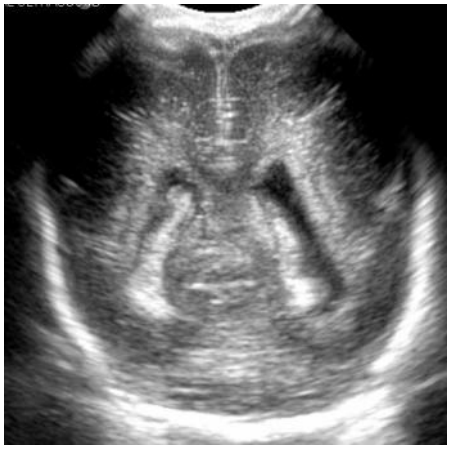

Figure 1. Cranial US performed at 8 days in a neonate born at 25 weeks gestation demonstrating increased periventricular white matter echogenicity.

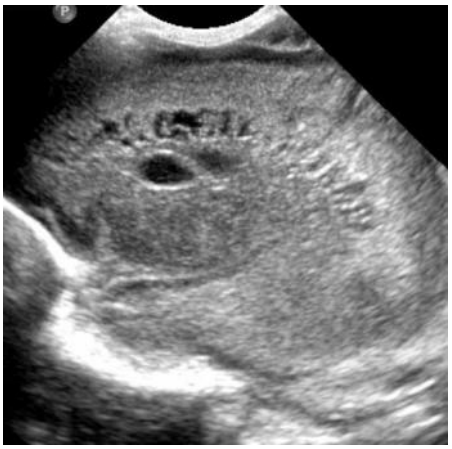

Figure 2. Cranial US performed at 25 days in a neonate born at 26 weeks gestation demonstrating increased periventricular cyst formation (Cystic WMIP).

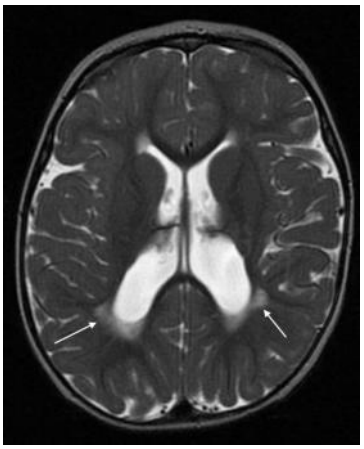

Figure 3. Axial T2 weighted MRI performed at 9 months of age demonstrating white matter loss with increased T2 signal (arrows) and secondary lateral ventricular dilatation associated indicative of WMIP.

genicities are non-specific and can also be seen as a transient phenomenon in normal premature infants. Thus, multiple sequential US are often required to detect and monitor these often subtle white matter abnormalities [6]. WMIP will produce white matter volume loss with secondary lateral ventricular dilatation and increased T2 white matter signal compatible with gliosis (Figure 3).

The non-cystic diffuse WMIP has been demonstrated to be the most difficult to detect on cranial ultrasound [7]. Frequently patients with normal serial cranial US will have WMIP diagnosed by term equivalent age (TEA) MRI. Cranial US can therefore be non specific and insensitive in the diagnosis of WMIP. This study was performed to determine the frequency WMIP occurs following normal serial cranial US.

METHODS

A retrospective study at a tertiary paediatric institution was performed by a word search of the Radiology Information System for subjects less than 18 years who had a post TEA MRI performed between 2006-14 with a diagnosis of "White Matter Injury of Prematurity", "WMIP", "Periventricular Leucomalacia" or "PVL". This identified 205 subjects. One hundred and thirty three of these were diagnosed with WMIP after review of the reports and imaging. WMIP was diagnosed on MRI by the white matter volume loss or the presence of increased T2 signal.

60 /133 (45%) subjects also had at least one neonatal cranial US performed (113 scans in total) and these were the study group (28 (47%) male, average gestational age 29 weeks). The US reports and images were reviewed for the absence or presence of abnormalities consistent with WMIP at 1-7 days (white matter echogenicity), 8-14 days (white matter echogenicity), and more than 14 days of life (white matter echogenicity, cysts and volume loss).

RESULTS

The results are summarised in Table 1. Neonatal US performed at 1-7 days, 8-14 days and more than 14 days of life in patients later diagnosed to have WMIP were normal in 16/45 (40%), 12/23 (52%) and 23/45 (51%) of cases respectively. Forty five percent (21/47) of US performed after 1 week were normal. Examples of these cases are illustrated in Figures 4 and 5.

DISCUSSION

Neuroimaging has become an integral component of

Age at which US performed	Number of Patients in which ≥1 US Performed	Number of Patients with Normal US
1-7 Days	45/60 (75%)	16/45 (40%)
8-14 Days	23/60 (38%)	12/23 (52%)
>14 Days	45/60 (75%)	23/45 (51%)
>7 Days	47/60 (78%)	21/47 (45%)

Table 1. Results of the number of normal cranial US performed at in the age groups that were studied.

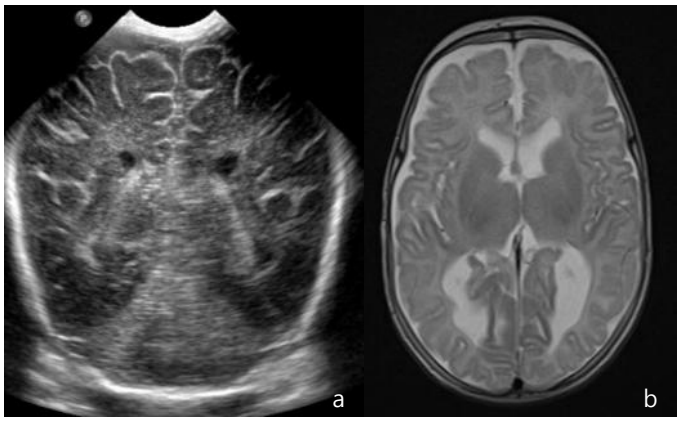

Figure 4. Coronal image (a) from a Cranial US performed in an ex 23.5 gestation neonate at 6 weeks of age demonstrating no abnormality. Axial T2 weighted image (b) of the same patient performed at 4 months of age demonstrating marked loss of white matter and secondary lateral ventricular dilatation predominantly effecting the parietal and occipital lobes.

preterm neonatal clinical care. Early detection and accurate characterisation of WMIP aids the clinician in counselling parents and predicts neurodevelopmental outcomes. The diagnosis of WMIP has relied on cranial US, which has the convenience of being portable and not using radiation. More recently TEA MRI has also been used in view of its greater sensitivity to this diagnosis [3].

This study demonstrates that almost 50% of premature infants with WMIP diagnosed by MRI will have a normal cranial ultrasound. More specifically, 40% of infants diagnosed with WMIP on MRI had a normal ultrasound performed before 7 days of life and 45% of these infants had a normal ultrasound performed after 7 days of life. This is concordant with other studies, which have concluded that the prevalence of WMIP based purely on US screening is significantly underestimated [3,8,9].

US has been shown to be sensitive and specific in detecting the severe, cystic form of WMIP. However, this form is now a rare complication in the premature neonate [2]. Conversely, US lacks the sensitivity and positive predictive value in detecting the non-cystic form of WMIP when compared to MRI [1,8,9,10]. The MR imaging features of the non-cystic form of WMIP include abnormal signal intensity in the periventricular white matter and posterior limb of the internal capsule and white matter volume loss, which can be difficult to confidently assess on US.

Inder et al further demonstrated that even if prolonged white matter echogenicity (echogenicity that persists for more than 7 days) is detected on ultrasound, it has a high false positive rate and low positive predictive value for WMIP [1]. In contrast, MRI provides a sensitive assessment for the more common, non-cystic diffuse WMIP and intracranial haemorrhage. Similar to US, MRI is also sensitive in detecting the cystic, less common type of WMIP. [6,8,9]

The main prohibitive factor to the use of MRI remains its availability and whether the infant is clinically stable to allow for transportation to and performance of the MRI. Although cranial ultrasound will likely remain a convenient modality for screening and detecting the less com-

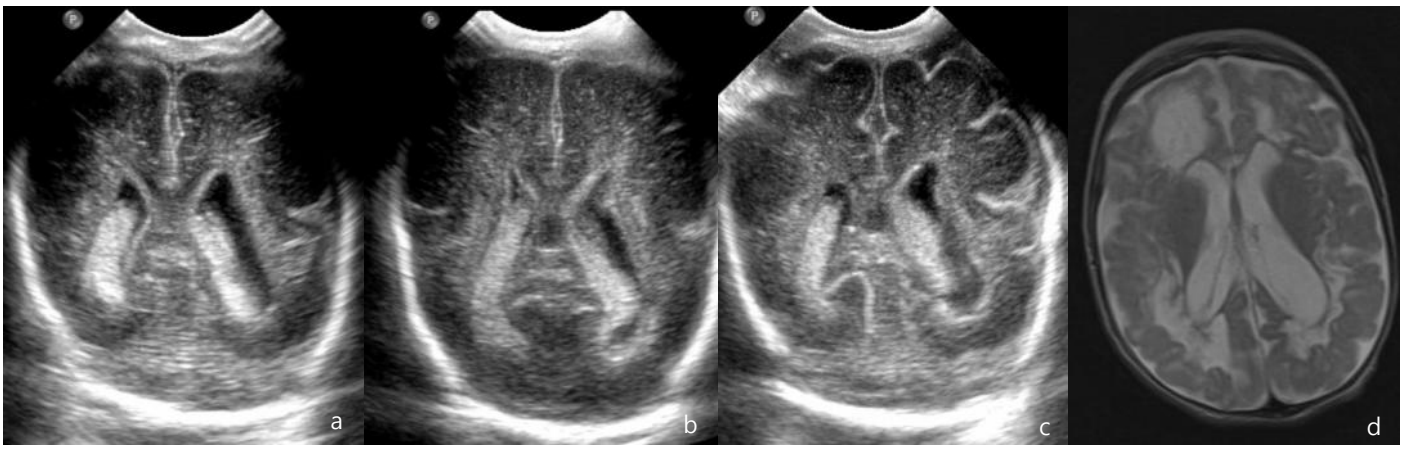

Figure 5. Coronal images from Cranial US performed in an ex 24 week gestation neonate at 6 (a), 12 (b) and 29 (c) days of age demonstrating no abnormality. Axial T2 weighted image (d) of the same patient performed at 5 months of age demonstrating marked signal change and loss of white matter and secondary lateral ventricular dilatation.

mon, severe cystic WMIP, we have shown that approximately 50% of preterm infants with WMIP will be under diagnosed and "slip through" the net of screening US.

The main limitations of this study include its retrospective nature and that not all patients had an US performed in all 3 of the studied time periods. However, the cohort represents the practice of a large tertiary centre. The study will also have some bias in that not all neonatal patients had an MRI and those that did may have a greater probability of an abnormal finding and WMIP.

CONCLUSION

Premature infants with a normal neonatal cranial US have approximately a 50% chance of WMIP demonstrated on a term equivalent age MRI.

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External Ear Cerumen Masquerading as Cholesteatoma.

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Abstract

This case reports a potential pitfall in the use of Diffusion Weighted Imaging (DWI) in the diagnosis of Cholesteatoma in the petrous temporal bone.

INTRODUCTION

Cholesteatoma is a common inflammatory condition that consists of ectopic keratinised epithelium that desquamates internally, continuously increasing in size as it accumulates keratin and epithelial debris. Middle ear cholesteatoma (MEC) is an important imaging diagnosis because it can cause serious labyrinthine and intracranial complications via local pressure effect and erosion, including loss of hearing. The treatment of choice is surgery, which aims to eradicate the cholesteatoma while maintaining hearing function and structural integrity of the middle ear. [1]

Once MEC is suspected, investigation commonly comprises of a dedicated temporal bone CT to characterise any bony erosion or deficiency and identify soft tissue or fluid within the middle ear cleft. However, CT lacks the specificity to differentiate MEC from other inflammatory conditions of the middle ear (which often do not require surgery). Dedicated MRI is often used to provide a specific diagnosis of MEC as diffusion weighted imaging (DWI) is highly sensitive and specific for MEC, although it does not provide the same anatomical detail as CT of the temporal bones. [1] In our institution, both CT temporal bones and dedicated MRI are utilised in order to accurately characterise MEC. Intravenous gadolinium based contrast material is not routinely administered as DWI alone has been shown to be highly sensitive and specific. [2] This is commonly considered straightforward, although there are some pitfalls that Radiologists should be aware of when interpreting such imaging. One such pitfall is presented; a case where cerumen in the external ear canals bilaterally provided for a false positive result on MRI.

CASE REPORT

This case was of an eleven year old girl with bilateral hearing loss with narrow external auditory canals in whom the Ear Nose and Throat Surgeon was concerned about middle ear disease and requested an MRI of the petrous temporal bones (figure 1). The MRI was performed on a 1.5 T Siemens Avanto and T1, T2 and Diffusion Weighted Sequences with ADC Maps were performed. The MRI demonstrated tissue of diffusion restriction in the region of middle ear cleft bilaterally and symmetrically. The diffusion restriction was concerning

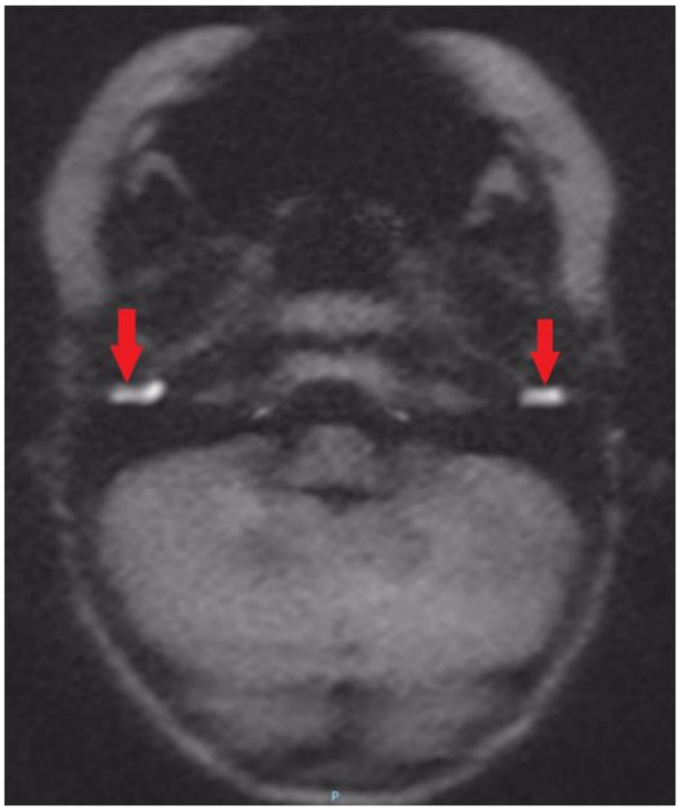

Figure 1. T2 haste axial diffusion weighted image showing foci of diffusion restriction (arrows) near each middle ear cleft, but actually within each external ear canal and consistent with cerumen.

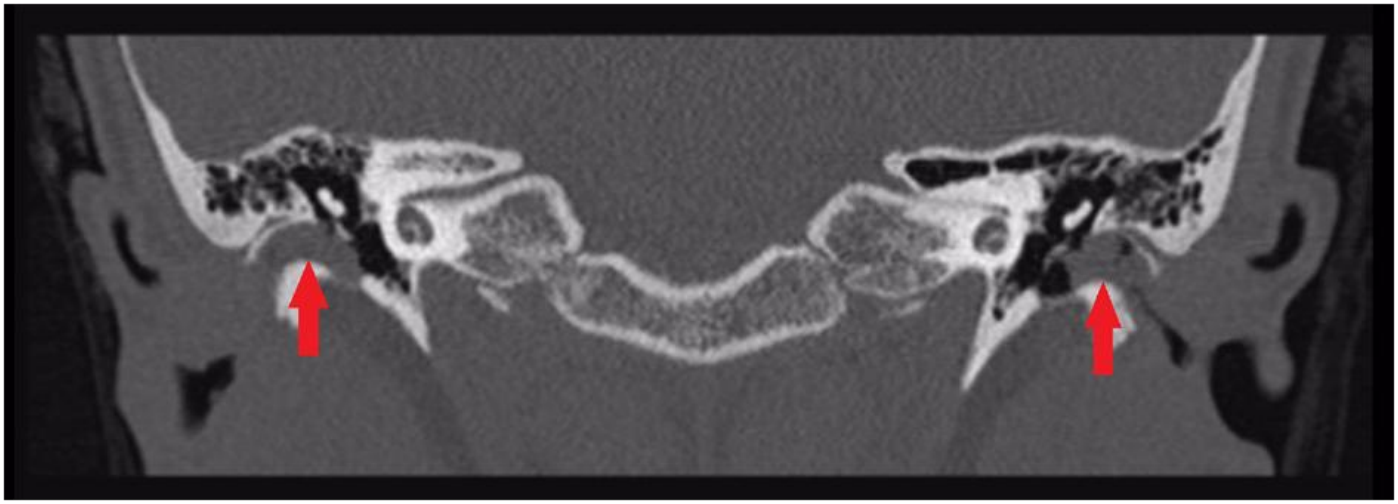

Figure 2. Fine coronal slice CT of the of the temporal bones shows no soft tissue within either middle ear cleft but soft tissue within each external ear canal (arrows), consistent with cerumen.

for cholesteatoma, however, the distribution of the diffusion restriction was felt to be atypical for this condition.

A temporal bone CT was performed (figure 2) and demonstrated well aerated middle ears and complete opacification of the external auditory canals. The diffusion restriction was elongated along the long axis of each external ear canal and with CT imaging correlation it was possible to determine that the restricted diffusion was confined to the external auditory canal. The CT also excluded bony erosion.

Cerumen demonstrates diffusion restriction (De Foer, et al., 2010) due to its waxy composition and therefore the correct diagnosis of cerumen within each external ear canal was made.

DISCUSSION

Certain false-positive results have been reported in the literature regarding the diagnosis of MEC using DWI. These include residual haemorrhage, Sialastic material within the middle ear, bone graft material, cholesterol granuloma, abscess and cerumen. [1] Awareness of these potential pitfalls is important to avoid misdiagnosis. Furthermore, this case demonstrates the importance of accurate anatomical location of abnormal DWI signal, paying particular attention to correlation with the CT imaging in order to accurately localise any abnormality.

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Absence of the Right Pulmonary Artery with Ipsilateral lung cysts —part of the congenital lung malformation spectrum? A case report.

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ABSTRACT

Unilateral absence of a pulmonary artery (UAPA) is a rare condition that can be associated with other congenital cardiac conditions or may occur in isolation. We report a case of absence of the right pulmonary artery associated with ipsilateral lung cysts. We explore the embryological origin of this condition and theorise that both of these findings may be part of the congenital lung malformation spectrum.

CASE PRESENTATION

A 7-year-old female presented in April 2008, to the Royal Children's Hospital in Melbourne, with a 3-day history of cough and fever. One week prior to presentation, she had an episode of sudden onset of coughing following ingestion of vegetable soup. She had no other relevant previous medical history. On clinical examination there was reduced air entry to the right hemithorax. A chest radiograph demonstrated reduced volume of the right hemithorax with elevation of the right hemidiaphragm, mediastinal shift to the right and compensatory hyperinflation and contralateral herniation of the left lung (Figure 1). The density of the right lung was diffusely mildly increased compared to the left lung. There was no evidence of air trapping on the expiratory radiograph. Apart from the compensatory hyperinflation, the left lung was otherwise normal. The appearances were felt to be due to right pulmonary hypoplasia, with airway obstruction by inhaled foreign body or mucus plug considered as a differential diagnosis. Bronchoscopy was normal with no airway obstruction seen. The patient was treated with antibiotics and was well on discharge.

The patient was lost to follow-up during a period of time between 2008 and 2014. Her parents gave a history of her having recurrent lower respiratory tract infections during this time, which were managed on an outpatient

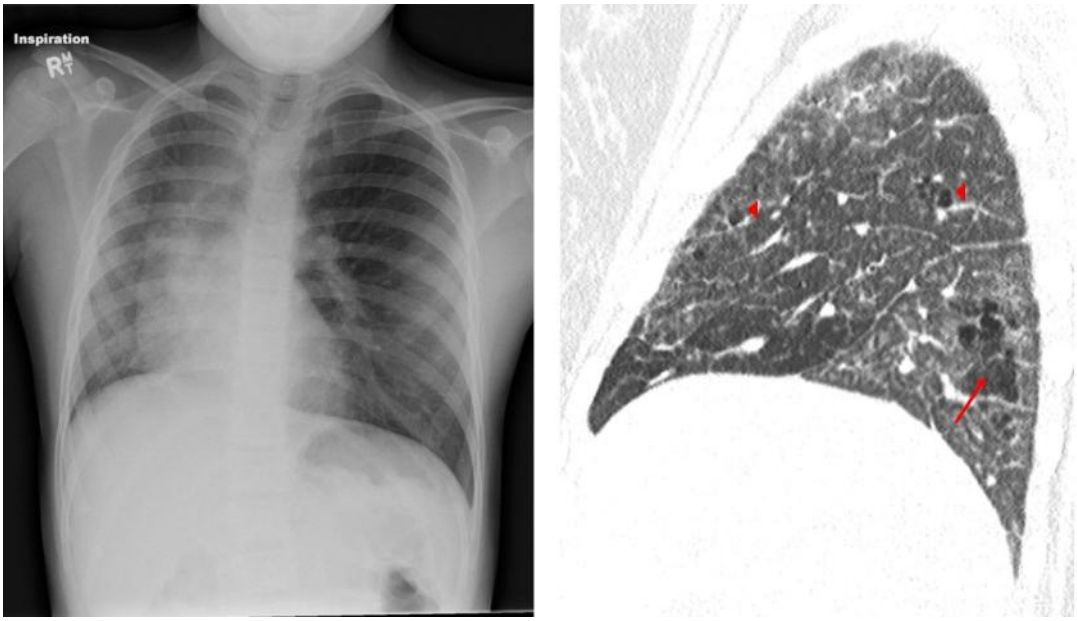

Figure 1: (a) Frontal chest radiograph acquired at time of presentation in 2008. The right hemithorax is smaller than the left with mediastinal shift to the right and compensatory hyperinflation of the otherwise normal left lung. (b) Sagittal reformat of the CT thorax in lung window shows a cluster of subcentimeter cysts in the right lower lobe (red arrow). Similar cysts are also noted in the right upper lobe (red arrowheads) and middle lobe (not shown).

basis with oral antibiotics. Subsequent chest radiographs have not demonstrated any interval change. Echocardiography is normal, with no evidence of pulmonary hypertension. Pulmonary function tests were normal.

In May 2015 a contrast-enhanced CT thorax was performed to further delineate the underlying anatomy and to assess for the presence of bronchiectasis in the setting of recurrent infection. The findings on CT were as follows:

1. Complete absence of the right main pulmonary artery. The pulmonary trunk was normal in appearance but terminated in a solitary left main pulmonary artery. There was a normal left-sided aortic arch (Figure 2).

2. There was evidence on the right of systemic collateral supply from the thoracic aorta. Enlarged bronchial, intercostal and internal mammary arteries were seen on the right side. There was also supply from the abdominal aorta, with a collateral arising from the right lateral wall of the coeliac trunk. Serrated pleural thickening was seen, presumed to represent trans-pleural collateral vessels. The right pulmonary veins were normal in configuration but smaller when compared to the left. There was no anomalous draining vein seen (Figure 2).

3. The right lung was smaller in volume, with multiple parenchymal cysts in all lobes, the largest measuring 7 mm in diameter. The intervening parenchyma contained patchy ground-glass densities with diffuse smooth thickening of the interlobular septa. There was inferior traction of the horizontal fissure indicating volume loss in the middle lobe. There was no evidence of bronchiectasis.

Our patient is currently being managed conservatively as she is otherwise well. A lung perfusion scan was organized but unfortunately she did not attend. She remains under ongoing follow-up by the paediatric respiratory team.

DISCUSSION

Unilateral absence of a pulmonary artery (UAPA) is also known interchangeably as unilateral agenesis or unilateral interruption of a pulmonary artery. It can be associated with other congenital cardiac anomalies or occur in isolation[1-6]. In the largest prevalence study to date by Bouros et al., from a series of young males presenting for military screening, the estimated prevalence was 1 in 100 000. Half of the cases from this series were isolated, while the other half had associated congenital cardiac anomalies[1]. The actual prevalence of isolated UAPA is difficult to estimate as more than 50% can be asymptomatic and incidentally discovered on chest radiographs performed for various other reasons[2].

The pathogenesis of UAPA has been discussed at length[2, 3,7, 8]. Embryologically, the pulmonary arteries arise from multiple origins[3]. The main pulmonary trunk arises from the truncoaortic sac while the distal intrapulmonary arteries arise from their respective lung buds. In between the main and distal pulmonary arteries, the extrapulmonary portion of these arteries arise from the proximal portion of the sixth aortic arch. It is the involution of this proximal portion of the sixth aortic arch that is thought to give rise to UAPA. In accordance to this, Apostolopoulou argues that the more appropriate term for this condition would be proximal interruption of the pulmonary artery[3]. This process is thought to occur in utero with the affected lung receiving blood supply from the ductus arteriosus which arises from the distal portion of the sixth aortic arch. In utero, the blood supply is thought to be adequate with ensuing normal fetal lung development[7]. After birth, with closure of the ductus arteriosus, the blood supply to the distal pulmonary artery is interrupted with subsequent pulmonary hypoplasia and development of collateral supply from systemic arteries[2, 8]. This theory is supported by angiographic findings of Apostolopoulou, who demonstrated ductus arteriosus ipsilateral to the absent pulmonary artery and progressive cessation of blood supply to the affected pulmonary artery due to closure of the ductus arteriosus[3].

The radiographic features of UAPA are a combination of the ensuing decreased blood supply, development of systemic collateral supply and superimposed complications such as infections. Chest radiographs can demonstrate a small lung and this may be accompanied by elevation of the ipsilateral hemidiaphragm, compensa-

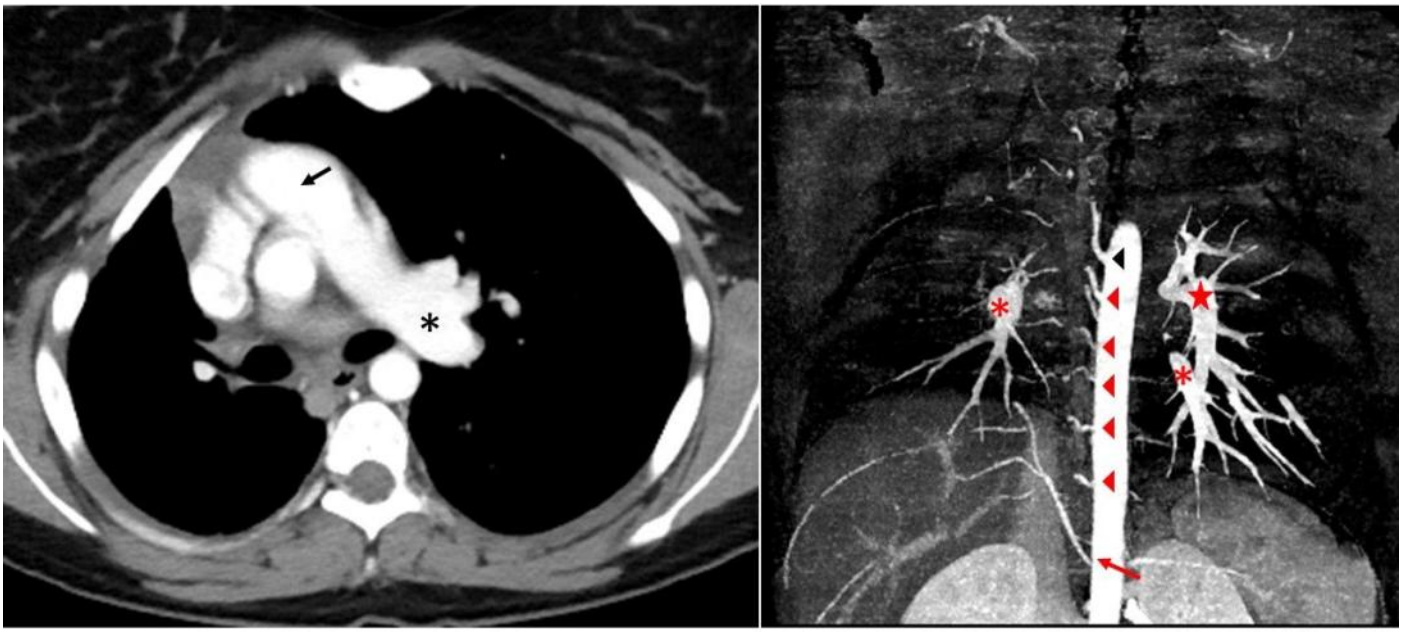

Figure 2: (a) An axial section of a contrast-enhanced CT of the thorax shows a complete absence of the right main pulmonary artery. The solitary left main pulmonary artery (black asterisk) is seen to arise from the pulmonary trunk (black arrow). Note is made of the mediastinal shift to the right. (b) A coronal MIP reformat of the same CT demonstrates systemic arterial feeders from the bronchial artery (black arrowhead), multiple intercostal arteries (red arrowheads) and from the coeliac trunk (red arrow). Both pulmonary veins (red asterisk) are demonstrated but only the left pulmonary artery (red star) is present.

tory hyperinflation of the contralateral lung and mediastinal shift to the affected side[4, 8, 9]. These features were seen in our patient. In addition, reduced pulmonary vascular markings can be seen[5, 9]. In our patient, contrast enhanced CT shows the multisource collateral supply. Besides collaterals from bronchial, intercostal, internal mammary and coeliac arteries, supply can also arise from subclavian, innominate and even coronary arteries[5, 9, 10]. Serrated pleural thickening and subpleural parenchymal bands are thought to be a reflection of the transpleural collateral vessels[11].

The parenchymal changes have been less well described. Sakai et al found cystic bronchiectasis, honeycombing and a mosaic attenuation pattern[6]. Interestingly, a mosaic attenuation pattern was also seen in the contralateral lung, which was thought to be due to increased pulmonary blood flow, development of pulmonary hypertension or compensatory hyperinflation[5, 6]. Cystic bronchiectasis and honeycombing may be due to recurrent infections. Sakai et al theorised that another possible cause may be the disruption of bronchial growth when the pulmonary arterial supply is disrupted. In our case there was no evidence of bronchiectasis or honeycombing, however there were multiple parenchymal cysts throughout the right lung, with the largest cluster in the lower lobe. To the best of our knowledge, this finding has only been described in a single case previously, with the origin of the cysts in this case unexplained[5]. However, the possibility of these cysts being part of the congenital lung malformation spectrum is considered. As early as 1987, Clements and Warner theorised that congenital bronchopulmonary-vascular malformations represented a spectrum of conditions arising from an insult to the developing lung. The type of malformation depends on the nature, the timing and the severity of the insult. This concept has been put forward in one form or another by various other authors[12]. Langston expanded on this theory suggesting that an obstruction malformation sequence with secondary pulmonary dysplastic changes is the underlying cause of many congenital bronchopulmonary foregut malformations[13]. In the context of such malformations the role of the radiologist is to "thoroughly describe all imaging findings of congenital lung anomalies rather than try to categorize the lesions by pathologic terminology"[14]. In the case of our patient these cystic parenchymal changes, together with the absent pulmonary artery, may represent part of the continuum of congenital bronchopulmonary-vascular malformations. Alternatively,

since the majority of alveolar septation occurs postnatally and appears highly dependent on branch pulmonary arterial supply[2], these cysts may reflect postnatal ischemic dysplasia following interruption of the pulmonary artery after closure of the ductus arteriosus.

The main symptoms of UAPA are recurrent chest infections, dyspnoea and reduced exercise tolerance and haemoptysis[9]. Our patient had reported recurrent chest infections that were managed on an outpatient basis with oral antibiotics. Although the echocardiogram was normal, she has a risk of developing pulmonary hypertension in the future. This is especially of concern as she reaches child-bearing age. In their analysis of 108 cases of isolated UAPA, Ten Harkel et al. found that 44% of patients had pulmonary hypertension[9].

Management depends on the age at presentation, symptoms and associated cardiac abnormalities. Patients presenting early enough may benefit from surgical intervention to revascularize the side of the absent artery and maximise postnatal lung growth[2]. Life threatening hemoptysis may be treated with embolization[15]. Pulmonary hypertension is treated either pharmacologically[4] or with surgical revascularization[9].

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Intra-articular osteoid osteoma of the elbow mimicking juvenile inflammatory arthritis.

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ABSTRACT

Osteoid osteomas are common benign bone tumours typically occurring in adolescence. When the classic symptoms and characteristic radiological features are present the diagnosis of osteoid osteoma is usually straight-forward. When the location is unusual, the diagnosis can be more challenging with non-specific and sometimes misleading symptoms and imaging findings which can mimic inflammatory and infective diseases. This can lead to a significant delay in diagnosis and inappropriate treatment.

We present a case of an intra-articular osteoid osteoma of the elbow which was initially mistaken for juvenile inflammatory arthritis and illustrate the subtle diagnostic features which can be easily missed unless there is an index of suspicion.

INTRODUCTION

Osteoid osteomas are relatively common and account for 10 - 12% of benign bone tumors [1]. They can present in any bone but have a predilection for the metadiaphyseal regions of the long bones, most commonly in the proximal femur. The location can be broadly divided into extra-articular and less commonly intra-articular. Intra-articular lesions are those that are located at the end of long bones surrounded by or close to the joint capsule [2]. Although rarely being reported, in some published series the intra-articular lesions account for up to 10-13% of osteoid osteomas [3].

We present a case report of a 15 year old female who, due to non-specific clinical signs and subtle imaging features, had a delayed diagnosis of intra-articular osteoid osteoma. We describe the circumstances when it is important to consider osteoid osteoma in the differential diagnosis and demonstrate the subtleness in identifying the hallmark osteoid osteoma nidus.

CASE REPORT

A 15-year old female presented to the paediatric rheumatologist with a 12 month history of isolated history of mono-articular left elbow pain, swelling and joint stiffness.. There was joint effusion with significantly limited and painful range of elbow movement, and tenderness of the joint line. There was no other evidence of arthritis elsewhere . Her past medical history was unremarkable and there was no significant travel history or relevant infectious contacts. Her father had a history of psoriasis. Her pain was localised to the distal humerus. She was ANA positive (1:1280), rheumatoid factor and HLA B27 were negative. Inflammatory markers were normal.

She had an external x-ray which was normal with no joint effusion or bony abnormality. She then proceeded to a non-contrast MRI which demonstrated a significant joint effusion, synovitis, and marrow oedema localised mainly to the distal humerus and also the proximal ulnar. The appearances suggested a diagnosis of an inflammatory juvenile-type arthropathy.

In the absence of an alternative diagnosis, Juvenile Idiopathic Arthritis was considered as possible diagnosis despite the uncommon presentation of monoarthritis of the elbow in this disease. Initially the attempt of two intra-articular steroid (triamcinolone hexacetonide) injectios was of some benefit but this was short lived. Trials of multiple different non steroidal anti inflammatory drugs proved ineffective. Repeat intra-articular steroid injection provided no clinical improvement. The patient was then lost to follow up for 8 months, during such time interval an alternative medical approach was sought.

When she returned, her signs and symptoms were unchanged with significant limitation of elbow movement

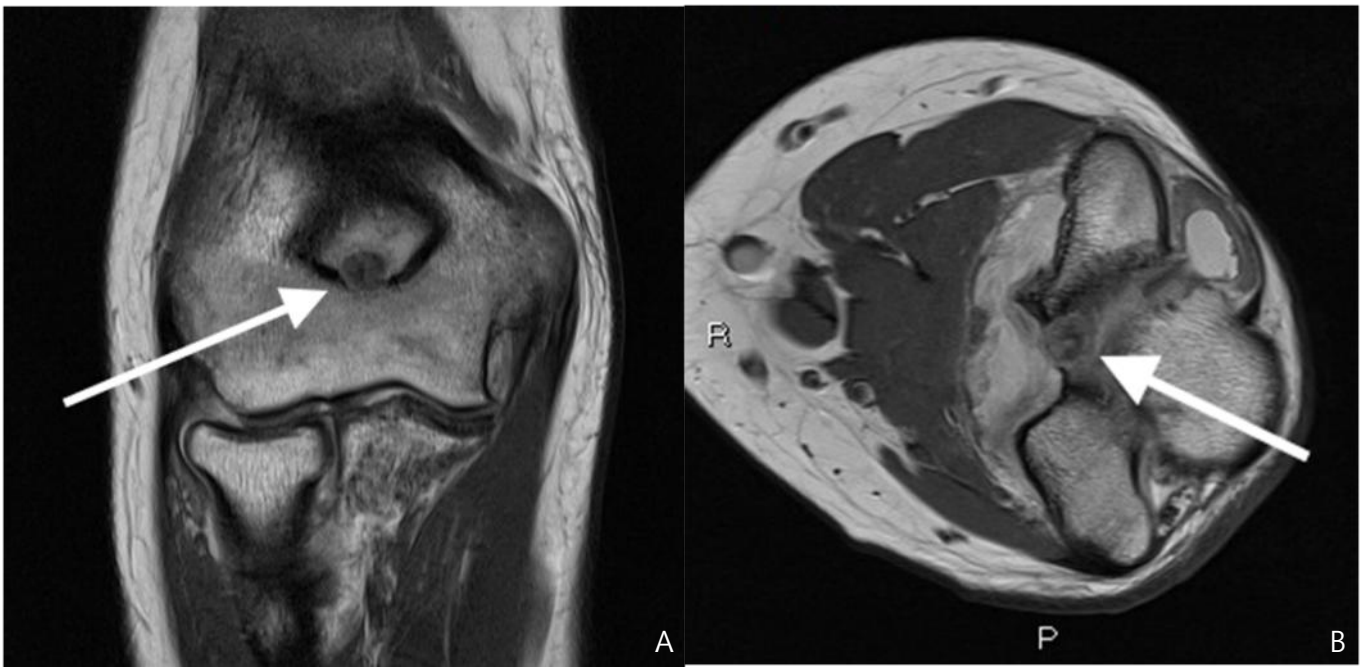

Figure 1. MRI of the left elbow (A) Proton density coronal and (B) proton density axial images show a 8 mm nidus (arrows) in the region of the olecranon fossa of the distal humerus with large joint effusion and synovitis.

with pain and tenderness of the joint line and of the distal humerus. There remained no other joint disease. Repeat MRI at our institution, this time with contrast, confirmed an osteoid osteoma with an 8mm enhancing nidus in the distal humerus in the region of the olecranon fossa. There was persistent distal humeral bone marrow oedema, joint effusion and synovitis [Figure 1 and 2].

DISCUSSION

Osteoid osteoma is a painful benign bone tumour that typically occurs in young patients aged 10 to 25 years, with males affected more frequently than females. Jaffe in 1953 described the osteoid osteoma as composed of two components: firstly, a core or 'nidus' composed of an interwoven network of osteoid trabeculae and richly

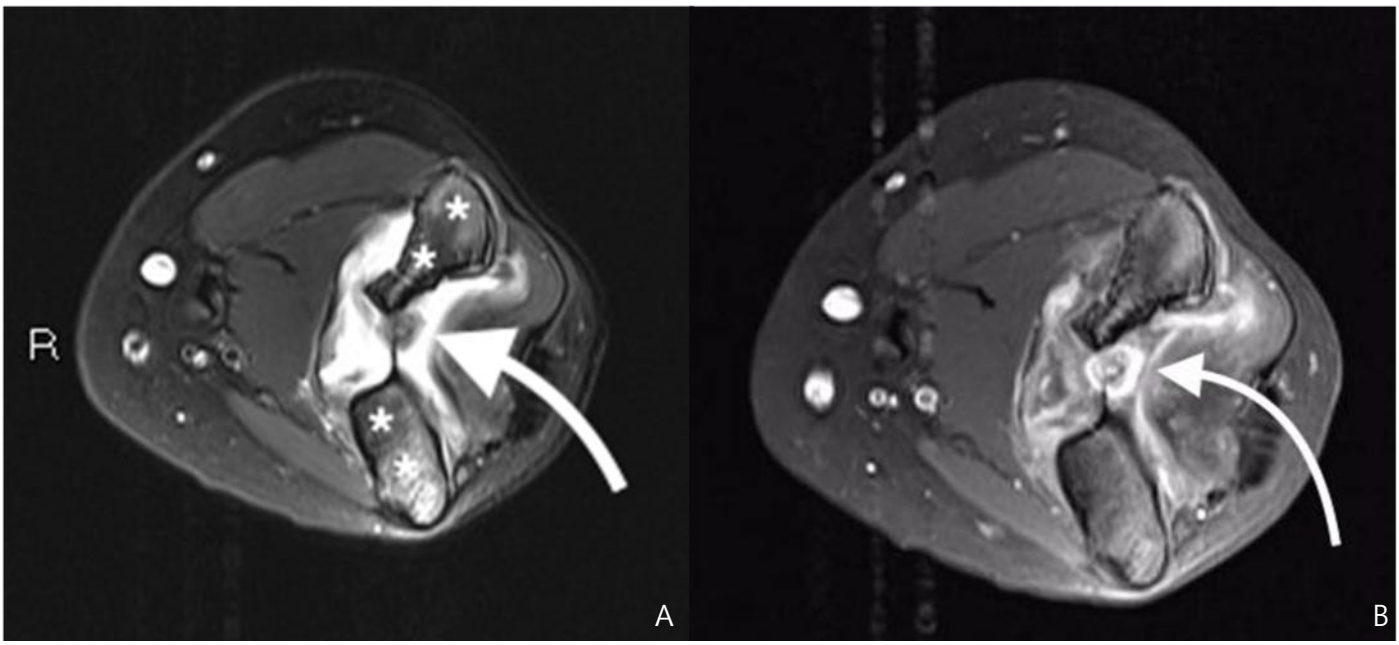

Figure 2. MRI left elbow axial images (A) T2 fat-suppressed shows the 8mm distal humeral osteoid osteoma nidus (arrow) and surrounding bone marrow oedema (*) with large elbow joint effusion. (B) T1 fat-suppressed with contrast show the enhancing nidus (arrow) and synovial enhancement.

vascularised connective tissue and secondly, the surrounding reactive perifocal sclerosis [4]. An osteoid osteoma on plain radiography therefore usually presents as a characteristic discernible lucent nidus that is less than 2 cm and may contain central mineralisation surrounding by sclerosis or periosteal reaction.

It is thought that the nidus releases prostaglandins which elicit a strong pain response. Thus classically, the patient presents with a history of night pain relieved with aspirin and non-steroidal anti-inflammatory drugs which suppress the synthesis of prostoglandins. This characteristic presentation most commonly occurs with extra-articular osteoid osteomas. Presenting as a separate clinical entity, the intra-articular lesions can have a more variable clinical manifestation with non-specific arthralgia, limited range of movement and swelling [5]. As a result, the intra-articular osteoid osteomas are often diagnostically challenging.

Intra-articular osteoid osteomas can occur in any joint, although the most commonly reported sites are the hip, ankle and elbow [3]. When present in the elbow, they tend to be located in the distal humeral epiphysis in the region of the olecranon fossa. The intra-articular osteoid osteoma is often occult on plain radiography. This is because the lucent nidus can easily be overlooked as in contrary to the more common cortical based extra-articular lesion, the intra-articular lesions are often intramedullary or subperiosteal. In this location they often lack the perifocal sclerosis which render lesions more discernible.

Osteoid osteomas are imaged with nuclear medicine bone scintigraphy, CT and/ or MRI. Although bone scintigraphy has been shown to have a high sensitivity, CT and MRI are more specific. CT can accurately identify the nidus and surrounding sclerosis however, radiation exposure remains a consideration and recently MRI has been increasingly used. On MRI, the features are of a nidus, synovitis, joint effusion, bone marrow and soft tissue oedema. The nidus has low to intermediate signal on T1 weighted and proton density sequences and can have low or high T2 signal intensity. The nidus is often difficult to discern against the background of perilesional and marrow oedema. Contrast administration can improve the conspicuity of the nidus as demonstrated in our case.

If the nidus is overlooked, the MRI features remain non-specific with marrow oedema, synovitis and joint effusion, which are often mistaken for more aggressive bone lesions, inflammatory arthritis, osteomyelitis and septic arthritis. Therefore, in the presence of synovitis as in our case, the predilection of marrow oedema to a single bone rather than oedema on both sides of the joint should alert the radiologist to search for a bone lesion or consider osteomyelitis rather than an inflammatory arthropathy.

The natural history of osteoid osteomas is poorly understood. Surgical excision has conventionally been the management option both for histological confirmation and curative treatment. Surgery however, is not without its morbidity. Minimally invasive techniques with percutaneous radiofrequency ablation have also been increasingly utilised. As such, many lesions are treated based on presentation and imaging findings with no histologic proof. Additionally, a small number of reports indicate that some osteoid osteomas spontaneously involute and hence there has been a recent movement towards conservative management with analgesia and hence, greater reliance on accurate imaging [6] [7].

CONCLUSION

Unlike extra-articular lesions, the intra-articular osteoid osteoma can be diagnostically challenging with a protean presentation and imaging features. Our case illustrates that without an index of suspicion, the hallmark nidus can often be missed on both plain radiography and MRI leading to misdiagnosis and inappropriate treatment. Therefore, in cases of persistent unexplained elbow pain and evidence of synovitis, especially when bone oedema is confined to one bone, an osteoid osteoma should be considered particularly in the paediatric / adolescent population.

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