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CONTENTS

Welcome Note
Hamzaini Abdul Hamid

Complicated Pneumonia: Imaging Algorithm
Che Zubaidah Che Daud

3D reconstruction CT in the evaluation of Fetus-in-Fetu
Fauziah Handar, MD, Che Zubaidah Che Daud, MD, Hasni Hamzah, MD, Hamzaini Abdul Hamid, MD, Faizah Mohd Zaki, MD, Mughni B. Bahari,MD, Azlan Mahmud,MBBS.

Case Series of Magnetic Resonance Imaging (MRI) In Pediatric Central Nervous System Tuberculosis.
Emilia RMR, Shahizon AMM, Noryati M, Hamzaini AH, Faizah MZ

QUIZ+ANSWER
Faizah Mohd Zaki, Rohazly Ismail, Noryati Muhammad, Hamzaini Abdul Hamid, Erica Hing Yee

Asian and Oceanic Forum for Paediatric Radiology

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Dear AOSPR friends,

On behalf of Malaysian Paediatric Special Interest Group, we are very honored to be given this opportunity to contribute in this 5th AOfPR issue. I am sure most of us agree with me that we had great and memorable memories during Special AOSPR Congress 2016 in Kuching, Sarawak. A part from excellent lectures, the congress also strengthened the relationship between member's countries.

We build a very warm relationship and all of us feel like a family. We pray and hope that this relationship will continue forever, which in turn will give great impact to the diagnosis and treatment of our children.

I would like to thank to all the contributors for this issue of AOfPR and hope these articles would be beneficial to all the readers.

Complicated Pneumonia: Imaging Algorithm

Che Zubaidah Che Daud

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INTRODUCTION

Chest imaging can be used to distinguish between the exudative and organizing phases of pleural effusion. Ultrasound has an advantage over CT scan in the identification and characterization of complicated effusion. However, CT scan gives more information than plain radiographs for complicated pulmonary infections with empyema, pleural effusion or broncho-pleural fistula. Early detection and intervention for pleural complications are critical as it will result in better outcome. Early Video Assisted Thoracoscopic Surgery (VATS) is more cost effective than thoracotomy in the treatment of empyema in children (1).

PNEUMONIA AND COMPLICATED PNEUMONIA

Lower respiratory tract infections are a challenge to clinicians caring children. Pneumonia can be due to bacterial, viral, fungal, mycobacterial and atypical causes. The incidence of specific pathogens differs by age and most common are *Streptococcus pneumoniae*, *Haemophilus influenza*, *Staphylococcus aureus* and *Streptococcus pyogenes*. Potential complications of pneumonia include parapneumonic effusion, empyema, necrotizing pneumonia and parenchymal abscess (1). Parapneumonic effusion is a collection of fluid in the pleural space secondary to pneumonia. Nearly one-half of all children with pneumonia will develop parapneumonic effusion and less than 5% of these effusion progress to empyema. In empyema, there is presence of pus, with polymorphonuclear leukocytes and fibrin. Incidence of empyema complicating community-acquired pneumonia (CAP) is increasing, and causes significant childhood morbidity (2). Progression of pleural effusion to empyema has three stages of evolution. In exudative stage, patient resolves with antibiotic therapy and most do not progress beyond this stage. In fibrinopurulent stage there is bacterial pleural invasion from contiguous pneumonic process, with polymorphonuclear cell accumulation, fibrin deposition, membrane formation and developing compartmentalization or loculation. In this stage the biochemistry of pleural fluid will show low pH and glucose with high LDH. If the patient is not adequately treated it will further progress into organization stage. In this stage, fibroblasts grow into the exudate from the visceral and parietal pleural surfaces, form an elastic membrane, and may encase the lung with potential to restrict respiration (1).

IMAGING OPTIONS

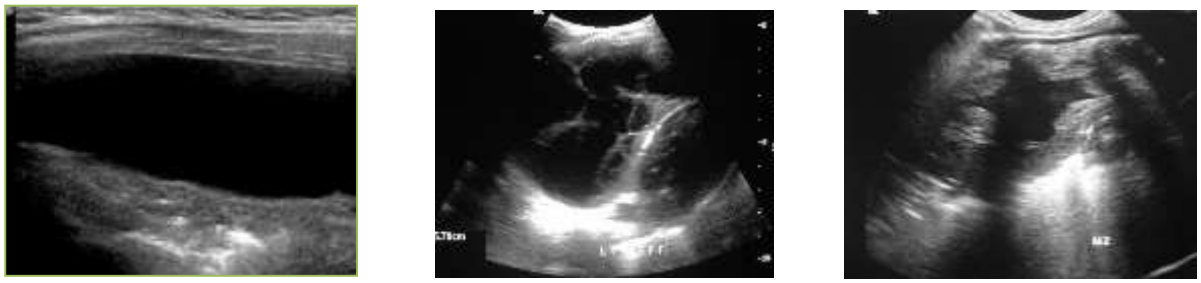
Imaging in complicated pneumonia include chest radiographs (CXR), ultrasound (US) and computed tomography (CT) scan. CXR is easily available and inexpensive. It can be done as portable or in the department with vertical or horizontal beam. It uses a low radiation dose, no sedation and no special preparation is required. CXR is sensitive and highly specific for the diagnosis of CAP. However, it has a limited value in differentiation between viral and bacterial lower respiratory tract infection. It is the standard first line imaging tool for suspected pneumonia. Frontal views are most useful when lobar pneumonia is present, with sensitivity and specificity of 100%. The presence of streaky or confluent opacity gives the frontal CXR a sensitivity and specificity of 85% and 98% respectively (1).

CT scan has high resolution and soft tissue contrast. Multiplanar and 3D reconstruction images make it superior than CXR in complicated pneumonia. However, the radiation dose is 100 times more than a CXR. Intravenous (IV) contrast and sedation are usually needed. The advantage of CT scan is its ability to demonstrate parenchymal abnormalities better than CXR. The disadvantage is its unability to detect complexity of pleural fluid compared to ultrasound (3). Donelly and Klosterman reviewed 56 patients with complicated pneumonia not responding to treatment, where CT results were compared to CXR findings performed earlier on the same day. CT scans were evaluated for cavitary necrosis, cavitation, abscess, loculated effusion, pericardial effusion, bronchial obstruction, broncho-pleural fistula and malpositioned chest tube. All 56 CT scans demonstrated at least one of the above findings that were not seen on CXR. A total of 110 findings were seen on CT. There was an average of two findings per CT scan, 40 with parenchymal complications and 37 with pleural complications. Another retrospective analysis of 17 children who underwent both CT scan and CXR shows that cavitary necrosis is often seen on CT before or in the absence of findings on chest radiography. CT assists in providing a global assessment, demonstrate size, possible loculations, and extent of lung involvement (abscess or necrosis) (4). Donnelly et al. performed a retrospective review of 30 patients who received chest CT with subsequent pleural fluid analysis. CT characteristics of a parapneumonic effusion do not readily distinguish empyema from a transudative process (5,6).

Chest imaging can be used to distinguish between the exudative and organizing phases of pleural effusion such as atypical layering pattern, complexity and loculations (3). US has more advantages over CT in identification and characterization of complicated effusion. It is helpful in both prognostication and treatment decisions. It can be done in the department or bedside. There is no radiation, contrast or sedation needed. It permits visualization of the lesions in real time and in different planes. In a retrospective analysis of 46 pediatric patients with empyema, Ramnath and colleagues show that early sonographic evaluation of parapneumonic effusions led to decrease hospital length of stay for high-grade effusions (7).

STAGES OF PARAPNEUMONIC EFFUSION

Stages of Parapneumonic Effusion	**Duration**	**Descriptions**
Exudative (Stage 1)	24-72 hours of illness	Pleuritis and inflammation Simple Exudative clear fluid
Fibrinopurulent (Stage2)	7 – 10 days of illness	Complicated pleural effusion or pus fluid Deposition of fibrin clots or membrane Septation or Loculation
Organizing (Stage 3)	2-4 weeks of illness	Fibroblast grows on parietal and pleural surface causing thickening of pleural surface. Intra-pleural fibrin membrane transforms to web.



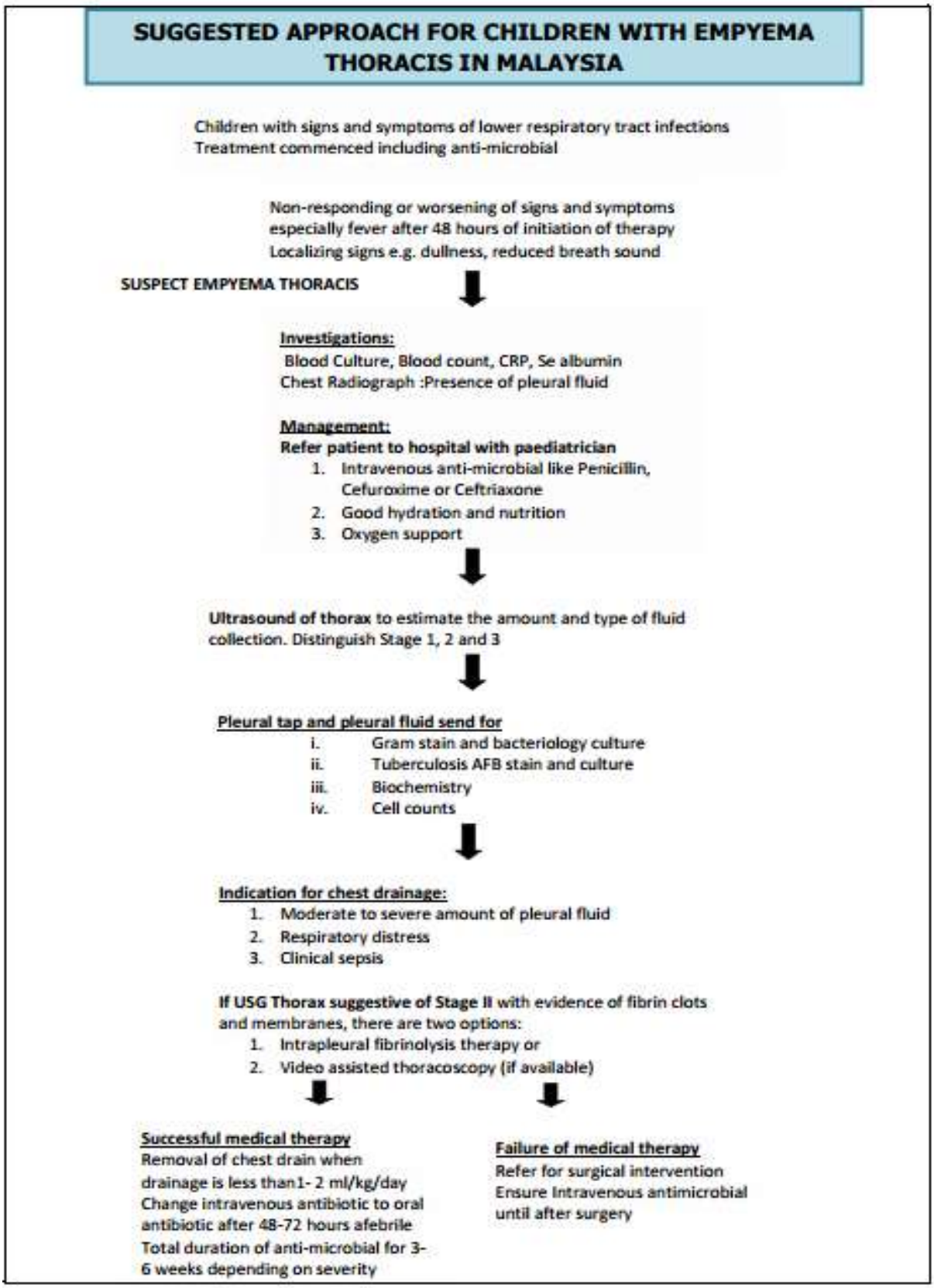
A B C

Figure 1. (A) Stage 1 parapneumonic effusion with clear pleural fluid. (B) Stage 2 parapneumonic effusion with more echogenic fluid and presence of septation. (C) Stage 3 parapneumonic effusion with echogenic fluid and very thickened of pleura.

Early detection of parapneumonic effusion by imaging with subsequent early intervention will result in better outcome. There are no statistical differences in the use of single chest tube placement or repeated thoracocentesis (2). There is a role of fibrinolysis when there is presence of loculations and fibrin development. Chest tube or pigtail insertion depends on the availability and on case-to-case basis.

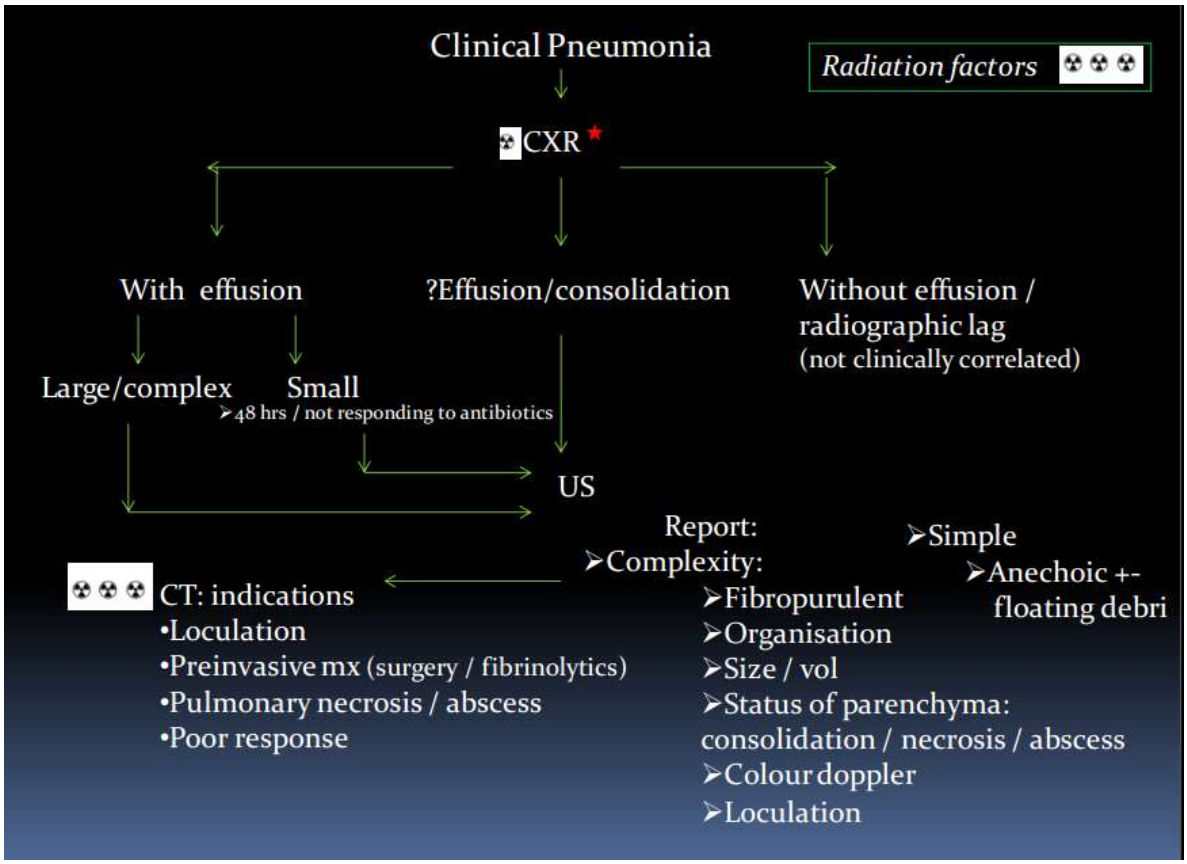
Surgery, either with VATS or open thoracotomy is done in cases not amenable to minimally invasive image-guided procedure (1).

We are presenting a Malaysian consensus guideline for children with empyema thoracis. The consensus was made during the Meeting of Paediatric Empyema Working Group 2013 in Kuala Lumpur



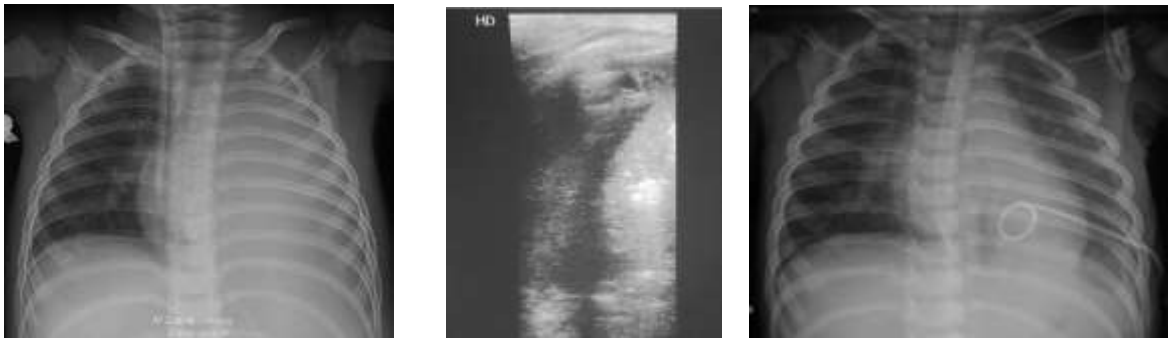
Algorithm of imaging in pneumonia and its complication

The consensus was made during the Meeting of Paediatric Empyema Working Group 2013 in Kuala Lumpur



CASE SCENARIO 1

One-year-old boy diagnosed with Hemolytic Uremic Syndrome complicated by acute renal failure secondary to Streptococcus pneumonia.

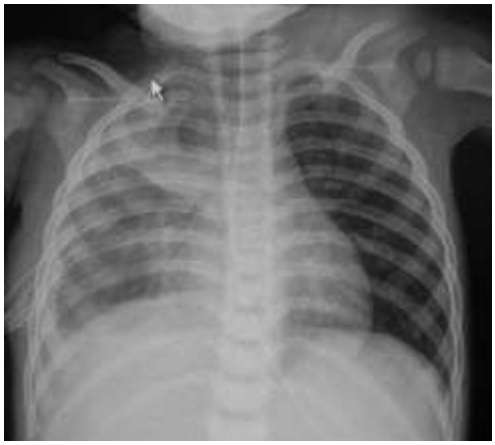


A B C

Figure 2. (A,) Chest radiograph shows massive left pleural effusion. (B,) Ultrasound demonstrates stage 2 parapneumonic effusion hence pigtail drain with fibrinolysis agent was introduced (C).

CASE SCENARIO 2

One-year-old girl with respiratory symptoms for 3 days, worsening and needing intubation.



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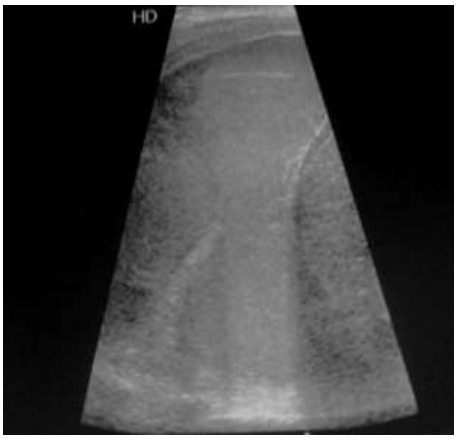
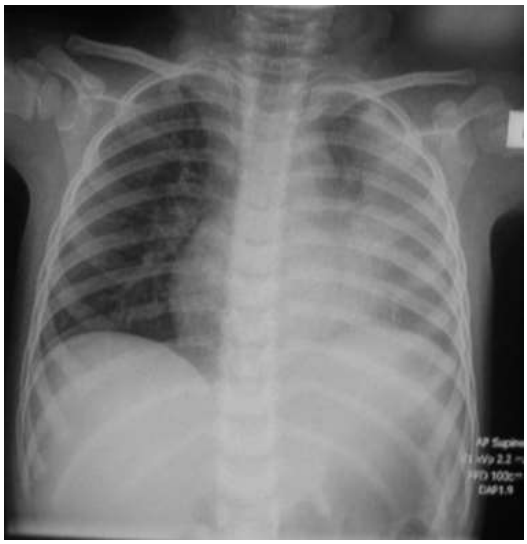


B

Figure 3. (A.) Chest radiograph show presence of right pleural effusion. (B,) ultrasound demonstrates stage 1 parapneumonic effusion. Chest drain was inserted without fibrinolysis agent.

CASE SCENARIO 3

A three-year-old boy with respiratory symptoms for six days.



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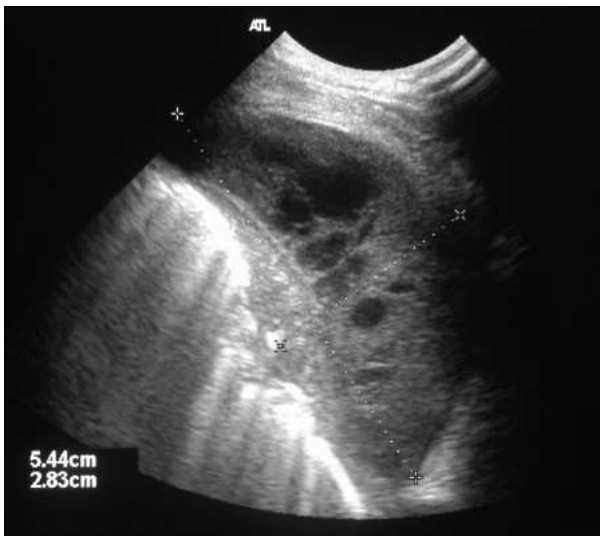

B

Figure 4. (A) Chest radiograph shows left pleural effusion. (B) Ultrasound demonstrates loculated stage 3 parapneumonic effusion. Thoracotomy and decortication was performed.

CASE SCENARIO 4

A five-year-old girl, two weeks post-splenic trauma had persistent tachypnea and fever.

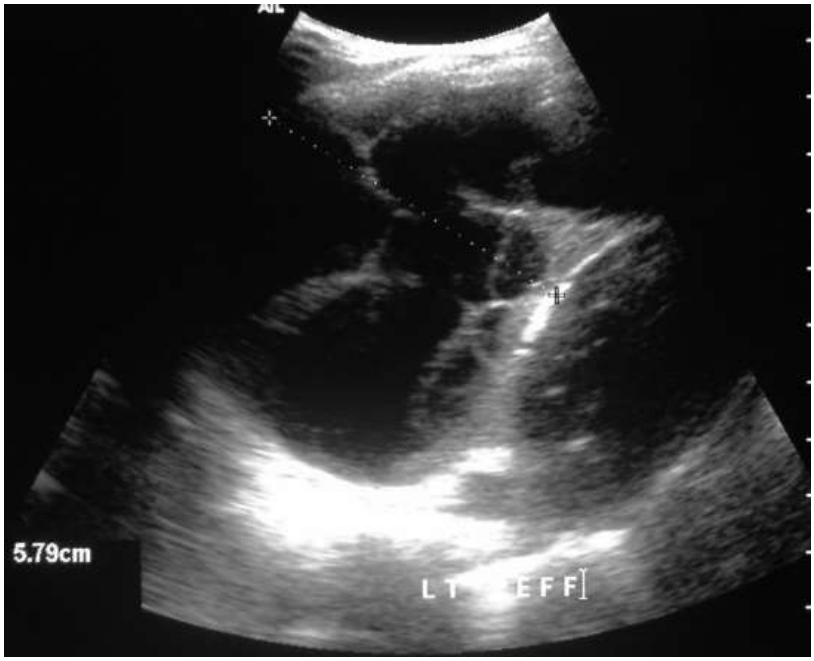

Figure 5. Ultrasound shows stage 2 parapneumonic effusion. Pigtail drain was introduced and fibrinolysis agent was given.

CONCLUSION

CXR is very sensitive and specific in the diagnosis of community-acquired pneumonia. Knowledge of the stages of parapneumonic effusion on US is very important. US has an advantage over CT in identification and characterization complex effusion. Good US with color Doppler can detect parenchymal necrosis. CT should be used judiciously in view of its ionizing radiation. Early detection and early intervention will result in better outcome. Chest drains either chest tube or pigtail (depending on availability) play an important role in stage 1 and stage 2 effusions. Fibrinolysis agent has a role in the management of parapneumonic effusion in late stage 1 and early stage 2. The role of surgery is for VATS in early stage 2 and open thoracotomy in stage 2, 3 or not responded to intrapleural fibrinolysis.

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3D reconstruction CT in the evaluation of Fetus-in-Fetu

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Abstract

Fetus in fetu (FIF) is an unusual medical entity with an incidence of 1 in 500000 births. It is due to abnormal embryogenesis in a diamniotic monochorionic twin pregnancy and it may grow from any tissue layer at various locations in human body, with the abdominal cavity being the most commonly involved. Diagnostic capabilities of the multi-planar 3D reconstruction images of multislices computed tomography (CT) has added superior value in diagnosing this rare pathology as CT further characterizes the soft tissue, bony component as well as the vascular supply of this entity. We presented a case of fetus in fetu and discuss on the value of multimodality imaging particularly utilizing 3D post-processing reconstruction CT technique in order to obtain definitive diagnosis in this case.

Keywords: Fetus-in-fetu (FIF), 3D reconstruction imaging CT

Introduction

Fetus in fetu (FIF) is a well-known rare entity in medical field. The rarity is presumed as 1 over 500000 births. It is due to abnormal embryogenesis in a diamniotic monochorionic twin pregnancy defined as a mass arranged on an axis around a vertebral column by Willis in 1958 [1].

In current radiology practice, CT reconstruction rendering technique has been proven to add diagnostic value in medical literature. With the utilization of the volumetric data acquisition and variable technique of volume image rendering, it improves the anatomic details and thus giving more information on the cases preoperatively [2]. Therefore, this report highlights the diagnostic contribution of 3D imaging technique in this unique and rare case.

Case report

An 18-month-old boy presented to clinic. He was found to have increasing abdominal size by his mother for 2 weeks. He was born at term via emergency caesarean section due to oligohydromnios with good birth weight and normal postnatal development. Clinically, he was afebrile, the abdomen was distended with no sign of peritonitis. There was neither ascites nor jaundice detected. Rest of the clinical history and examination were unremarkable.

Tumor markers which include Beta-HCG and Alpha feto-protein were normal. Other blood parameters showed normal blood count, (Hemoglobin : 11.4 g/l, white cell count of $12.4x10^9/L$ and platelet of $300x10^9/L$). Liver function test was normal. Urinalysis and renal profile were also unremarkable.

Initial ultrasound imaging showed a complex multi-cystic lesion at porta hepatic region (Figure 1) with no biliary dilatation, which raised the suspicion of non-obstructive choledochal cyst. Multiplanar 3D CT scan of the abdomen further showed a large complex retroperitoneal mass multiple limb-like structures/ organs resembling a fetus (Figure 2-6). Intra-operative finding showed the tumor in retroperitoneal region, adhered to the duodenal mesentery and head of pancreas, stretching the proximal duodenum D1/D2 as well as the superior mesenteric vein. Biliary system was normal. Histopathology confirmed the diagnosis of mature teratoma composed of three germ layers of ectoderm; bone, cartilage and fatty tissue from mesoderm and intestinal mucosa from endoderm layer. The cyst wall was ruptured and associated with foreign body granulomatous inflammation. Patient was re-admitted 5 day after discharge due to sepsis and hemorrhagic pancreatitis. Re-laparotomy done and confirm the diagnosis and adhesiolysis was performed. Subsequently, he improved and was discharged home well.

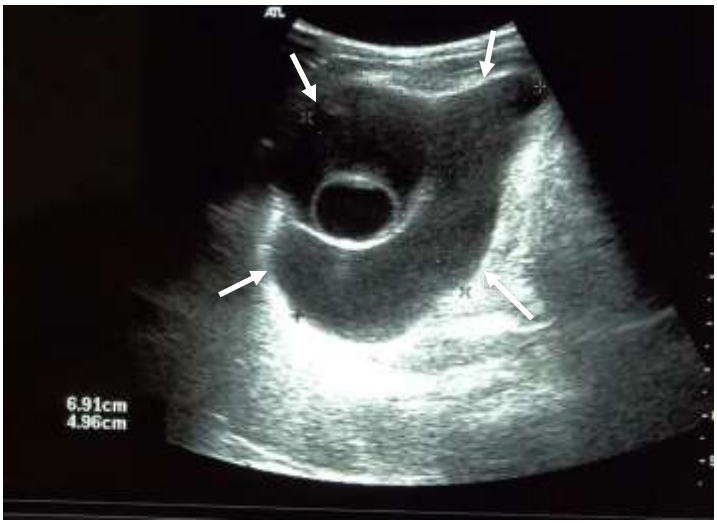

Figure 1. Axial USG shows multiseptated cystic lesion (white arrows) at right upper abdomen

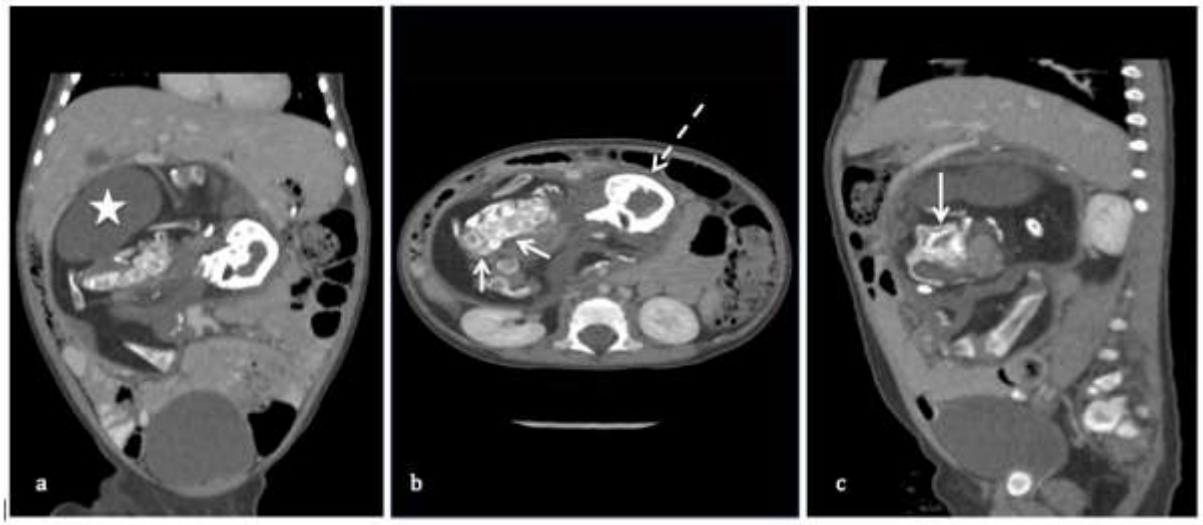

Figure 2. a) Coronal, b) axial and c) sagittal CT abdomen images show a complex mass with variable soft tissue density within its own cavity. Bony component resembling skull (white dotted arrow) and cystic lesion arising from adjacent vertebra-like structure (white arrows); which is presumed to represent a myelomeningocele (white star), but turns out to be granulomatous inflammation.

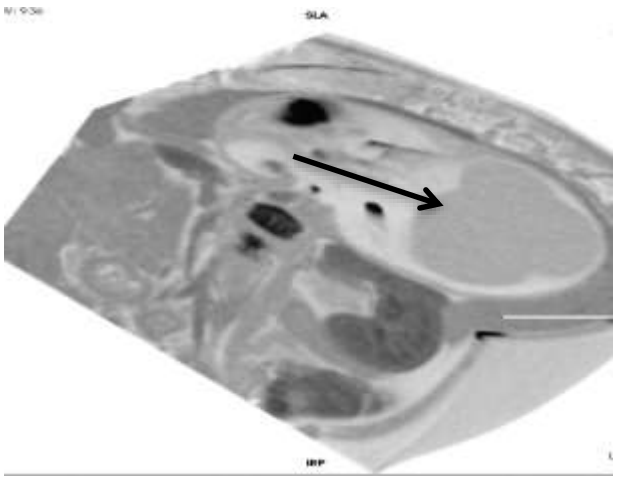

Figure 3. 3D volume rendering technique show the lobulated cystic lesion (black arrow) at right upper abdomen/ suprarenal region as previously depicted in ultrasound.

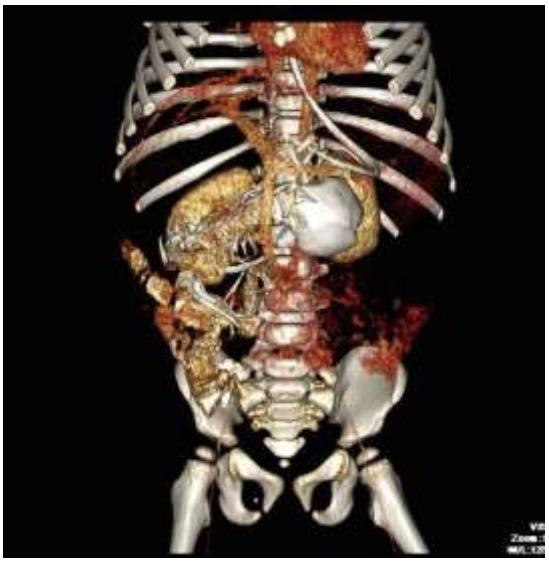

Figure 4. Shaded surface display (SSD) rendering technique with advanced software enhances the appearance of the bony structure of fetus-in fetu.

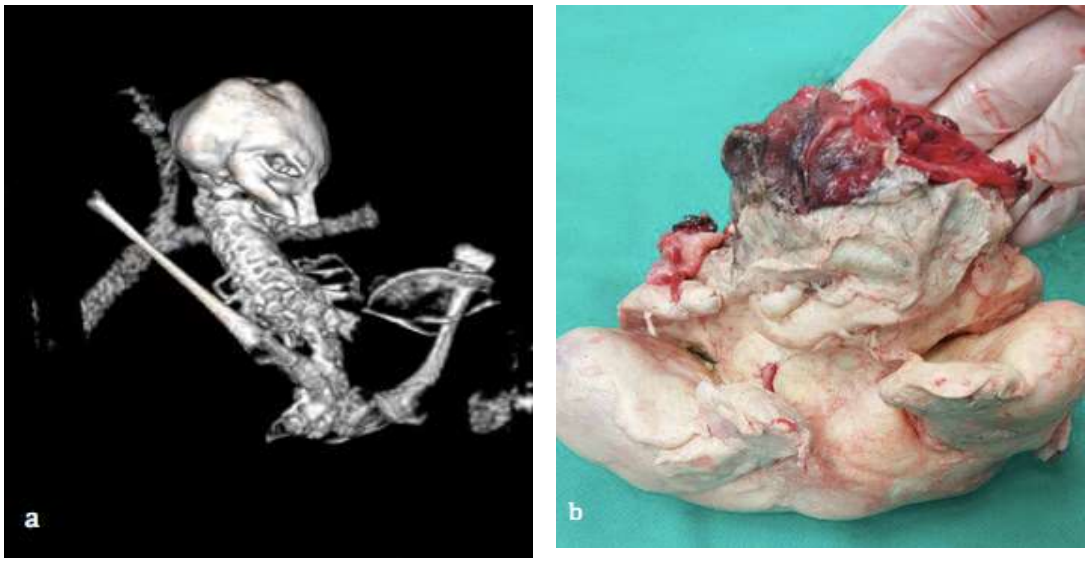

Figure 5 a) 3-Dimensional bone reconstruction view of the tumor after 3D processing showing the orientation of the long bones and spine. b) Corresponding gross pathology of the tumor excised intra-operatively.

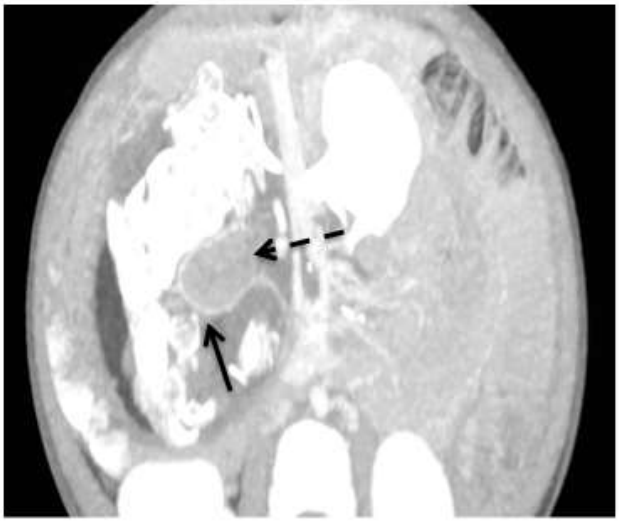

Figure 6: Maximum intensity projection (MIP) images processing shows a long tubular structure with similar density to the main vessel (black arrow), most likely representing one of the blood supplies to the fetiform tumour with homogenous enhancement at central part of the fetiform (dotted black arrow).

DISCUSSION

Fetus in fetu (FIF) is a rare anomaly. This entity was historically discovered by Lord in 1950 [3]. Current diagnosis of FIF is made based on Willis in 1958 with the presence of indubitable vertebral column which requires radiographic and dissectional evaluation to support the diagnosis[1]. The complexity of this anomaly is also believed to be a form of matured teratoma or a fetus-like containing structure labelled as parasitic twin as it grows inside the parent body[2].

The clinical presentation of FIF is always non-specific, according to the location it grows. It had been reported in oral cavity [4], as an intracranial mass in a 4-month-old boy [5], nevertheless it is commonly found in the abdominal cavity. It can stay harmless and asymptomatic in the body and has been reported to be detected as an incidental finding after 45 years of life [6]. Most reportable cases are benign and completely curable post-operatively. However, there is rare occasion where FIF was reported to cause recurrence and malignant transformation [7].

The most practical radiological method of imaging in current practice for suspected FIF is by performing multiplanar 3D CT imaging of abdomen with intravenous contrast. CT helped to characterize bony and soft tissue component of FIF in this case. In fact, most of the case with intra-abdominal FIF, the diagnosis was not suspected pre-imaging due to its rarity as in this case. This technique has shown better characterization of the multi-component structures resembling a fetus in the retroperitoneal space.

There are multiple types of images rendering in daily radiology practices [8]. These include maximum intensity projection (MIP), minimum intensity projection (MinIP), shaded surface display (SSD), volume rendering (VR) and virtual endoscopy (VE) technique [8,9]. Different tissue compositions have significant difference in radiological densities in the data volume, which allow the computer to generate images of medical diagnostic value. In radiology, every tissue density is recorded as algorithm number and this numerical data will be further explored to generate image mapping for reconstruction and anatomic details [9]. Some of these 3D rendering techniques have been applied in this case to better delineate the anatomic component of the mass which comprises bone, cystic and soft tissue component and possible vascular supply of the FIF which are crucial in the diagnosis of FIF.

Initially our case was technically challenging as the 2D imaging ultrasound depicted cystic mass in the right upper abdomen, whichmade the diagnosis of choledochal cyst as primary concern at that time. Its agreeable as ultrasound has superior quality in differentiating solid and cystic component as the sound waves echoes in two different tissue composition. However, sound waves are not able to penetrate compact tissue like bony structures, resulting in posterior shadowing and further compromise the imaging quality. A dedicated volume rendering 3D image processing revealed the fetiform tumor was enclosed in its own sac. A complex cystic lesion at right upper abdomen was abutting the bony vertebral structure presumably to be a meningocele. The bony component in FIF was overlook by ultrasound as it might have been interpreted as calcification or even gas in adjacent loops of bowel. Therefore, CT has a superb role to distinguish different tissue structures thus improving diagnostic capabilities in this case.

Technically shaded surface display (SSD) was performed to further characterize tissue with specific density such as bones by selecting the Hounsfield range of tissue to be highlighted. The diagnostic ability of SSD depends on its capacity to represent with great detail, selected tissue types in isolation from other surrounding structures [9]. In this case, several well-formed bony structures such as malformed skull, vertebras and a few long bones were visualized which was important in diagnosis of FIF.

Maximum intensity projection (MIP) algorithm is a CT software which enables visualization of the abnormal vessel supplying this FIF. This is also valuable information to the surgeon as it provides preoperative details. MIP technique is able to differentiate the hyperdense structure in relation to surrounding tissue and improve anatomic spatial resolution[8,9]. Therefore, in this case there was a vascular supply from the host artery leading to homogenous enhancement of the soft tissue within the fetiform organ, which was subsequently comfirmed to be bowel component as shown in Figure 6.

Conclusion

FIF though rare, can be diagnosed optimally using advanced cross sectional imaging as it is crucial to identify different structures of FIF namely the skeleton, vertebral column, soft tissue as well as vasculature to reach the diagnosis. Diagnostic capabilities of the multiplanar 3D reconstruction images has added value in diagnosing FIF as it improves delineation of anatomic detail and can better characterize the soft tissue, bony component as well as the vascular supply of the fetiform teratoma.

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Case Series of Magnetic Resonance Imaging (MRI) In Pediatric Central Nervous System Tuberculosis.

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Abstract

There has been increasing trend of CNS tuberculosis in Asian population while children are among the vulnerable subgroup being affected. The purpose of this case series is to illustrate the various magnetic resonance imaging (MRI) findings of the brain and spine in children who had laboratory proven central nervous system (CNS) tuberculosis. We will also discuss the various CNS presentation of tuberculosis in children with positive MRI features.

Keywords:

Tuberculosis, central nervous system, pediatric

Introduction

Pulmonary tuberculosis (TB) is a common disease in our country and also in our neighbouring countries in Asia, affecting both immunocompromised and immunocompetent patients [1]. However, extrapulmonary tuberculosis manifestation in the central nervous system is uncommon and remains the most severe form of TB in children. There has been an increase in the prevalence of CNS TB with children less than 5 years old being most commonly affected. Neurological sequaele such as marked cognitive and motor impairment in CNS TB survivors warrant accurate and prompt diagnosis and treatment which include recognizing the imaging pattern of this disease [1,2]. We aim to highlight three case series demonstrating different clinical presentation as well as imaging pattern of CNS TB among children and discuss the variety of CNS TB presentation based on previous literatures.

Case series

Case 1

A 14-year old girl presented with 3 weeks history of intermittent low grade fever which progressed to right lower limb weakness, requiring walking aid. Power of the ankle flexion and extension was 0/5 while the rest ranges from 3/5 to 4/5. Her chest radiograph showed miliary lesions. Subsequently a CT and MRI brain/ whole spine (Figure 1 & 2) was performed which showed multiple rim enhancing lesions representing tuberculomas in the supratentorial region. CSF lumbar puncture showed positive TB Polymerase Chain Reaction (TB-PCR). Although antituberculosis treatment was commenced, repeated MRI 11 months later showed worsening of these lesions. Patient then underwent craniotomy and excision biopsy of the lesion which showed hyperaemic dura with adhesion around the arachnoid layer to the dura. The lesion appeared rubbery and adherent to the surrounding tissue. Cheesy material was found within the lesion after the removal. Histopathological (HPE) finding was documented as chronic granulomatous inflammation. Child had continuation of the antituberculous treatment and was well until this case report was written.

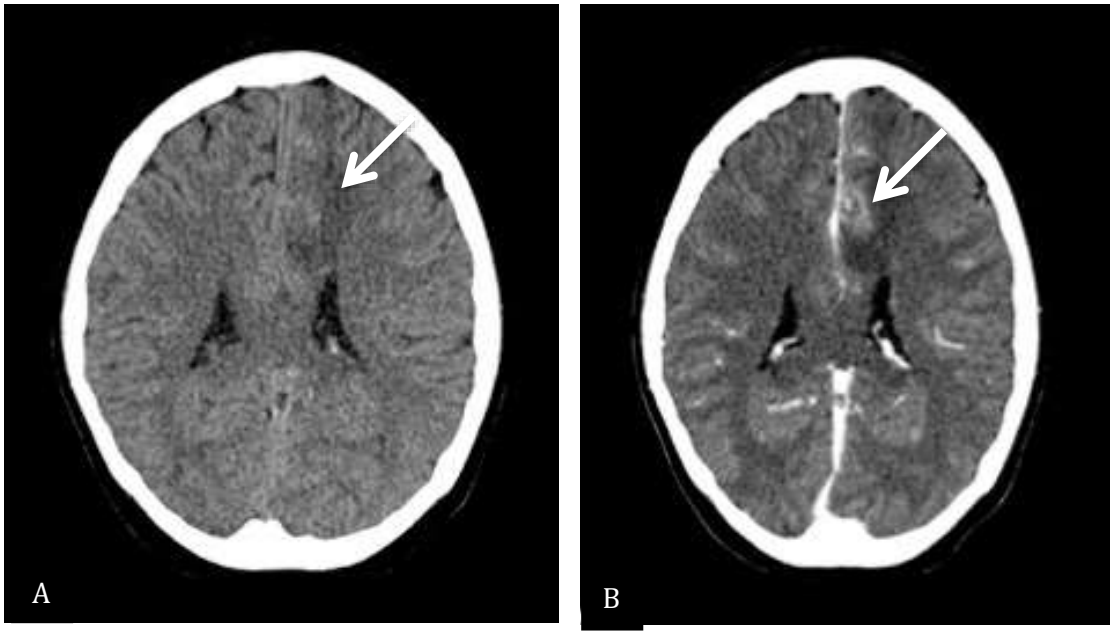

Figure 1. Case 1; 14yo/girl. (A) Non-enhanced CT brain shows an ill-defined white matter oedema in the parasagittal region of the left frontal region (arrow). (B) On post-contrast CT, there is a rim-enhancing lesion at this region on contrasted CT brain (arrow).

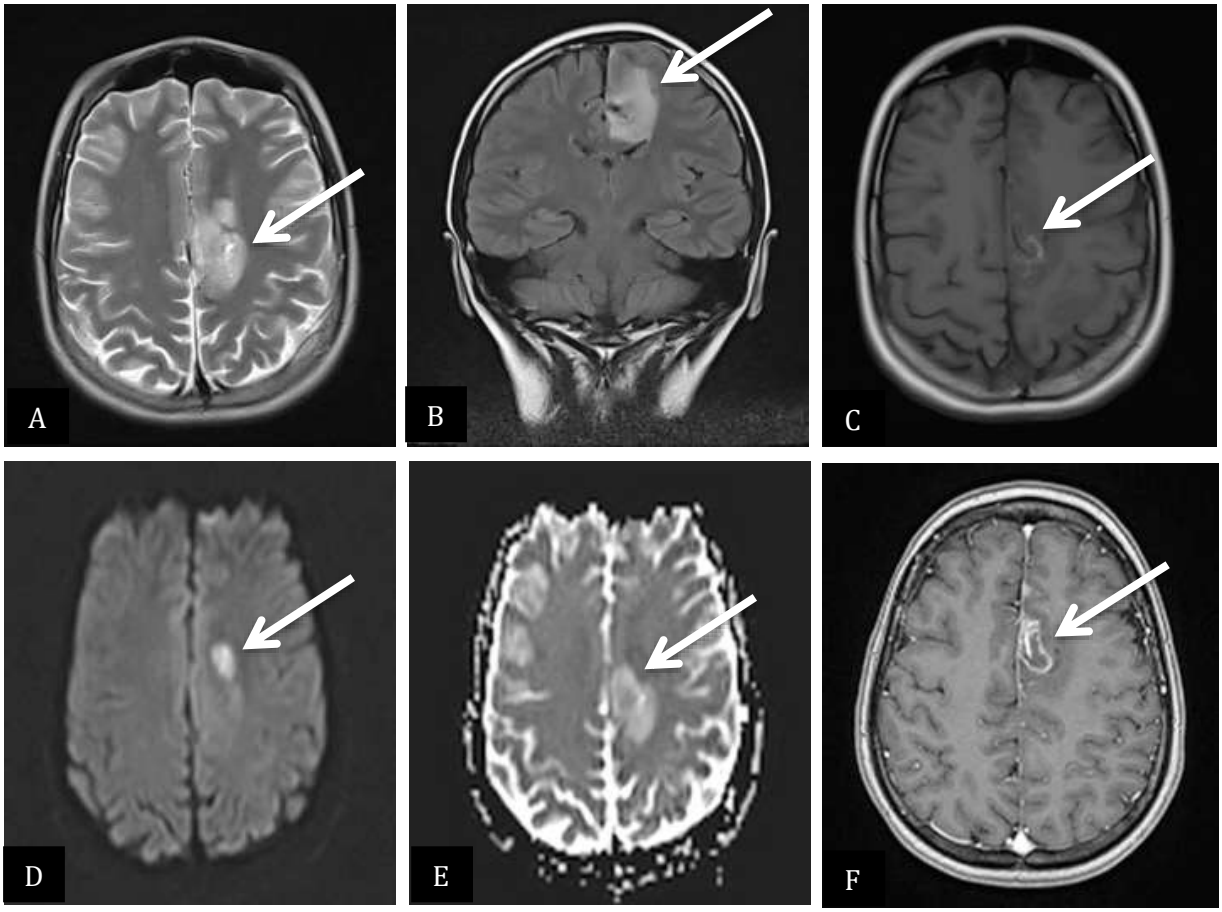

Figure 2. Case 1; 14yo/girl. (A) Axial T2W (B) Coronal FLAIR (C) Axial T1W (D) Axial DWI (E) Axial ADC (F) Axial T1 Post-gadolinium. There is a rim enhancing lesion in the grey white matter - junction at parasagittal region of the left high fronto-parietal region which demonstrates low signal intensity on T1W, high signal intensity on T2W and restricted diffusion on DWI/ADC suggestive of tuberculous abscess (arrow). There is associated adjacent white matter and cortical edema as well as leptomeningeal enhancement

Case 2

A previously well 7 months old infant presented with macrocephaly as well as developmental delay. He was unable to bear weight and could only crawl at 7 months of age. Neurological examinations showed bilateral upper and lower limbs hyperreflexia and slight hypertonia. The powers of all limbs were 4/5 generally. CXR performed was normal. MRI brain showed obstructive hydrocephalus caused by multiple basal and craniocervical junction leptomeningeal enhancing lesions that showed T2 hypointensity and no diffusion restriction. There were also multiple anterior and posterior enhancing intradural lesions in the spinal canal (Figure 3). A

provisional diagnosis of infective process such as tuberculosis was made. The patient underwent posterior fossa craniotomy, lesion excision and ventriculoperitoneal (VP) shunt insertion which showed solitary whitish lesion with some parts having soft consistency while other parts having firm consistency. This lesion was highly vascularised with distinct margins from the surrounding parenchyma. HPE result from the frozen section showed necrotising granulomatous inflammation. He was treated with anti-tuberculous treatment for 9 months and was well after even though he had delayed motor development as compared to his age-match group.

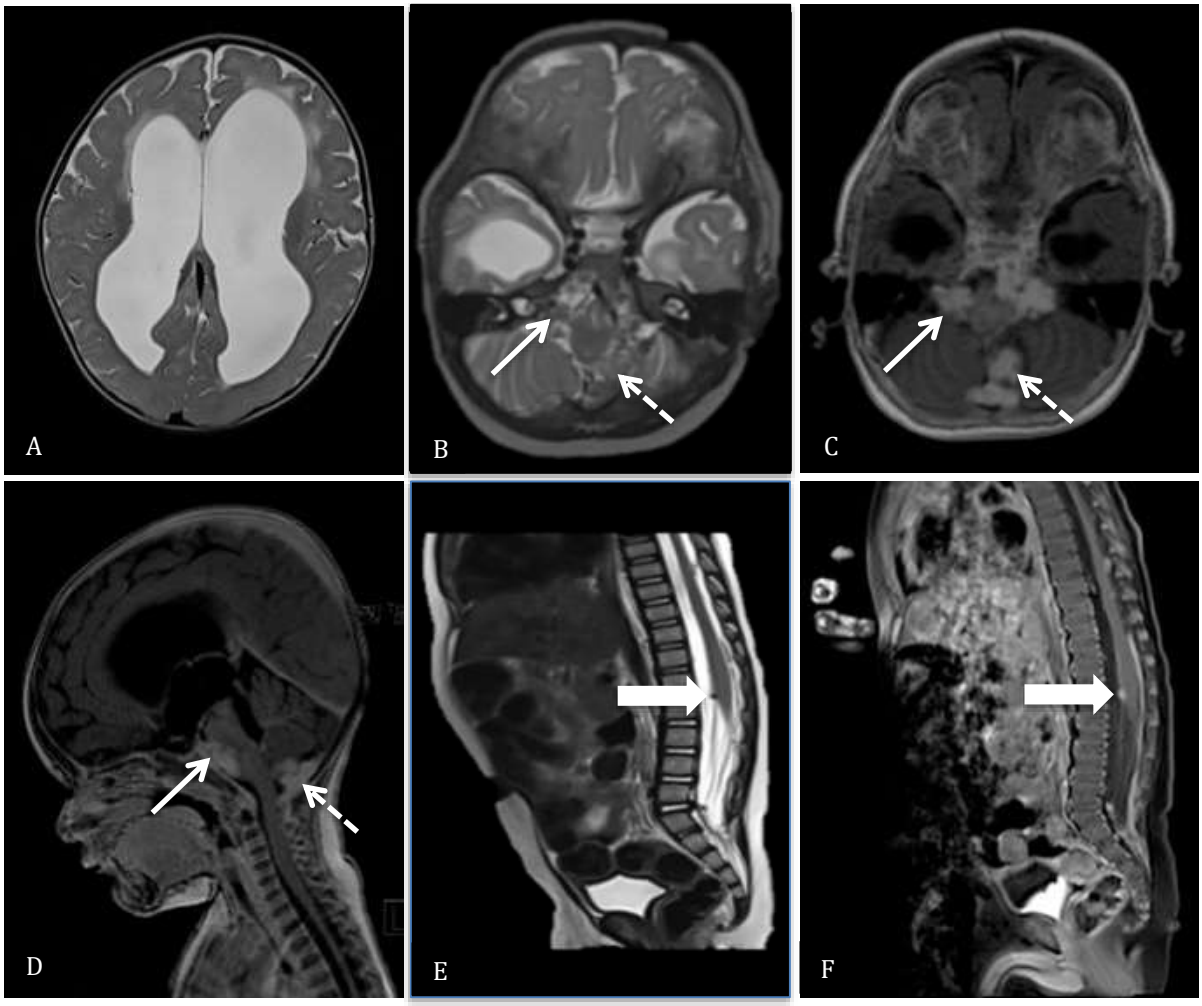

Figure 3. Case 2; 7m/boy. (A) Axial T2W. There is marked hydrocephalus causing thinning of the white matter and associated CSF seepage. (B-D; Axial T2W, Axial T1 Post-gadolinium, Sagittal T1 Post-gadolinium).There are multiple enhancing nodules in the basal cisterns of pre-pontine (arrow) and cisterna magna (dotted arrow) which show T2W hypointensity and enhancing on post-gadolinium indicating leptomeningeal lesions. These result in obliteration of the craniocervical junction and hydrocephalus. (E & F; Sagittal T2W and T1 post-gadolinium). Similar intradural extramedullary nodules are seen in the spinal canal (thick arrow).

Case 3

A 16-year old boy, with no known medical illness came to emergency department with status epilepticus. He complained of headache and fever for the past 1 week prior and denied history of contact with tuberculosis patients. CT brain was performed and showed generalized cerebral edema. Subsequently, MRI demonstrated thick leptomeningeal lesions predominantly in the basal cisterns and both sylvian fissures where tuberculous meningitis was suspected (Figure 4). Cerebrospinal fluid (CSF) culture was positive for Mycobacterium tuberculosis after 6 week. He was treated with antituberculosis and improved initially but later succumbed as he defaulted treatment and follow-up.

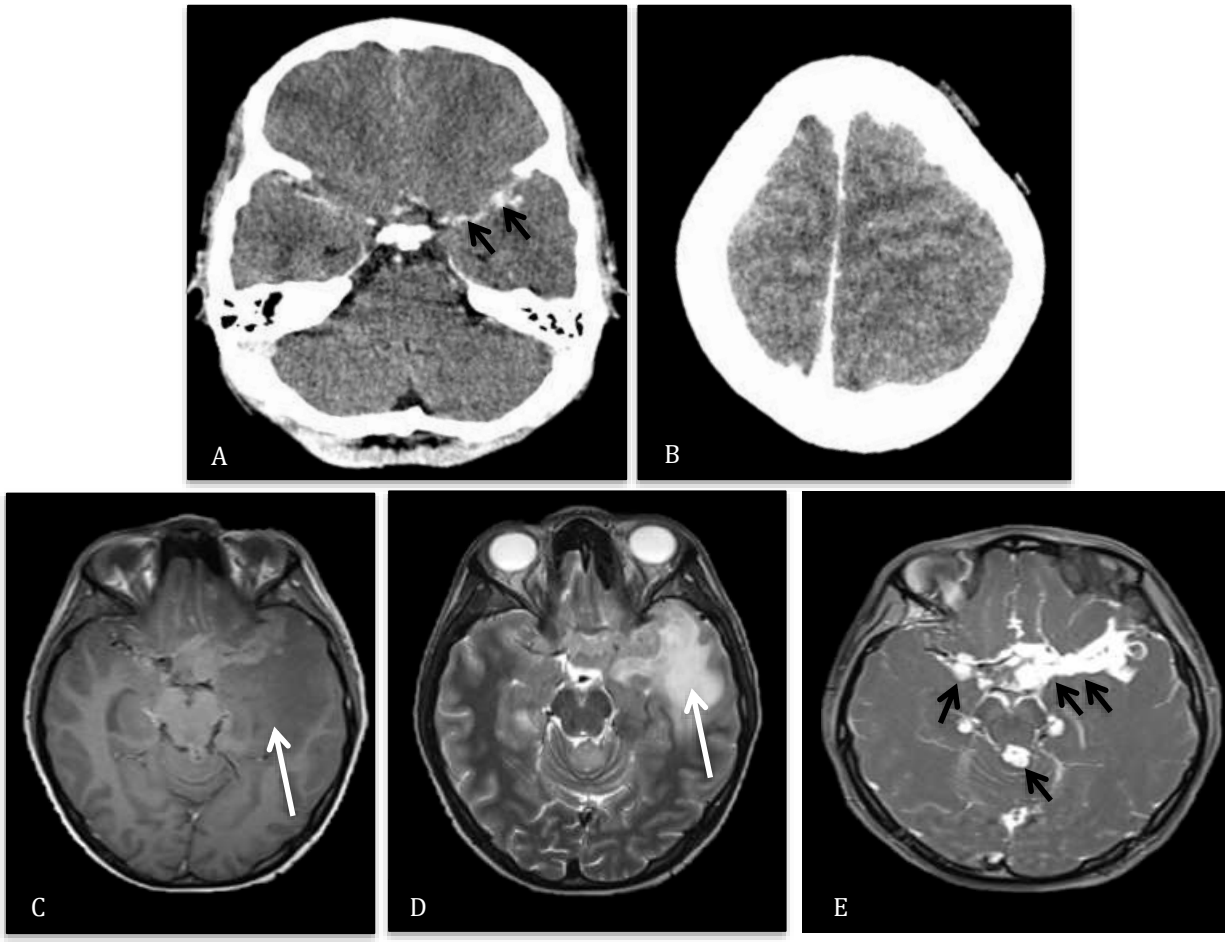

Figure 4. Case 3; 17/boy. (A-B) Contrast enhanced CT brain shows nodular enhancement within the left basal cistern (arrows) with associated generalized brain edema. (C-D) Axial T1W and T2W brain of the child show huge area of white matter edema in left temporal lobe (arrow). (E) Axial T1 post-gadolinium shows thick nodular enhancement of the basal cisterns along the course of middle cerebral artery bilaterally as well as in the ambient cistern posteriorly (arrows).

Discussion

Tuberculosis (TB) is caused by Mycobacterium tuberculosis and it has a high morbidity and mortality each year. TB manifests itself as pulmonary or extrapulmonary lesions. Among all the extrapulmonary manifestation of TB, central nervous system tuberculosis (CNS TB) remains the most severe form in children with prevalence of 10% and an extra 5% higher in immunocompromised patients [1,2].

The clinical manifestation of the disease varies and is non-specific [2]. Central nervous system tuberculosis has a wide range of presentation that may mimic other illness. The most common presentations are headache followed by fever, seizures and altered level of consciousness as well as neurological deficit [3] as seen in our case 1 and 3. These symptoms are associated with hydrocephalus and meningeal lesions seen on imaging. However, our case 2 patient presented with developmental delay and presented at a relatively young age which is known to present with a more non-specific clinical symptom. Previous literature had highlighted that young age and late presentation of the disease is associated with poor outcome which include marked cognitive and motor impairment [4].

In neuroimaging, MR brain is superior to contrast enhanced CT brain as it is able to distinguish tuberculoma from tuberculous abscess as demonstrated in case 1; as well as differentiating with other lesions such as neurocysticercosis and neoplasm [5-7]. MRI is able to demonstrate in more details the enhancement pattern and distribution in leptomeningeal disease as depicted in case 3 when the nodular leptomeningeal enhancement was more marked on MRI as compared to CT. Nevertheless, CT can be the first line of imaging tool especially in diagnosing hydrocephalus and cerebral edema [7]. On the other hand, CT and MRI may be normal at the early stage of the disease which was reported to be up to 11% of cases [3, 5].

Communicating hydrocephalus and basal cisterns enhancement (90%) are the most common findings in CNS TB. Meningeal enhancement is found predominantly involving the interpeduncular fossa, pontine cisterns, perimesencephalic and suprasellar cisterns [5]. Case 2 shows peculiar conventional MRI signal lesions especially on T2W which showed T2 hypointensity that enhances on post-gadolinium. We postulated that this could be due to

evolution of the lesions from active granuloma to fibrosis/calcification, resulting in the enhancement. Sulci over the convexities and of the Sylvian fissures can also be affected. However cerebellar meningeal and tentorial involvement is uncommon. Presence of basilar meningeal enhancement with tuberculoma is highly sensitive and specific for CNS TB [5, 7].

Tuberculosis meningitis (TBM) is the most severe form of CNS TB. 20-45% of TB cases in children are affected by TBM, 9 times higher compared to adults. TBM is common in children less than 5 years old with the mean age ranges from 23 to 49 months old [5,6]. Meanwhile, tuberculoma is the most common parenchymal lesion in CNS TB (40%). It can either be solitary, multiple or military with sizes varying from few millimeters to 8 cm in size. In children, lesions are usually found at the infratentorial region. If the lesion involves the supratentorial region, it is likely seen in the frontal and parietal lobes [5].

Cranial neuropathies affecting cranial nerves II, III, IV and VII are commonly associated with TB. These are partly due to vascular compromise; due to nerve ischaemia and entrapment of nerves by the exudates or by large tuberculomas compression [4].Other rare manifestations of CNS TB include tuberculous abscess, tuberculous encephalopathy, cerebritis, vasculitis as well as infarction [5, 8]. Lenticulostriate arteries are common end arteries affected by TM meningitis, frequently causes bilateral basal ganglia infarctions that are associated with poor prognosis. The middle cerebral artery territories are also commonly affected and are frequently bilateral as seen in our case 3 [8].

It would be advisable to image the spine as well once there are intracranial manifestations suggestive of TB as it can manifest as extramedullary or intramedullary lesions. Our case 2 patient had intraspinal extension of his extensive leptomeningeal disease. Meanwhile, intramedullary tuberculomas (IT) are very rare with 2 in 100000 cases of TB reported at which cervico-thoracic cord is commonly affected [5]. Other than that, TB myelitis or epidural/subdural abscess can occur but less common than the intradural lesion. Another spinal manifestation of TB would be the TB spondylitis which commonly involves thoracic vertebrae. When the vertebrae are involved, there would be associated soft tissue component/ abscess in the majority of cases [5].

Conclusion

CNS tuberculosis is known to be a great mimic and therefore, it can present in various ways not just on clinical presentation but also imaging patterns. Radiologists need to be aware of this and invariably include tuberculosis as one of the differential diagnosis especially when the imaging finding is not typical of certain pathology. Since we live in this part of Asia, where tuberculosis is endemic; it is important to recognize CNS TB at its early stage, understand the different imaging manifestation that can be demonstrated by CNS TB, as it has a high morbidity and mortality.

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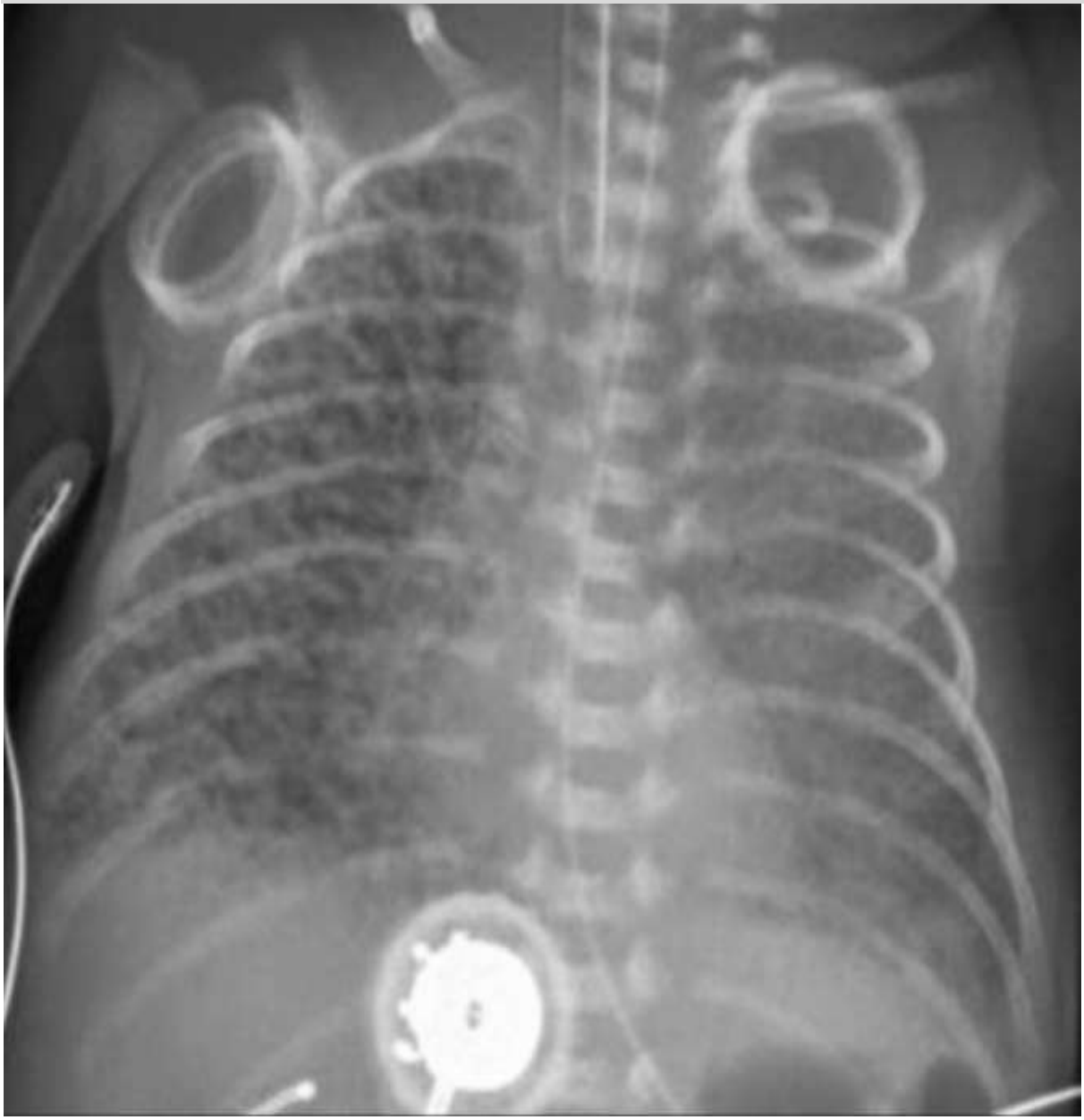
Special AOSPR 2016 Quiz

Faizah Mohd Zaki, Rohazly Ismail, Noryati Muhammad,
Hamzaini Abdul Hamid, Erica Hing Yee

CASE 1



Premature baby at 31 week of gestation who was intubated for respiratory distress syndrome (RDS). What is the abnormality?

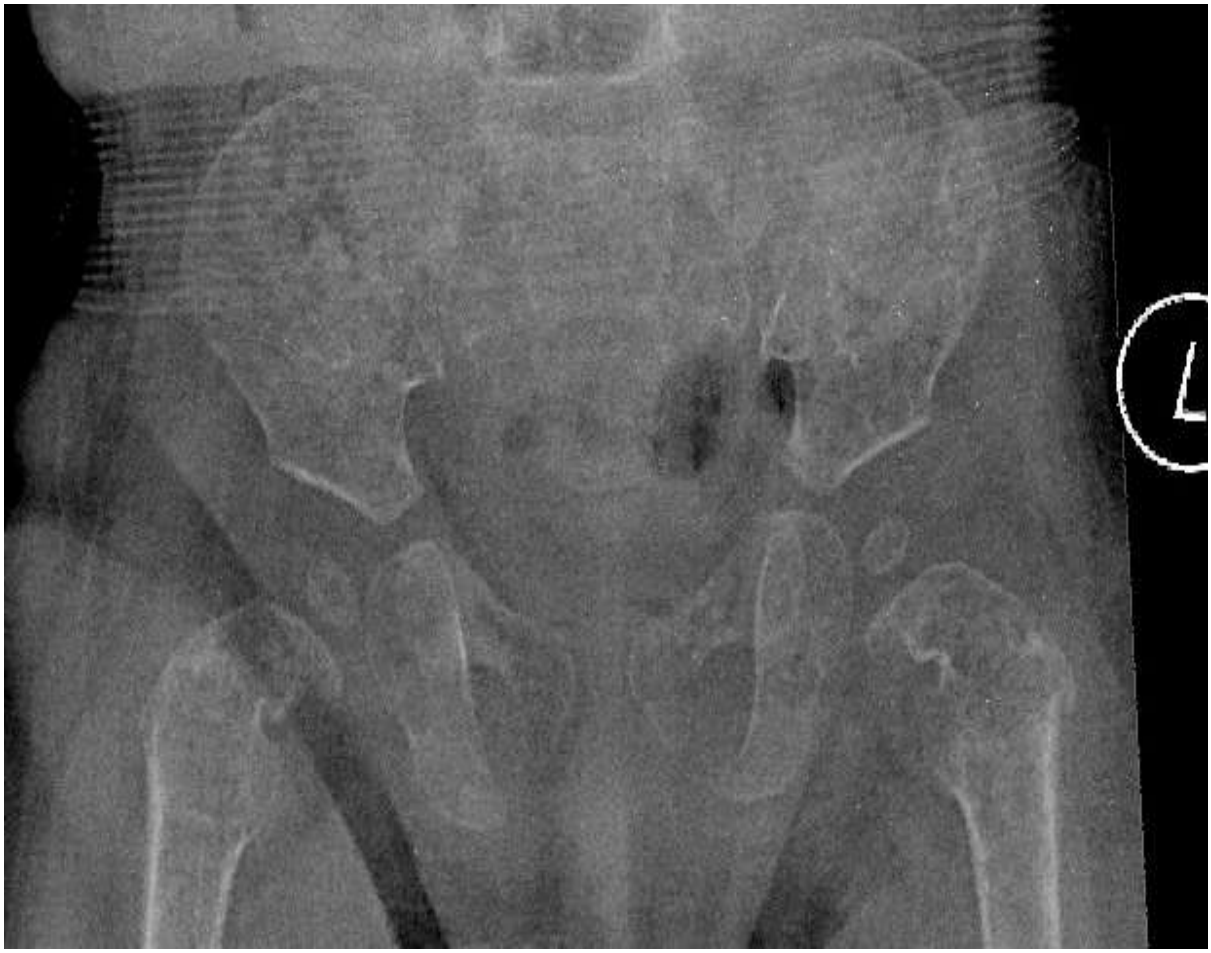
A. Pulmonary interstitial emphysema (PIE)

B. Spontaneous (idiopathic) pneumothorax

C. Congenital lobar emphysema (CLE)

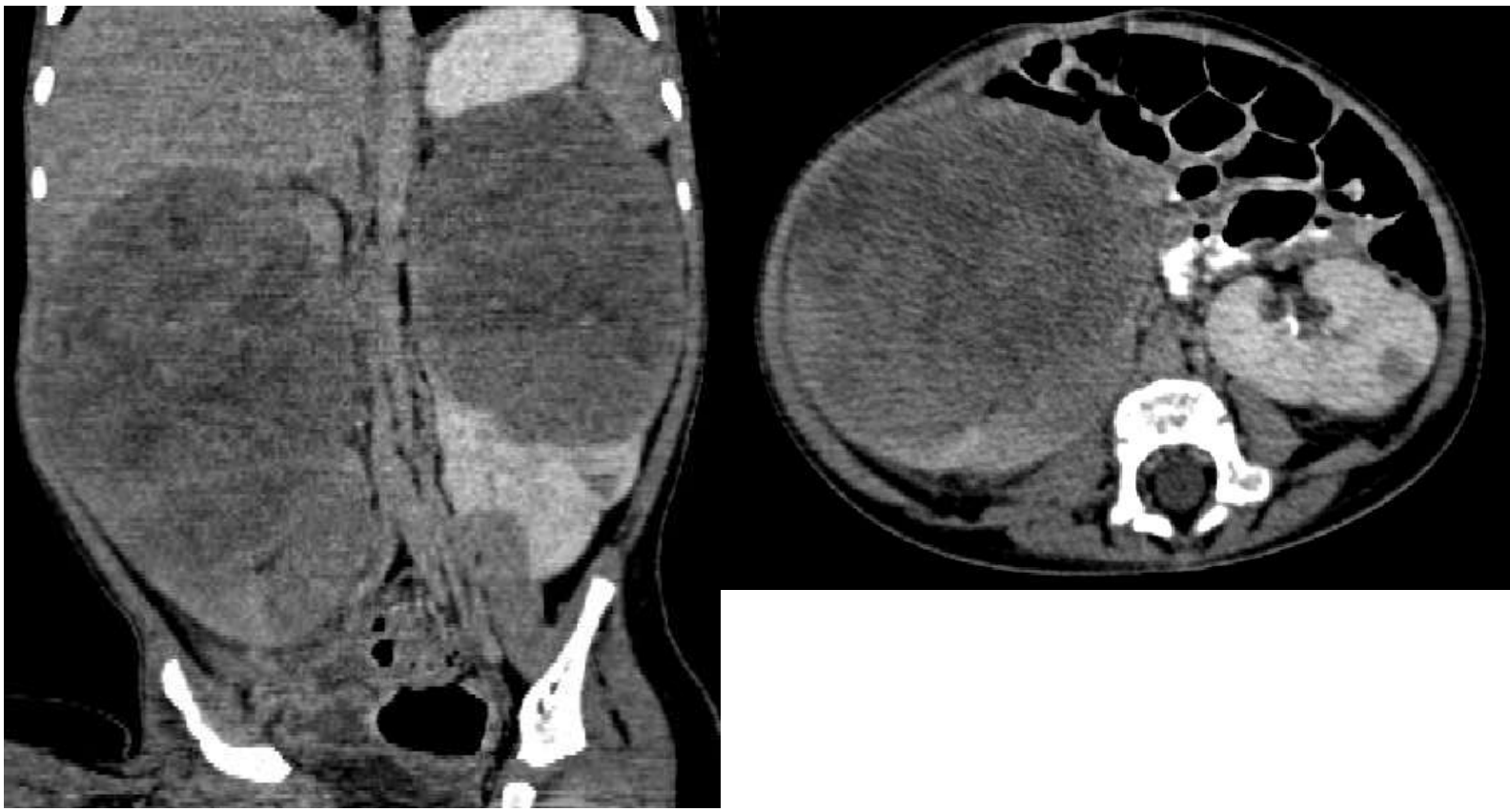
D. Congenital pulmonary airway malformation (CPAM)

CASE 2



- 10 month old boy, refused to weight bear. What is the most likely diagnosis?
- A. Metastatic Wilms tumor
- B. Leukemia
- C. Avascular necrosis right hip
- D. Osteosarcoma

CASE 3



8-year-old girl presented with progressive abdominal distension. Given the likely diagnosis, what can be the underlying disease in this patient?

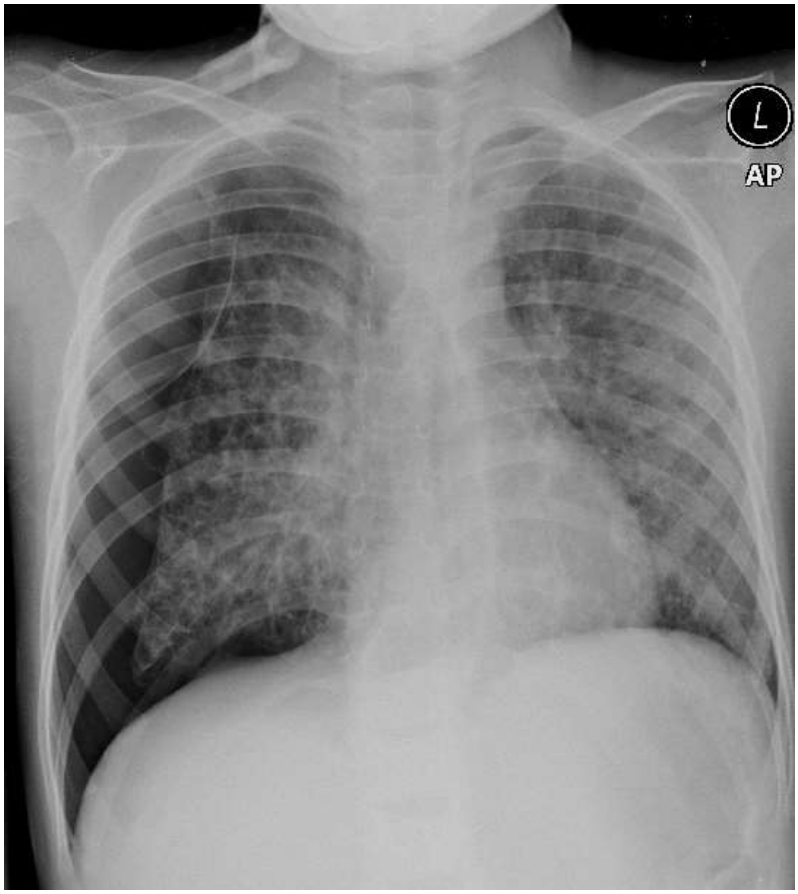
A. Denys-Drash syndrome

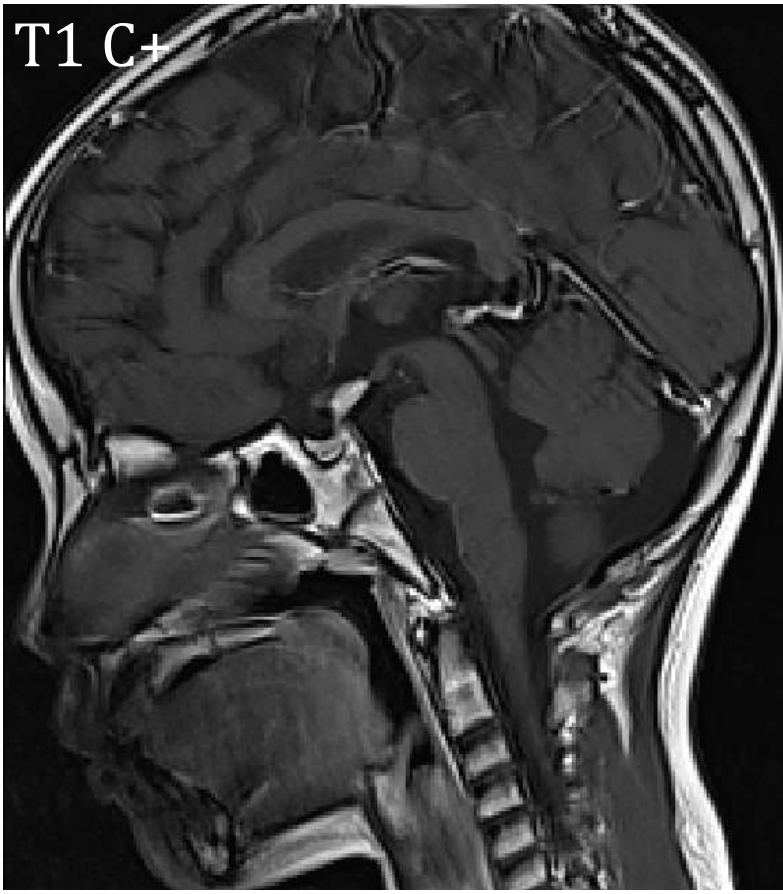
B. Beckwith-Wiedemann syndrome

C. Nephroblastomatosis

D. Trisomy 18

CASE 4





A 4-year-old boy presented with recurrent pneumothorax. What is the most likely diagnosis?

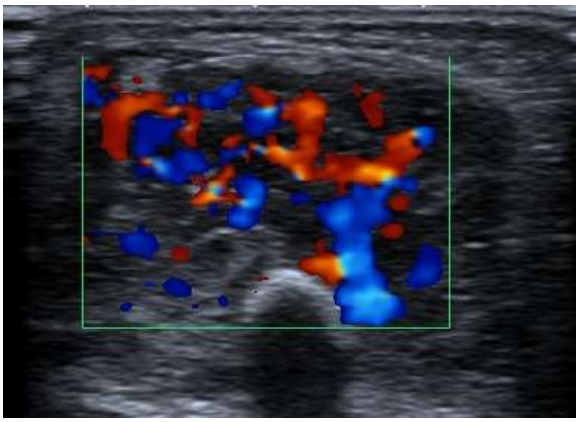
A. Sarcoidosis

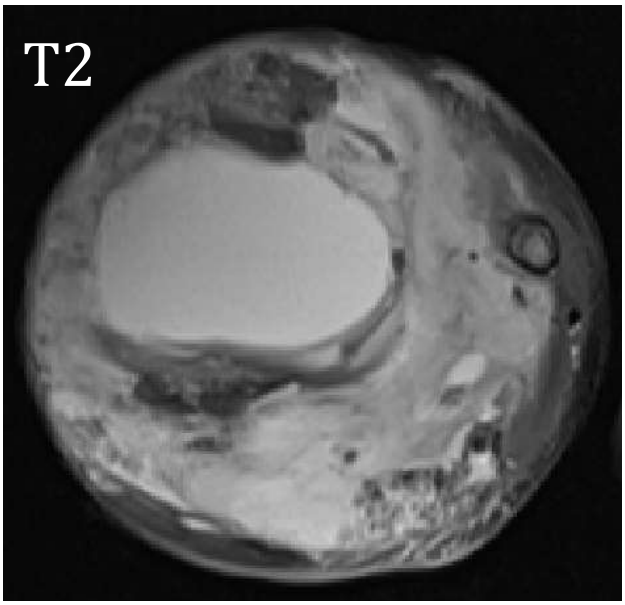
B. Langerhan cell histiocytosis

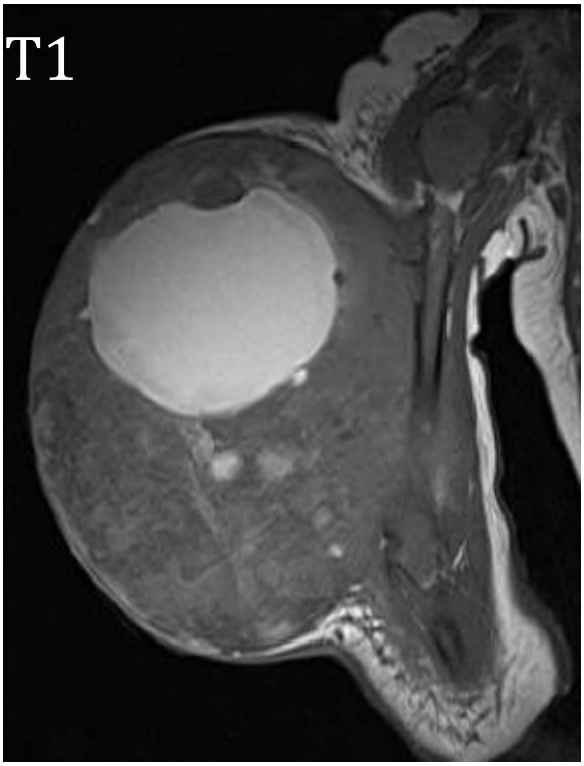
C. Lymphangioleiomyomatosis

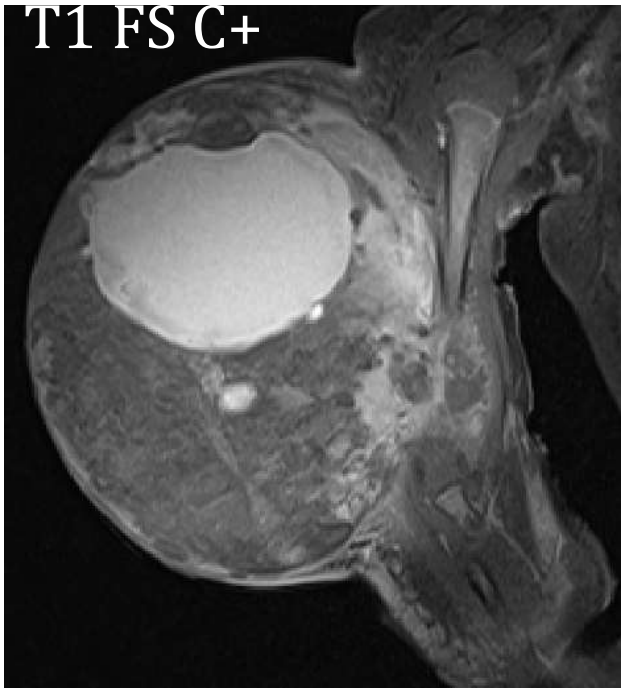
D. Birt-Hogg-Dube syndrome

CASE 5









Day 21 OL boy with progressive right arm swelling, noted since birth. The MRI was performed 3 weeks after the ultrasound. What is the most likely diagnosis?

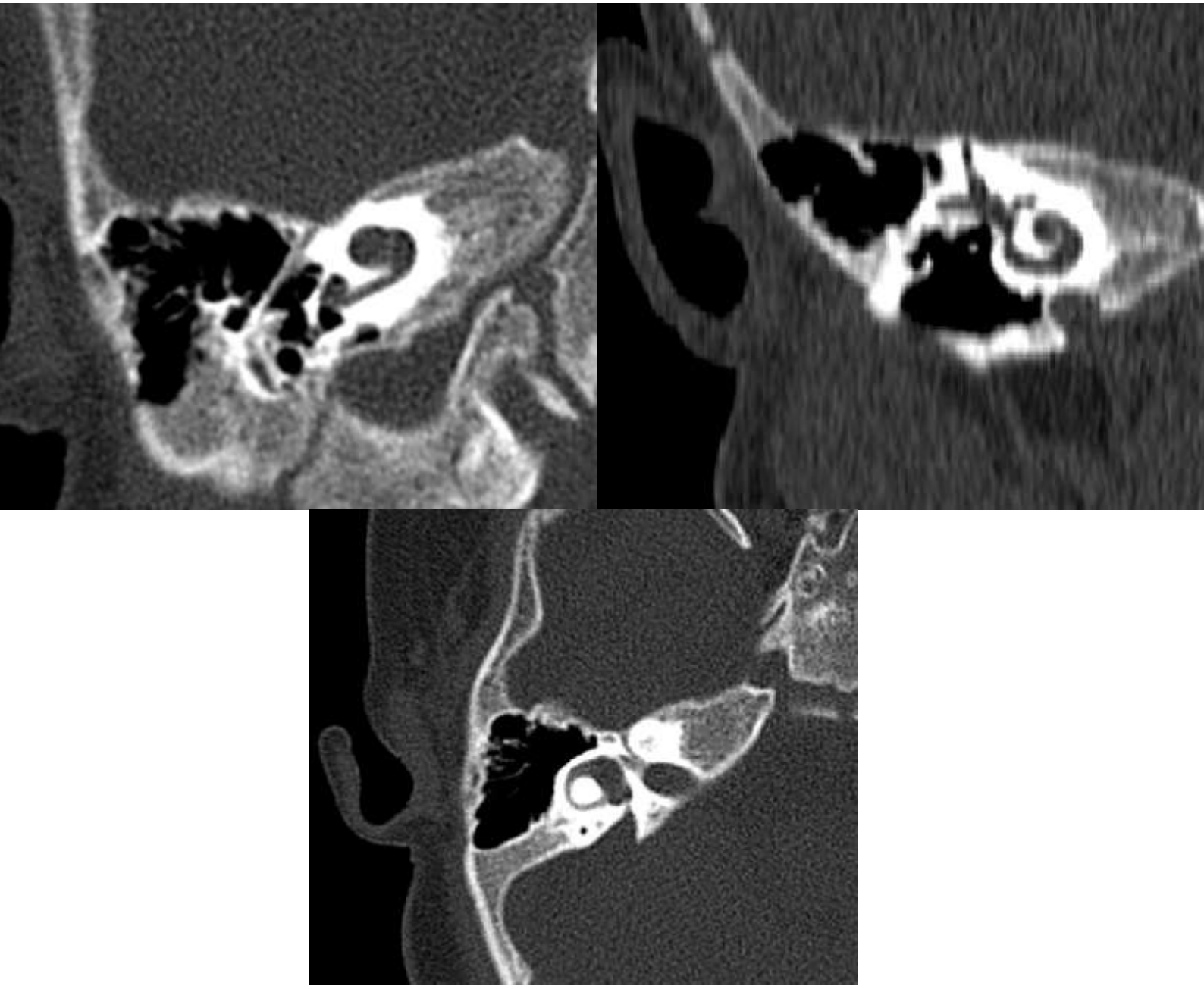
A. Congenital hemangioma

B. Dermatofibrosarcoma protuberans

C. Infantile fibrosarcoma

D. Intramuscular venous malformation

CASE 6



2-year-old boy with sensorineural hearing loss. What is the most likely diagnosis?

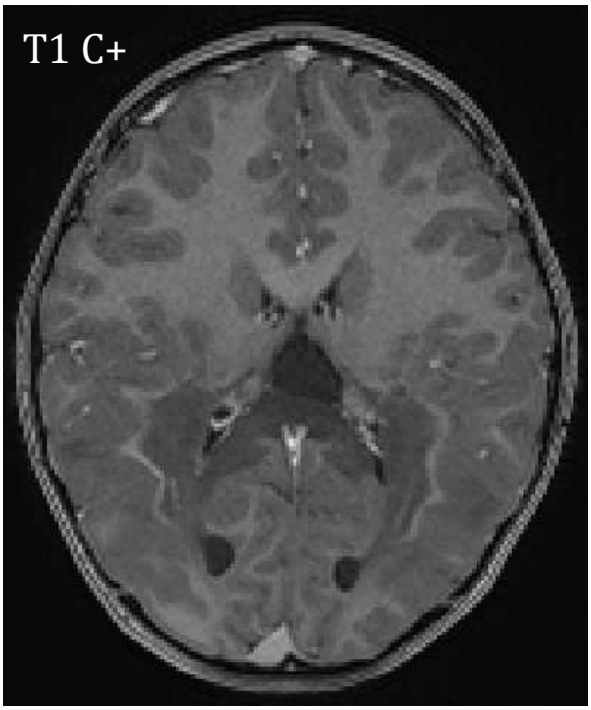
A. Incomplete partition type I with enlarged vestibular aqueduct

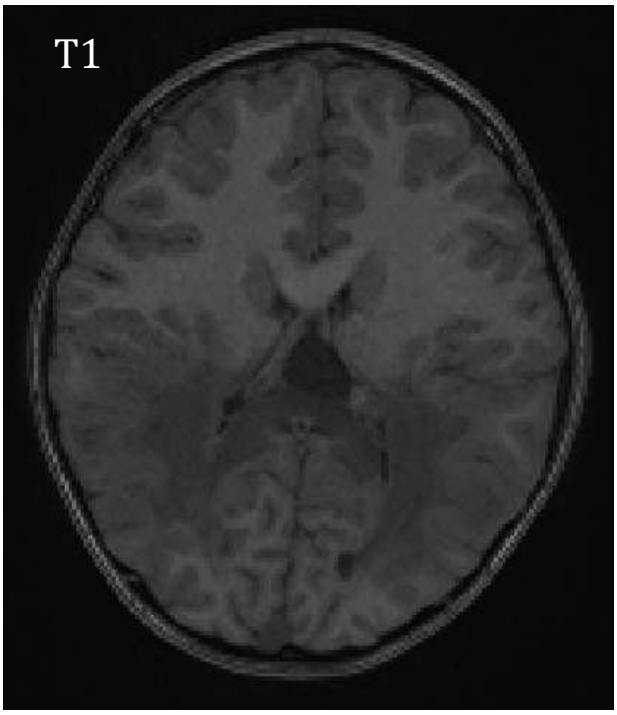
B. Incomplete partition type II with enlarged vestibular aqueduct

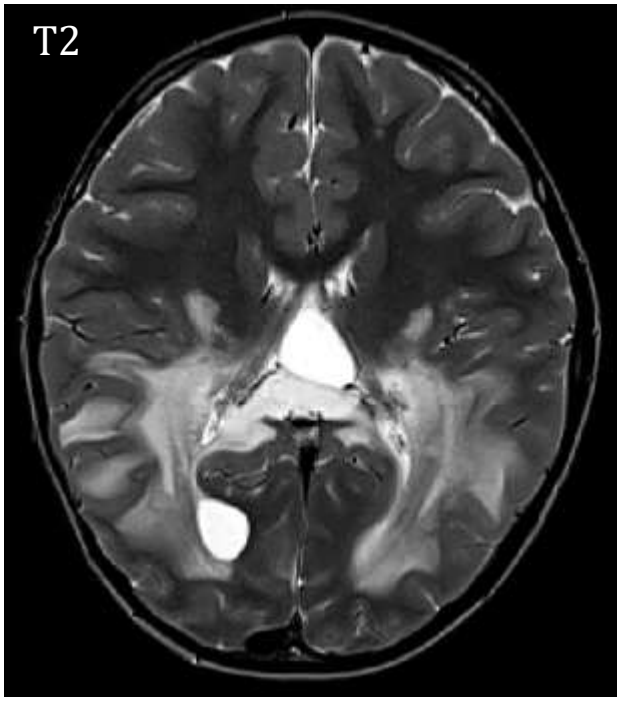
C. Cochlear aplasia with enlarged vestibular aqueduct

D. Common cavity malformation with enlarged vestibular aqueduct

CASE 7







6-year-old boy with progressive visual loss. What is the most likely diagnosis?

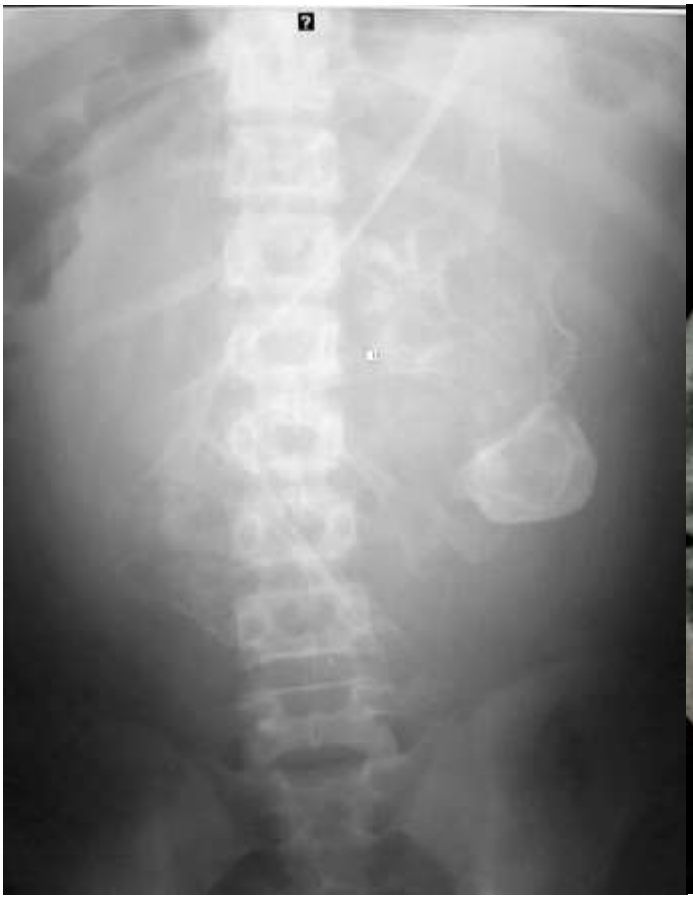
A. Metachromatic leukodystrophy

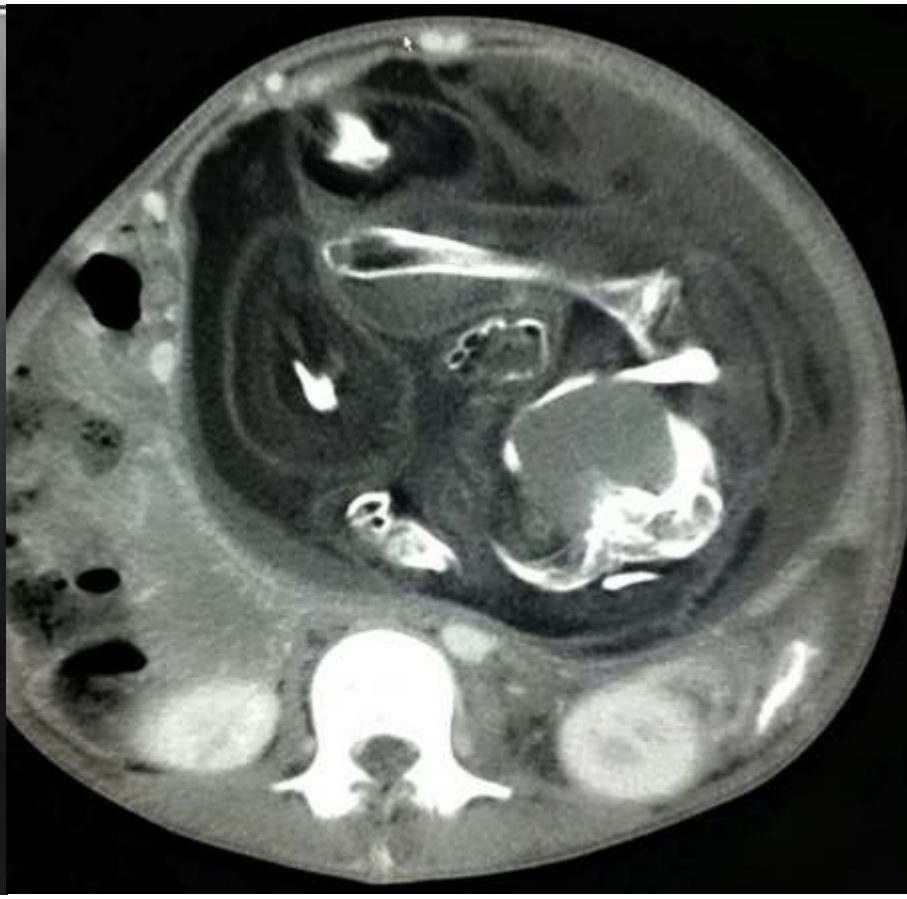
B. X-linked adrenoleukodystrophy

C. Pelizaeus-Merzbacher

D. Canavan disease

CASE 8





16 years old girl complained of gradually increasing abdominal mass. What is the most likely diagnosis?

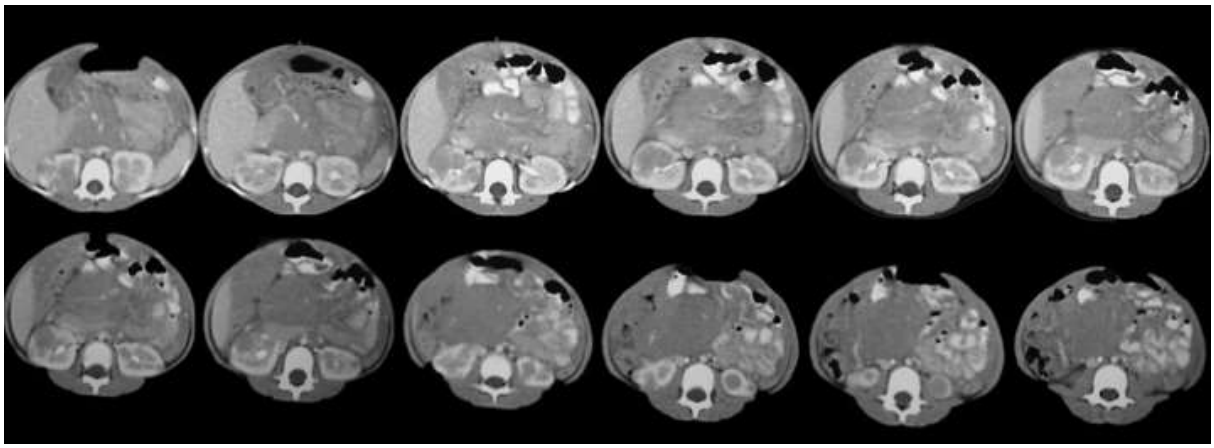
A. Intraperitoneal teratoma

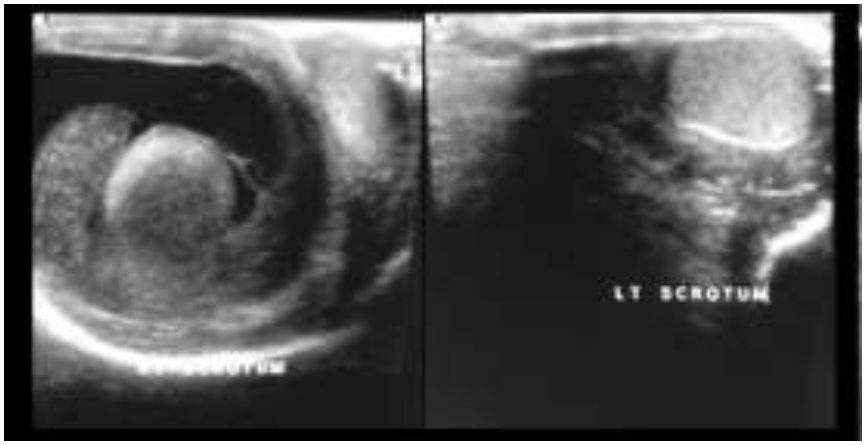
B. Peritoneal hamartoma

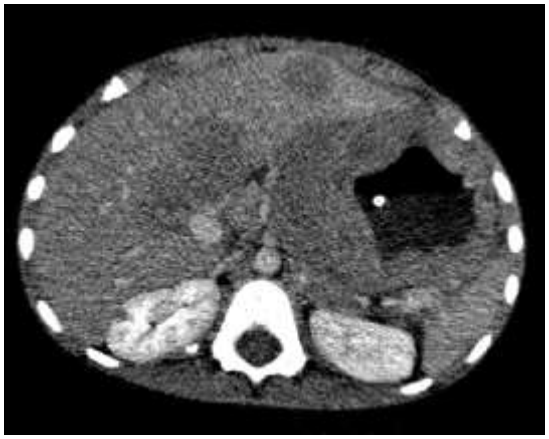
C. Lithopaedion

D. Fetus in fetu

CASE 9







5-year-old boy with history of progressive abdominal distension. What is the most likely diagnosis?

A. Metastatic neuroblastoma

B. Lymphoma

C. Metastatic testicular germ cell carcinoma

D. Crohn's disease

Special AOSPR 2016 Quiz Answer

Faizah Mohd Zaki, Rohazly Ismail, Noryati Muhammad,
Hamzaini Abdul Hamid, Erica Hing Yee

CASE 1

A. Pulmonary interstitial emphysema (PIE)

- Air leak phenomena
- Happens in pre-term babies with high ventilator setting
- Rupture bronchioloalveolar junctions, gas perivascular and peribronchial spaces
- Tubular and cystic lucencies – not branching pattern of air bronchograms
- Focal or diffuse, unilateral or bilateral

CASE 2

B. Leukemia

- Bone lesions are common in ALL (40- 60%)
- However, diffuse osteopenic changes with multiple pathological fractures are rare.
- Skeletal radiographic changes that can occur in a child with acute leukemia include
    - Diffuse osteopenia
    - Metaphyseal bands
    - Periosteal new bone formation
    - Geographic osteolysis, osteosclerosis, mixed osteolysis and sclerosis
    - Permeative destruction

CASE 3

C. Nephroblastomatosis

- "Nephrogenic rest" as a focus of abnormal nephrogenic cells persisting beyond 36 weeks' gestation
- Beckwith reported a 41% association between Wilms tumor and nephrogenic rests
- Beckwith-Wiedemann and hemihypertrophy
- On CT, the abnormal tissue enhances slightly and homogeneously (in comparison to the intense enhancement of the normal renal parenchyma).
- Diffuse nephroblastomatosis can be impossible to distinguish from autosomal recessive polycystic kidney disease, leukemia, or lymphoma.

CASE 4

B. Langerhan cell histiocytosis (LCH)

- LCH is the most common dendritic cell disorder
- Divided into three groups on the basis of the number of lesions and systems involved
- Lung involvement occurs in approximately 10% of LCH cases.
    - Centrilobular micronodules
    - Predominantly bilateral symmetric upper- to mid-lung distribution
    - The costophrenic angles are usually spared
    - Cysts vary in size but usually are less than 1 cm
    - Spontaneous pneumothorax
- CNS is involved in approximately 16% of LCH cases. Higher if facial bone and BOS involved
- MRI brain
    - Loss of the normal posterior pituitary bright spot
    - 70% of patients will also show thickening of the pituitary stalk

CASE 5

C. Infantile fibrosarcoma

- Malignant proliferation of fibroblasts
- First 5 years of life. 1/3 present at birth
- Extremities in 74%
- Ultrasound – Invariable, sometime hypervascular
- MRI – non-specific
    - T1 isointense
    - T2 heterogenously hyperintense
    - C+ - heterogenous enhancement
- Should be included if soft tissue tumor
    - Presenting in infancy
    - Located in an extremity
    - Show tumoural hemorrhage

CASE 6

B. Incomplete partition type II with enlarged vestibular aqueduct

- The most common type of cochlea anomalies found in children with sensorineural hearing loss
- Cochlea is only 1 1/2 turns with coalescence of the apical and middle turn, forming a cystic apex
- Basal turn of cochlea is normal
- When associated enlarged vestibular aqueduct (VA), it was formerly known as Mondini anomaly
- Always evaluate enlarged VA because it can cause intra-operative CSF gush during CI due to the communication with subarachnoid space of posterior fossa

CASE 7

B. X-linked adrenoleukodystrophy (X-ALD)

- Clinical: exclusive male, adrenal insufficiency, skin problem
- Diagnosis: VLFA to LCFA blood assay
- Demyelination spread outward and cephalad
- Contrast enhancement++
- **3 different zones:**
    - **Central/inner* - burned-out (scar)
    - **Intermediate* - active inflammatory demyelination (enhance post contrast)
    - **Peripheral* - leading edge active demyelination (not enhance post contrast)
- MRS in periphery: reduced NAA and high Choline peak

CASE 8

D. Fetus in fetu

- Rare (1 in 500,000 births)
- 1st theory: Malformed parasitic twin located in the body of its host twin
- Benign, normal alfa fetoprotein
- 80% in upper retroperitoneum
- Differential dx – Teratoma; common location at lower abdomen, ovaries or sacrococygeal
- Component: 91% spine, 83% limbs, 56% CNS, 45% GIT, 40% vessels, 27% GIT
- Willis criteria for FIF: **advance development of the mass**
  Vertebral axis + appropriate arrangement of limb and organ

CASE 9

B. Lymphoma

- NHL - heterogeneous
- large variety of diseases, nodal and extranodal
- Group (CHOP):
  1. Burkitt lymphoma (50%)
     - large abdomen mass, +/- marrow + CNS
  2. Lymphoblastic lymphoma (30%) - mostly T cell
     - neck, chest nodes, thymus +/- marrow, CNS
  3. Large cell (20%)

     - Anaplastic large cell lymphoma (usually T): neck nodes, nodes elsewhere, organ (lung, CNS, skin, bone, others)

     - Diffuse large B-cell lymphoma (B): large mediastinum mass, nodes elsewhere, bones

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