CALVARIAL TUBERCULOSIS

EFRAIM CULMINAS, LUCY KATHRINA BANTA- BANZALI

ABSTRACT

Skeletal tuberculosis accounts for 1-3% of TB cases, and of these only 0.2-1.3% had calvarial involvement.[1] Calvarial TB is most likely secondary to a primary focus. Diagnosis is confirmed through findings of *Mycobacterium tuberculosis* via microbiological, histopathological or cytopathological methods. This case report presents Primary Calvarial Tuberculosis in a five-year old male presenting with multiple cranial masses and initial diagnosis of Langerhans cell histiocytosis (LCH).

CASE REPORT

A five-year-old male presented with a two-year history of a left frontal head mass. He had a history of head trauma few days prior to the appearance of the mass. The mass was noted to gradually enlarge along with the appearance of multiple masses at the right and left parietal areas. Physical examination showed multiple, soft, fluctuant, non-erythematous, nontender, non-movable masses on the left frontal and both parietal areas of the skull. The rest of his physical and neurologic examination was unremarkable.

A skeletal survey revealed multiple varisized lucent lytic foci with non-sclerotic rims in the skull and a cranial Computed tomography (CT) scan showed multiple osteolytic changes scattered on the calvarium (Figure 1). Imaging findings were consistent with LCH. Two months into the work-up, the patient had cough and undocumented fever. Chest radiography revealed pneumonia with minimal left pleural effusion. Tuberculosis work-up was done and a Mantoux test showed an 11-millimeter induration while sputum Acid Fast Bacilli (AFB) showed negative findings. He was then started on Isoniazid and Rifampicin for three months. The patient has unremarkable past medical, surgical, family, birth, and maternal

history. He had completed his primary series of vaccination including the Bacille Calmette-Guerin (BCG) vaccine. He has no known allergies. His developmental milestones were at par with age.

He then underwent left frontal craniectomy and excision. Intraoperatively, the tumor was soft, yellowish, and thinly circumscribed with cystic fluid within. There was no infiltration to the inner table of the skull. Histopathology reported chronic granulomatous inflammation with multinucleated giant cells and extensive necrosis consistent with calvarial tuberculosis. The CD1a immunohistochemistry staining for LCH was negative. AFB testing of the cystic fluid was also negative. The Estimated Sedimentation Rate (ESR) was noted to be thrice elevated. Patient was subsequently managed as a case of calvarial tuberculosis and was given anti-tuberculosis treatment for 12 months. Three months post craniectomy, the patient already completed the intensive phase of the anti-tuberculosis regimen and is currently on the first month of the continuous phase. There was regression of the cranial masses and there was no evidence of appearance of new lesions.

CASE DISCUSSION

On the background of a normal neurologic examination, metastatic neuroblastoma and calvarial tuberculosis were considered in a patient who presented with bone lytic lesions. Initially, LCH was considered due to his clinical presentation and history that was supported by the imaging findings. Eighty percent of patients with LHC has skeletal system involvement of which 50% involves the skull. Radiologic studies will also reveal punched-out lytic lesions.[3] However this patient had a negative CD1a expression in immunohistochemistry and biopsy showed chronic granulomatous inflammation consistent with calvarial Tuberculosis. The rarity of calvarial tuberculosis put this diagnosis aside until the biopsy results were obtained. Usually this is transmitted via the

hematogenous or lymphatic routes. The spread through a lymphatic route makes this rare as the skull has a generous vascular supply. The primary event is the lodging of the bacilli in the diploic skull bones from an extracalvarial focus. Trauma is postulated to be a predisposing factor. This may be due to direct inoculation[4] or an increased vascularity, decreased resistance, discovery of a latent infection or the attraction of inflammatory cells to the area of trauma.[5,4]

Calvarial tuberculosis has a higher incidence in the young, aged 15-16 years old with predominance in males.[2,6] The most common presentation is a painless, soft fluctuant scalp swelling. This is usually followed by sinus discharge, localized pain, seizures and meningitis.[7] The most common sites of involvement are frontal and parietal bones and is due to the greater amount of cancellous bone with diploe channels at these sites.[2,3,5,6] Raut et al. found that the lesions may appear singly or multiple and has three types: The circumscribed or perforating type, the diffuse type and the least common, the circumscribed sclerotic type.

Our patient is a five-year-old male with a cranial mass who had the same presentation as with other cases of calvarial tuberculosis. Although vaccination was complete and no known exposure to tuberculosis infection were named, the history of head trauma is a strong predisposing factor. His cranial masses also involved frontal and parietal bones and is a perforating type of lesion. Imaging studies help to delineate lesions. A skeletal survey usually detects 80% of lesions and show punched-out defects with both osteolytic and osteoblastic areas. In plain cranial CT imaging, small, circumscribed and punched out, lytic or sclerotic lesions are commonly found in the parietal, frontal or occipital area of the skull. A magnetic resonance imaging (MRI) is highly specific and allow for a conclusive diagnosis as it can delineate subtle parenchymal involvement.

Our patient had lytic lesions in frontal, parietal and occipital area and cranial CT scan demonstrated bone destruction which is noted in 85% of calvarial tuberculosis.[6] Although the patient had no other symptoms such as night sweats, generalized weakness and loss of appetite, he developed fever and cough during his disease. His chest radiograph showed pleural effusion which hinted at a possible primary pulmonary tuberculosis or may represent a reactivation of tuberculosis, as pleural effusions may occur in the absence of a radiologically apparent tuberculosis.[8] A positive Mantoux test and an elevated ESR, as in the case of this patient may give a diagnostic clue to the diagnosis of tuberculosis. Calvarial tuberculosis can be confirmed through the isolation of Mycobacterium bacilli in culture or a positive AFB.[5] The patient's clinicoradiological presentation combined with histopathological evidence of a caseating granuloma is often sufficient to diagnose tuberculosis.[7] Once there is a strong clinical suspicion, the patient can be started on anti-tuberculous treatment and a good response will confirm the diagnosis. Our patient still satisfied the criteria for tuberculosis based on the National Tuberculosis Protocol of the Philippines (NTP). Given the history, physical examination, diagnostic studies, and the histologic confirmation through biopsy, the patient was clinically diagnosed with extrapulmonary tuberculosis.

The management comprises of completion of an anti-Tuberculosis regimen; in some cases, surgical intervention may be necessary. Surgery is indicated for large lesions and if neurological deficits are present.[5] In this case, surgery was vital in the patient's diagnosis as it led to the histologic finding of calvarial tuberculosis. As per NTP guidelines for skeletal tuberculosis, a two-month intensive phase, followed by ten months of the continuation phase is recommended. The patient's progress can be monitored through

serial ESR and cranial CT scans after the intensive phase of anti-Tuberculosis treatment. The prognosis of calvarial tuberculosis is generally good. Currently, the patient has no evidence of new-onset lesions or recurrence of lesions for seven months now since diagnosis. On follow-up patient is generally well, with no significant findings on MRI (Figure 2) and is on the first month of continuous phase anti-tuberculosis treatment.

SUMMARY

Calvarial tuberculosis is a rare form of skeletal tuberculosis, and it is important to diagnose early. This case shows that it is important to consider tuberculosis, which is a common disease in the Philippines but can often be missed due to its varying presentation. A thorough clinical history and physical examination are important as it can provide practical cues to arrive at the right diagnosis and management. Surgery is indicated for obtaining tissue for histological analysis or for removal of bony sequestra. Other surgical indications include cases with large extradural collections causing neurologic deficits or lack of response to treatment. Prognosis is dependent on the provision and compliance of appropriate pharmacotherapy. Hence patient education is vital as tuberculosis has implications to the community.

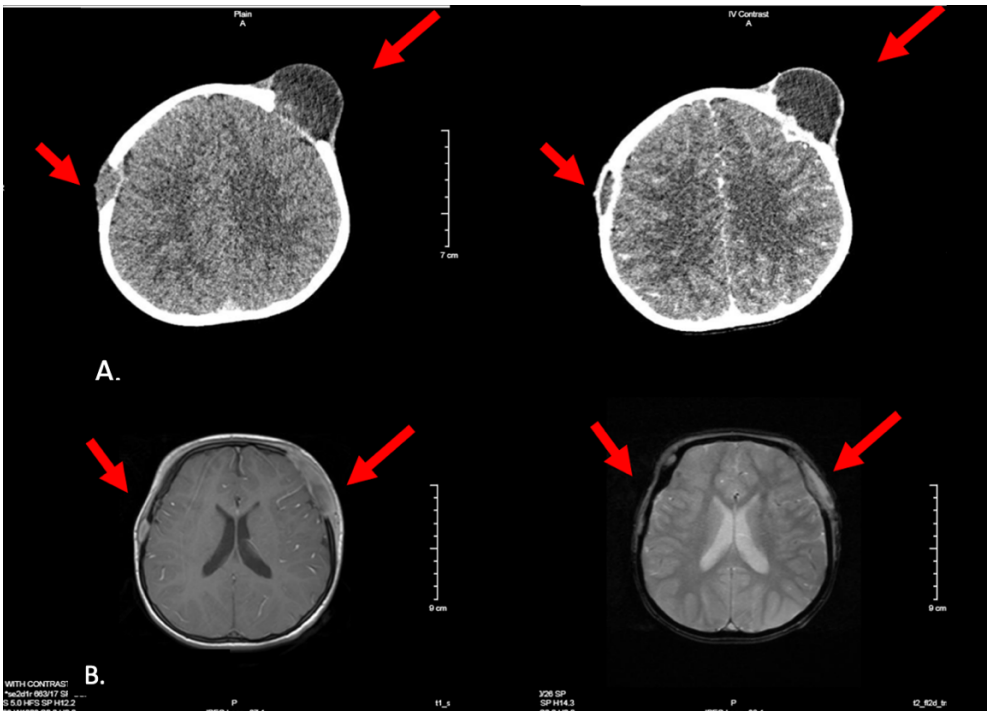

Figure 1. ***A.*** Cranial Computed Tomography (CT) scan preoperatively (without contrast, left. with contrast, right). Multiple osteolytic changes scattered on the calvarium associated with extra-cranial masses through the lytic defects, with most of them exhibiting epidural extensions. An avidly enhancing predominantly solid lesion (arrows) is also evident on the right frontal lobe ang left anterior temporal lobe with associated surrounding vasogenic edema and effacement of adjacent sulci. ***B.*** Cranial MRI post operatively (T1, left. T2, right). Destruction of both inner and outer tables of the skull with multiple soft tissue components extending into the subgaleal region with associated thickening of the dura. Multiple bulging scalp masses were noted at the left posterior frontal convexity, right anterior parietal bone, left posterior temporal to anterior occipital bone.

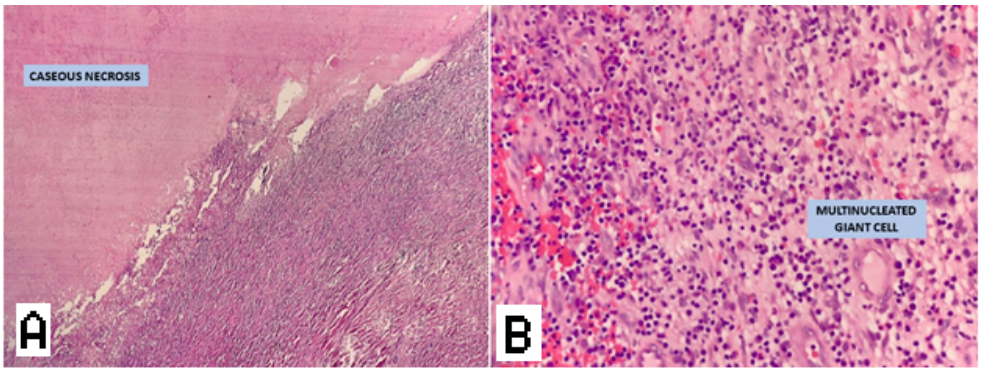

Figure 2. Hematoxylin and Eosin Stain of the calvarial mass (left, frontal) showing A. Caesous Necrosis and B. Granulomatous inflammation with multinucleate.

REFERENCES

1. Houda Ben A, Makram K, Chakib M, Khaoula R, Fatma H, Fatma S, et al. Extrapulmonary tuberculosis: Update on the epidemiology, risk factors and prevention strategies. International Journal of Tropical Diseases. 2018;1(1).
2. Saha DK, Ahsan ASMA, Fatema K, Faruq MO, Saha M, Ahmed F. Calvarial tuberculosis - a case report. Bangladesh Critical Care Journal. 2016;4(1):41–3.
3. Jezierska M, Stefanowicz J, Romanowicz G, Kosiak W, Lange M. Langerhans cell HISTIOCYTOSIS in children – a disease with many Faces. recent advances in pathogenesis, diagnostic examinations and treatment. Advances in Dermatology and Allergology. 2018;35(1):6–17.
4. Chand K, Singh A, Awasthy N. Calvarial tuberculosis: Review of six cases. Annals of Indian Academy of Neurology. 2006;9(4):227.
5. Diyora B, Kumar R, Modgi R, Sharma A. Calvarial tuberculosis: A report of eleven patients. Neurology India. 2009;57(5):607.
6. Abhijit A. Raut, Arpit M. Nagar, Datta Muzumdar, Ashish J. Chawla, Ranjeet S. Narlawar, Sudhir Fattepurkar, Veena L. Bhatgadde. American Journal of Neuroradiology Mar 2004, 25 (3) 409-414;
7. An unusual presentation of calvarial tuberculosis. Postgraduate Medical Journal. 2002;78(917):188–9.
8. Santra A, Sen A, Chatterjee A. Calvarial tuberculosis in a Preschool-Aged Child: An UNCOMMON ENTITY. Sultan Qaboos University Medical Journal [SQUMJ]. 2018;17(4):484.
9. Cohen LA, Light RW. Tuberculous pleural effusion. Turkish Thoracic Journal/Türk Toraks Dergisi. 2015;16(1)