

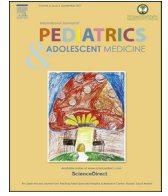
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Case report

The many faces of tuberculosis

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ABSTRACT

Tuberculosis (TB) presenting as multifocal cystic lesions of bone (Jungling's disease) with an intra-orbital mass and calvarial involvement is rare, especially in an immunocompetent host. A 3-year-old boy presented with acute-onset, painless, intra-orbital swelling of the right eye and bony lesions over the right tibia and frontal bone. Tubercular etiology could be established with the help of the Xpert MTB/RIF test, which was performed on pus aspirated from the orbital lesion. The child improved upon antitubercular treatment. Atypical presentations of TB in children are highly variable and might include multifocal osteolytic lesions and intraorbital abscesses. Use of appropriate diagnostic methods, along with timely intervention, leads to a favorable outcome.

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1. Introduction

Approximately 25% of pediatric tuberculosis cases are extrapulmonary, with only a small proportion presenting with bone and joint involvement [1]. Multifocal TB osteomyelitis is a rare disease, comprising 4.6–10% of osteo-articular TB cases [2]. Cystic bone TB, also termed Jungling's disease or osteitis tuberculosa multiplex cystoids, is a rarer variety of TB osteomyelitis [3]. The lesions predominantly involve the metaphysis but can occur anywhere in the skeleton and may mimic infections (bacterial or fungal) and malignancies (primary or metastatic). Tubercular orbital and calvarial involvements are exceedingly rare and create a diagnostic dilemma. Biopsy is the gold standard for the diagnosis of osteo-articular TB; however, with the advent of nucleic acid amplification assays, the diagnosis can be established in less time and in a less invasive manner.

We treated a child with Jungling's disease with calvarial involvement and an intra-orbital, extra-conal tubercular abscess that was diagnosed by CBNAAT (cartridge-based nucleic acid amplification test) and treated successfully with Anti-tubercular

therapy (ATT). We report this case in light of its value as a rare and complex presentation of TB in a pediatric patient; we also aim to highlight the possible role of newer diagnostic modalities in achieving good clinical outcomes coupled with the use of standard ATT.

2. Case report

A 3-year-old, appropriately immunized, premorbidly asymptomatic boy presented with a painless swelling at the outer corner of his right eye for 3 months and two painful bony swellings: one over the right shin for 2 months and the other over his forehead for 15 days. There was no history of trauma, visual disturbances, fever, weight loss, cough, bleeding, blood component therapy or contact with TB. Examination revealed a soft, erythematous, fluctuant swelling approximately 3 × 3 cm in size at the outer canthus of the right eye. There was a slightly tender, pea-sized, firm swelling over the forehead with no evidence of inflammation (Fig. 1A). There was also an ill-defined, firm, tender, bony swelling over the shaft of the right tibia (Fig. 2A). A Bacillus Calmette–Guérin (BCG) scar was present. There was no lymphadenopathy, organomegaly or other mass lesions. A complete hemogram and liver and renal function tests were within normal limits. A Mantoux test was positive (14 mm). A CT scan of the head without contrast revealed a small lytic lesion in the frontal bone and a hypodense, focal, lytic lesion with an associated periosteal reaction involving the right zygoma that extended posteriorly into the right infra-temporal fossa with

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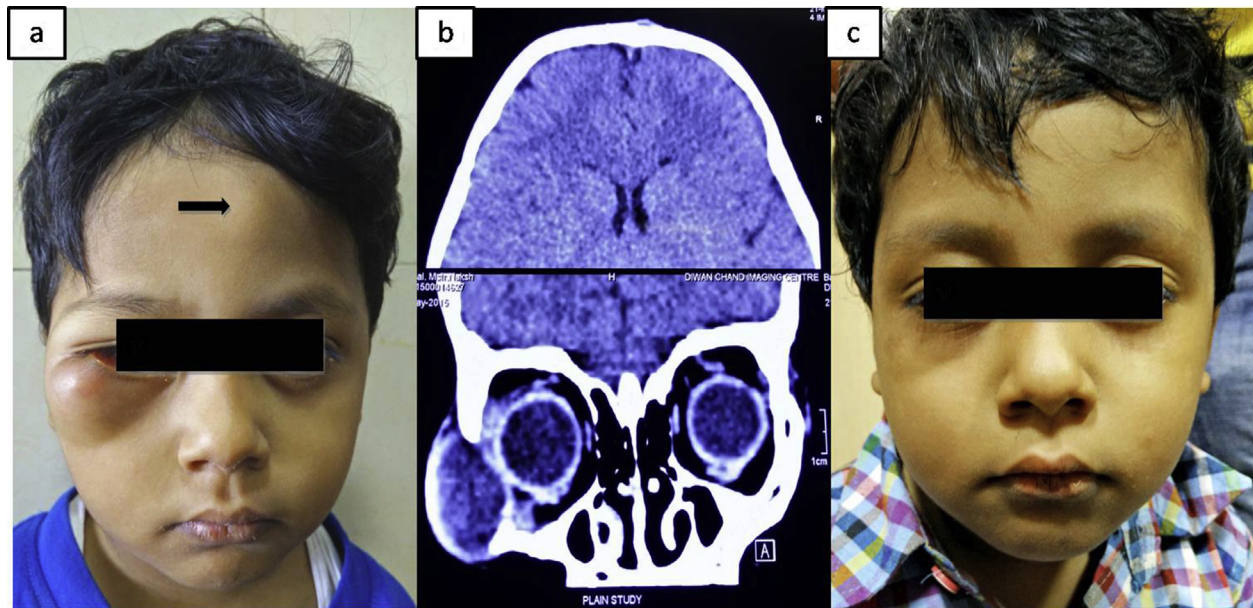


Fig. 1. A. Soft, fluctuant swelling at the outer canthus of the right eye with slight redness and a pea-sized, firm swelling over the forehead (black arrow); Image B. Plain CT scan of the head and orbits showing a hypodense, focal, lytic lesion with an associated periosteal reaction involving the right zygoma with contiguous extra-conal soft tissue thickening and a small lytic lesion in the frontal bone with associated mild soft tissue thickening; C. The swelling in both the right eye and forehead completely subsided after 6 months of antitubercular therapy.

extra-conal soft tissue thickening in the infero-lateral aspect of the right orbit (Fig. 1B). Ultrasound showed an intra-orbital, extra-conal lesion with few echoes within it. X-ray images of the right tibia showed an ill-defined, lytic bony defect in the medulla of the mid-diaphysis with a thick periosteal reaction and thickening of the surrounding soft tissue (Fig. 2B) USG-guided aspiration of the eye lesion yielded 7 ml of purulent material. Gram staining showed only pus cells; bacterial and fungal cultures were sterile. Cytopathology was suggestive of acute inflammatory exudates and necrosis, the stain for acid-fast bacilli was negative, and there were no malignant cells. GeneXpert testing of the pus was positive for

Mycobacterium tuberculosis, and it was sensitive to Rifampicin. His chest X-ray was within normal limits, and a contact survey was negative for exposure to TB. He was started on a category 1 treatment for TB, pending the mycobacterial culture. The work-up for Human immunodeficiency virus (HIV) was negative. The Mycobacterium growth indicator tube (MGIT) culture grew *Mycobacterium tuberculosis*, which was sensitive to all the first-line antituberculous drugs. After 6 months of follow up, the child remained asymptomatic with complete subsidence of the swellings in the eye and bones (Fig. 1C).

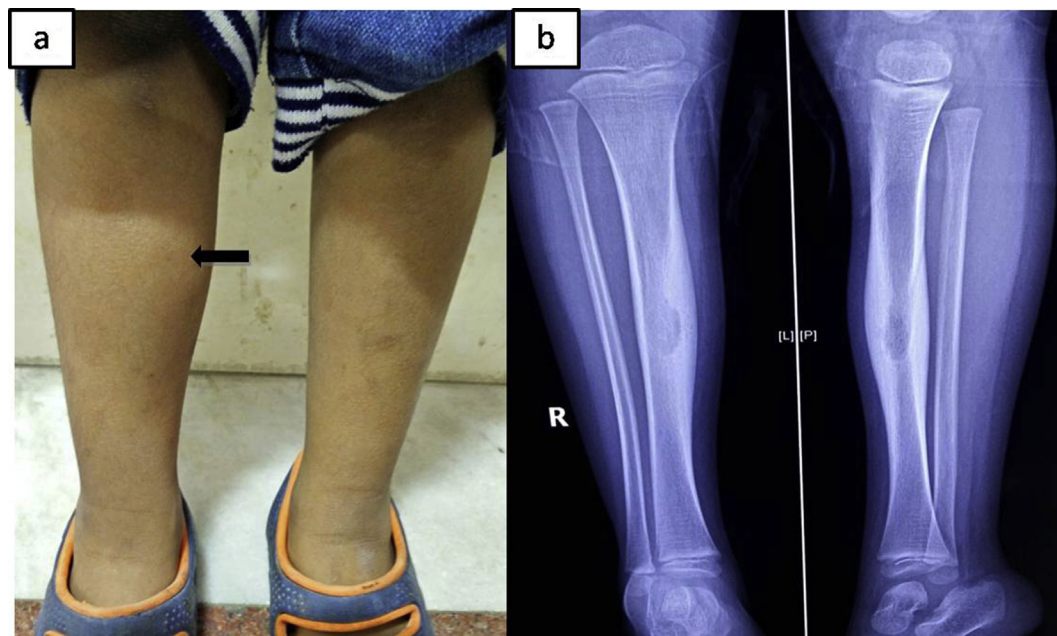


Fig. 2. A. An ill-defined, firm, tender swelling over the right tibia (black arrow) without any evidence of inflammation; B. X-ray images (lateral and antero-posterior) of the right tibia showing an ill-defined lytic bony defect in the medulla of the mid-diaphysis with a surrounding thick periosteal reaction.

3. Discussion

The child in our case presented with an intra-orbital, extra-conal cystic swelling and multifocal, bony lytic lesions. The differential diagnosis for multiple bony lytic lesions in children includes fibrous dysplasia, Ollier's disease, Langerhans' cell histiocytosis and metastases (e.g., neuroblastoma, primitive neuroectodermal tumor, leukemia, primary bone tumors). The differential diagnosis for acquired cystic lesions in the orbit includes lymphangioma, lacrimal gland cysts, hematic cysts, hydatid cysts, cysticercus cysts and a non-enhancing area of mucocele [4,5]. Inflammatory condition such as orbital pseudotumor and malignant diseases such as Langerhans' cell histiocytosis/juvenile xanthogranuloma and granulocytic sarcoma can also present similarly [5].

The most common presentation of osteo-articular TB in children consists of spondylitis, arthritis of large joints and osteomyelitis, with spinal involvement being the most common presentation (50%) [6]. A multifocal form of skeletal TB is very uncommon and has been limited to case reports and series only [2]. In these cases, the spine is the site most commonly affected, followed by the long bones; however, spinal involvement was absent in our case. Reported cases of orbital tuberculosis (OTB) number less than 100 in the literature [7]. Orbital involvement with a cystic, intra-orbital, extra-conal lesion containing purulent material similar to our case has been reported by Babu K et al. [7] Our patient had the 'OTB with evidence of bony destruction' variety, the second most commonly observed variety of orbital TB. TB with calvarial involvement is very rare, representing just 0.2–1.3% of all cases of skeletal tuberculosis [8]. These cases present with painless swelling over the scalp with a discharging sinus that is often preceded by a history of trauma [9]. Our patient had a painful swelling with no discharge and without a history of trauma. Such a combination of rare presentations in a pediatric patient with TB has not been previously described in the literature.

The gold standard for the diagnosis of skeletal TB is histopathology analysis and demonstration of Acid-Fast Bacilli (AFB) either by Zeihl-Neelsen staining or culture; however, nucleic acid amplification techniques are presently being used with increasing frequency to increase the yield [8]. In our case, the right orbital cystic swelling aspirate was negative for AFB, but the diagnosis of tuberculosis was confirmed using the Xpert MTB/RIF technique (GeneXpert); it also showed Rifampicin sensitivity. The Xpert MTB/RIF test is a cartridge-based nucleic acid amplification technique that is currently recommended by the WHO as a replacement for conventional testing practices (e.g., microscopy, culture or histopathology analysis) of specific non-respiratory specimens in the diagnosis of extra-pulmonary TB [9]. Approximately 50–70% of osteo-articular TB patients are found to have concomitant evidence of pulmonary involvement. Our patient did not have any previous history of tuberculosis, and there were no pulmonary lesions suggestive of old or active TB.

Mendelian susceptibility to mycobacterial diseases is an important predisposing condition for atypical (and often disseminated) TB in otherwise healthy hosts [10]. A mutation analysis could not be conducted in our case. The patient was started on the

standard 4-drug ATT, as per category 1 of World Health Organisation (WHO) guidelines, and showed good response with decreased orbital swelling and decreased pain over the right tibia. He is scheduled for 12 months of treatment as per recommendations [9]. A few atypical features were recognized in our case, namely, a relatively short course, absence of constitutional symptoms, and involvement of scalp and orbit in an immunocompetent host.

4. Conclusion

Tuberculosis should be suspected in cases in which an apparently normal host presents with osteomyelitis at unusual or multiple sites.

Author contribution

Both the authors were involved in the management of the patient, literature search and manuscript writing. All the authors approve the final manuscript.

Patient consent

Written consent was obtained from the father of the patient for publication of the case including images.

Conflict of interest

None stated.

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