International Journal of Science and Research (IJSR)
ISSN: 2319-7064
SJIF (2019): 7.583

Vertebra Plana in Paediatric Patient with Paraplegia; Diagnostic Approach and Management

Soman S

Assistant Professor, Department of Orthopaedics, Smt Kashibai Navale Medical College, Pune, Maharashtra, India

Abstract: ***Introduction: Vertebra plana is a typical feature of eosinophilic granuloma. But in endemic countries tuberculosis is always a probability especially when there is neurological involvement. Other causes are also to be considered for the appropriate management. Histology sometimes comes inconclusive for which immunohistochemistry helps to reach the diagnosis. Case presentation: We present a case of 12 years old girl with vertebra plana and neurological compromise being treated as tuberculosis outside. We did posterior instrumented fusion with decompression and sent a biopsy. But it failed to give a diagnosis. Later immunohistochemistry proved it to be eosinophilic granuloma. The patient recovered completely to normal neurology. Conclusion: This case demonstrates a rare case of eosinophilic granuloma presenting as vertebra plana and causing neurological deficit. Hence patients should be closely monitored and should undergo decompression at the earliest evidence of neurological compromise. Also it highlights the role of immunohistochemistry in diagnosis.Key-words: vertebra plana, eosinophilic granuloma, tuberculosis, decompression***

1. Introduction

Vertebra plana is the term given to a vertebral body which has lost almost its entire height anteriorly and posteriorly. It is also called pancake vertebra[1]. It can be caused by a variety of conditions like trauma, osteoporosis, Langerhans cell histiocytosis, osteogenesis imperfecta, leukaemia, vertebral metastasis, multiple myeloma, lymphoma, Ewing's sarcoma, osteomyelitis, vertebral hemangioma[2,3,4,5,6] etc. Its most common cause in children is Langerhans cell histiocytosis in which the patient presents with local pain and insidious symptoms. Neurological deficit is very rare; in fact there is no literature of it with gross neurological involvement. However in endemic countries, tuberculosis can also present with similar findings. Biopsy thus becomes important for the diagnosis. But it often comes inconclusive for which immunohistochemistry will help to reach the correct diagnosis. The aim of presenting this case report was to show a patient with vertebra plana presenting with neurological deficit where biopsy was inconclusive. However the diagnosis was confirmed with immunohistochemistry suggesting its use in such cases.

2. Case Presentation

A 12 years old girl presented with history of back pain for 14 days and inability to move both of her lower limbs for 10 days. Sensations were intact with no bowel and bladder involvement. There was no history of fever,weight loss or other constitutional symptoms.Upper extremities were normal. She had consulted a local hospital before and was being treated as spine tuberculosis based on radiographs. She had tenderness at lower thoracic spinal processes without paraspinal fullness. Neurological examination showed increased tone with ASIA Scale B with neurological level D8. No clinically significant past history was present. She was admitted and investigated. Routine blood investigations were normal. A dorsolumbar spine X-ray (Figure 1) showed vertebra plana of T8 vertebra with sclerosis and maintained disc spaces without paraspinal shadow. MRI showed vertebra plana of T8 vertebra with complete marrow replacement of vertebral body and posterior elements with associated homogenously enhanced soft tissue component in adjacent pre- and para-vertebral space and in ventral and dorsal epidural spaces leading to severe cord compression. The differential diagnosis of this entity was assessed (Table 1).The patient underwent posterolateral decompression with instrumented fusion to decompress the neural structures and for biopsy for diagnostic purpose. Intraoperatively circumferential engulfment of cord with granulation tissues was encountered which was removed and sent for histopathological examination. But it came inconclusive following which immunohistochemistry was done which proved the diagnosis of eosinophilic granuloma on the basis of S100 positive. The patient improved remarkably after surgery to ASIA Scale E at 1 months post operative. At final follow up of 6 months,patient showed normal neurology with early signs of spontaneous reconstitution of vertebral body.

3. Discussion

In endemic countries, tuberculosis presents with a wide range of clinical and radiological features. Osseous tuberculosis often resembles other destructive bony lesions[7]. It often is a probable diagnosis even if the clinic-radiological features are atypical. So confirmation of the diagnosis is essential for the proper management of the patient (Table 1).

Langerhans cell histiocytosis is a spectrum of diseases of the reticuloendothelial system. One of its form, eosinophilic granuloma, presents as a single self-limiting lesion in children. It constituted less than 1% of all bone tumours but 75% of all cases of Langerhans cell histiocytosis[8].it is primarily a disease of the bone but can also involve other regions[9]. Patients complain of local pain, back stiffness, decreased range of motion. Vertebra plana is a typical radiological feature. Neurological involvement is extremely rare[10] and can occur due to vertebral collapse or spread to peridural region causing compression of the spinal cord[11]. Since its clinical and radiological features are similar to other conditions as well, histopathological examination and immunohistochemistry are important for its diagnosis. Histopathological examination shows Langerhan's cells, mixed inflammatory cells and giant cells. There is no nuclear atypia and no atypical mitosis. Detection of SP 100

protein on Langerhan's cells by immunohistochemistry is diagnostic. Treatment options include observation with or without bracing, curettage, wide excision, local radiation and chemotherapy[12]. But conservative methods are favoured as the condition is self-limiting, can undergo spontaneous resolution[13] and no particular medical or surgical treatment is proved to be better than observation.Surgery is needed in rare cases for severe persistent pain, spinal instability or neurological compromise.

Our case also demonstrates the natural history of vertebra plana by causing neurological deficit. Hence patients should be closely monitored and should undergo decompression at the earliest evidence of neurological compromise.

4. Conclusion

Vertebra plana in paediatric population presents a diagnostic challenge to spine surgeons. These patients should be meticulously examined and differential diagnosis other than those endemic to that region should be considered. Eosinophilic granuloma can present with varied clinical features and should be managed appropriately after diagnosis where immunohistochemistry is an important tool in case with inconclusive biopsy report. Posterior instruments fusion with decompression is an appropriate modality even in patients with gross neurological compromise.

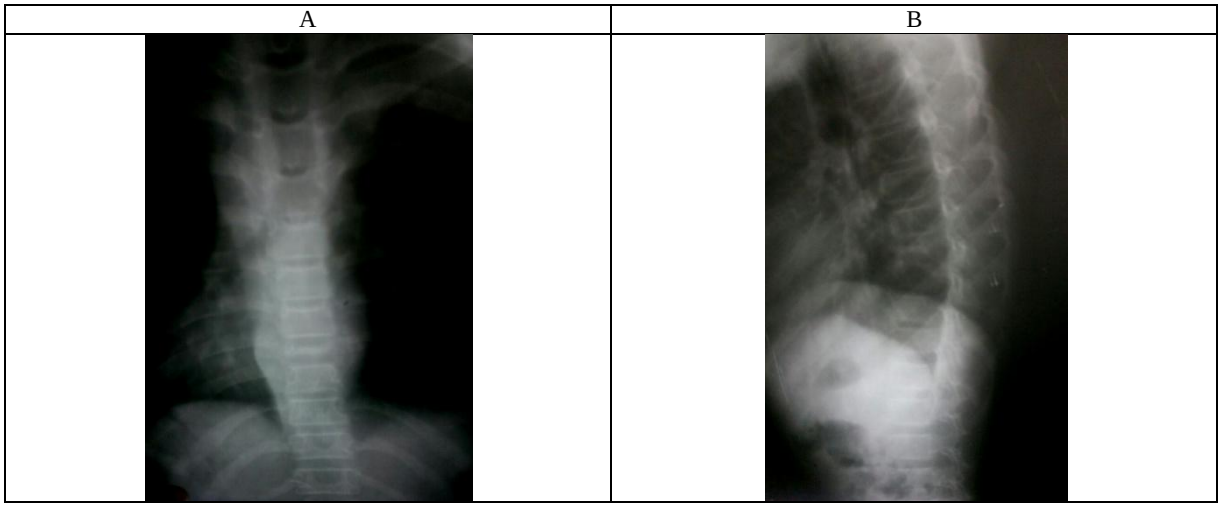
5. Clinical Message

Eosinophilic granuloma should be a differential diagnosis in paediatric age group with vertebra plana even in tuberculous endemic areas. Also immunohistochemistry has a key role in diagnosis of eosinophilic granuloma. Surgical decompression has a favourable outcome in patients presenting with neurological deficit.

References

[1] Roche CJ, O'Keeffe DP, Lee WK, Duddalwar VA, Torreggiani WC, Curtis JM. *Selections from the buffet of food signs in radiology.RadioGraphics* 2002; 22:1369-1384.
[2] Reeder MM. *Gastrointestinal tract and abdomen. Reeder MM, Bradley WG, eds.* Reeder and Felson's gamuts in radiology. 3rd ed. New York, NY: Springer-Verlag, 1993.
[3] Dahnert W. *Radiology review manual* 3rd ed. Baltimore, Md: Williams & Wilkins, 1996.
[4] Swischuk LE. *Differential diagnosis in pediatric radiology* Baltimore, Md: Williams & Wilkins, 1984.
[5] Wickenhauser J, Sunder-Plassman M, Zaunbauer F, Flemmich K, Hohenberg G. *Vertebra plana Calve. Neuroradiology* 1979; 18:221-225.
[6] Ippolito E, Farsetti P, Tudisco C. *Vertebra plana: long-term follow-up in five patients. J Bone Joint Surg Am* 1984; 66:1364-1368.
[7] Haghighatkhah H, Jafroodi Y, Sanei Taheri M, Pourghorban R, Sadeghian Dehkordy A. Multifocal Skeletal Tuberculosis Mimicking Langerhans Cell Histiocytosis in a Child: a Case Report With a Long-Term Follow-Up. *Iranian Red Crescent Medical Journal*. 2015;17(12):e19942. doi:10.5812/ircmj.19942.
[8] Sweasey TA, Dauser RC. Eosinophilic granuloma of the cervicothoracic junction. J Neurosurg 1989; 71: 942 ± 944.
[9] 5 Huvos AG. Bone tumors. Diagnosis, treatment and prognosis. 2nd edn. Philadelphia: WB Saunders, 1990.
[10] Lazio BE, Stambough JL. An unusual posterior element spine tumor. Orthopedics 1994; 17: 725 ± 728.
[11] Bilge T, Barut S, Yaymaci Y, Alatli C. Solitary eosinophilic granuloma of the lumbar spine in an adult. Case report. Paraplegia 1995; 33: 485 ± 487.
[12] Mirra J. Bone tumors: diagnosis and treatment. Philadelphia: Lippincott, 1980.
[13] Villas C, Martinez-Perric R, Barrios RH, Beguiristain JL. Eosinophilic granuloma of the spine with and without vertebra plana: Long term follow-up of six cases. J Spinal Disord 1993; 6: 260 ± 268.

Figure 1



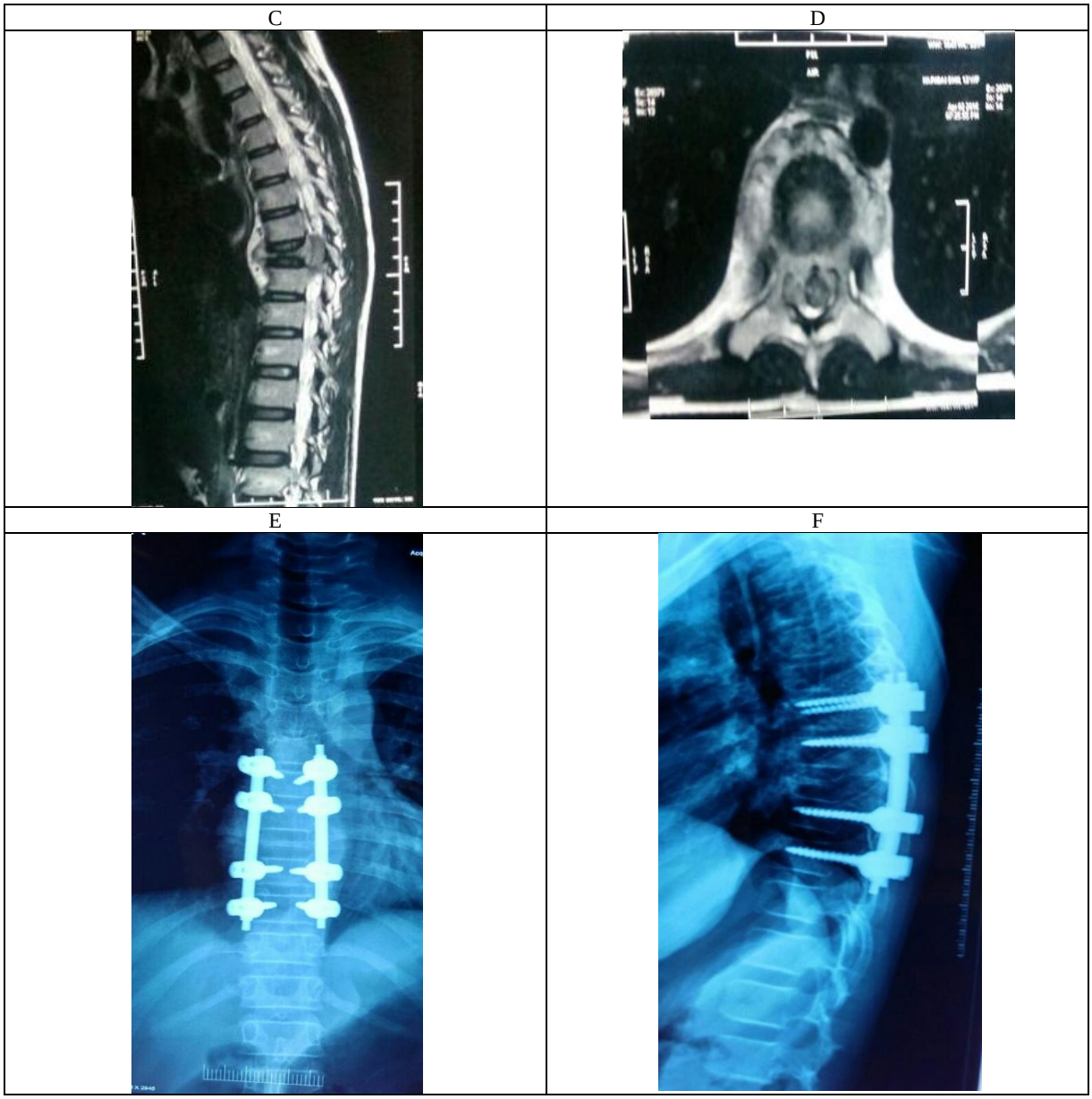


Figure A,B- shows the Antero-posterior and Lateral view of thoracic spine showing the almost complete collapse of the D8 vertebra
Figure C,D – shows T2 weighted sagittal and Axial images with D8 vertebra plana, note ht associated hyper-intense paraspinal and anterior epidural collection and also the maintained proximal and distal disc spaces. The compression involving the spinal canal is evident.
Figure E,F- showing the post operative AP and Lateral images showing the fixation from D6-D10 using pedicle screws.

Table 1

Differential Diagnosis	Features
Spine tuberculosis	Local pain, tenderness, stiffness, paraspinal muscle spasm, cold abscess, constitutional symptoms, neurological involvement, X ray: involvement of 1 or more vertebra with intervening disc
Eosinophilic Granuloma	Local pain, tenderness, no constitutional features, neurological involvement rare, X ray: vertebra plana, osteolytic lesions
Trauma	High velocity injury, neurological involvement
Osteogenesis Imperfecta	History of multiple fractures, limb bowing, ligament laxity, short stature, blue sclera, hearing loss, brownish opalescent teeth, X ray: cod fish vertebra, thin cortices, generalised osteopenia, saber shins, wormian bones in skull films
Acute Lymphoblastic Leukaemia	Fever, frequent infections, anaemia, generalised weakness, weight loss, peripheral smear examination and bone marrow biopsy for diagnosis
Hemangioma	Present as incidental finding or with pain or pathological fracture, X ray: lytic lesion, vertical striations giving honeycomb or jail bar appearance, may be multifocal
Ewing's Sarcoma	Pain, fever, mimics infection, local swelling, tenderness, raised ESR and WBC count, X ray: large destructive lesion in diaphysis or metaphysis, moth-eaten appearance, onion skin or sun burst type periosteal reaction