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# **UNUSUAL PRESENTATION OF OSTEOLASTOMA AS VERTEBRA PLANA – A CASE**

## **REPORT AND REVIEW OF LITERATURE:**

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### **Conflict of Interest**

1 None

2

3 **ABSTRACT:**

4 **Background:**

5 Osteoblastoma is rare and accounts for 3% of all benign tumours and 1% of all bone tumours.

6 The spine is the most common site of occurrence, constituting 32 to 45 % of all osteoblastomas.

7 It has a strong predilection for the posterior elements, most often occurring in the lumbar spine.

8 **Method:**

9 In this case report, we describe an unusual presentation of spinal osteoblastoma presenting as  
10 thoracic T9 vertebra plana in a 20-year-old female. She presented with discomfort over the  
11 midback with unsteadiness of gait. The patient underwent detailed investigations including CT,  
12 MRI and CT guided Biopsy. To our knowledge, this is the first case report of vertebra plana due  
13 to spinal osteoblastoma in the English literature.

14 **Result:**

15 The patient successfully underwent posterior decompression of T9 with laminectomy followed  
16 by MIS (Minimally Invasive Surgery) posterior instrumentation from T7 to T11. Histopathology of  
17 the intraoperative specimen was consistent with osteoblastoma. The patient had an uneventful  
18 postoperative recovery and no evidence of tumour recurrence could be demonstrated on PET  
19 scan at 15 months' follow-up.

20 **Conclusion:**

21 In conclusion, the differential diagnosis for vertebra plana is extensive and we add spinal  
22 osteoblastoma as another etiology to the existing list. Diagnosis and treatment of vertebra plana

involves multimodality radiological imaging, and careful histological and surgical evaluation to identify the underlying etiology.

**Keyword**

*Plana, Presentation, Osteoblastoma, Vertebra, excision*

### Introduction:

The term "vertebra plana" is the radiological description for partial destruction and flattening of a vertebral body with relative preservation of the adjacent intervertebral disc spaces. The commonest cause of vertebra plana in the pediatric and adolescent population is Langerhans cell histiocytosis. Malignant causes of vertebra plana include metastatic disease, lymphoma, leukemia, multiple myeloma and Ewing's sarcoma. Other, less common etiologies include trauma, Gaucher's disease, osteomyelitis, coccidioidomycosis, tuberculosis, myofibromatosis, aneurysmal bone cyst (ABC) and giant-cell tumour (GCT) [1]. We describe an unusual presentation of vertebra plana secondary to an osteoblastoma in a 20-year-old female. We have also briefly reviewed the pertinent literature.

### Case Report:

In February 2015, a 20-year-old female patient was referred to us from a general practitioner complaining of midback pain for 2 months duration. She also complained of intermittent numbness of both feet with occasional loss of balance on walking resulting in falls. She was both bowel and bladder continent. There was no significant past medical history. On examination, she had a gibbus deformity at T9 with localized tenderness on palpation. Detailed neurological examination revealed full power of both lower limbs with no objective sensory loss to touch and pain. However, proprioception and vibration were reduced in both lower limbs. Bilateral knee and ankle jerks were hyper-reflexic. In addition, a positive tandem and broad based gait were apparent.

Radiographs of the thoracolumbar spine showed solitary T9 vertebra plana with more than 80% loss of vertebral height and sclerosis. [FIGURE 1] This prompted us to do detailed workup to

rule out infection, primary tumour (both benign and malignant) and metastasis of T9 vertebra. Blood investigations for total cell count, ESR, and CRP were normal. Workup for multiple myeloma was negative. Tumour markers levels (CEA, CA125, AFP, beta hCG) were also within normal limits. Bone scintigraphy revealed intense radioactive tracer uptake (technetium-99m-MDP) at the T9 vertebra indicative of high bone turnover. [FIGURE 2] A CT scan showed T9 vertebra plana with expansile, mainly lytic soft tissue replacement of the bony trabeculae throughout the body, pedicles, laminae, transverse and spinous processes. Cortical breach was seen predominantly at the left pedicle with evidence of hyperdense soft tissue extension into the spinal canal.[FIGURE 3a] An MRI scan demonstrated isointense T1W signal with heterogeneous iso to hyperintense T2W signal throughout the collapsed T9 vertebral body and posterior elements.[FIGURE 3b] There was corresponding intense post contrast enhancement with pre and paravertebral soft tissue extension, and circumferential epidural extension enveloping and compressing the cord. The differential diagnosis included a GCT/ABC, Langerhans cell histiocytosis or a malignant entity such as lymphoma. CT guided biopsy was performed using a T9 right transpedicular approach. Histopathology was reported as an osteoblastic lesion most likely an osteoblastoma. Gram staining, aerobic, anaerobic, fungal and AFB (Acid Fast Bacillus) cultures were negative.

Upon discussion with the patient and their family the decision was made for surgical decompression. Posterior decompression of T9 with laminectomy followed by MIS posterior instrumentation from T7 to T11 [Longitude; Medtronic] was performed. In addition to thorough decompression of the thoracic cord and excision of the expanded posterior elements, transpedicular curettage of the vertebral body was done to ensure maximal removal of tumour. [FIGURE 4]

Histopathology of the intraoperative curetted specimens confirmed the diagnosis of osteoblastoma. It showed prominent new osteoid and anastomosing trabeculae of woven bone

formation by plump osteoblasts with vascular stroma. No significant atypia or plasma cells were noted. [FIGURE 5] Immunohistochemistry (CD1a, CD138, and AE1/3) were negative for Langerhans cell histiocytosis or metastatic carcinoma. After discussion at the multidisciplinary tumour board and with the patient, it was decided not to proceed with anterior vertebrectomy and to closely observe the patient for future recurrence with 6 monthly CT.

CT scan at 1-year follow-up showed evidence of reossification and did not reveal any evidence of tumour occurrence. [FIGURE 6] In addition, a PET scan at 15 months' follow-up did not show any suspicious tumour activity. [FIGURE 7]

## Discussion:

The term "vertebra plana" was first coined by Calve in 1925 to describe the pathology of aseptic necrosis of the vertebral body or osteochondritis [2, 3]. The classic radiographic findings of vertebra plana as described by Calve [3] and Buchman [2] include 1) involvement of only one vertebra, 2) lack of adjacent intervertebral disc involvement 3) widened intervertebral disc, 4) greater opacity or sclerosis of the involved collapsed vertebral body. The differential diagnoses of vertebra plana are many and a detailed workup including CT, MRI, bone scan and CT guided biopsy is of paramount importance.

Osteoblastoma is rare and accounts for 3% of all benign tumours and 1% of all bone tumours [8]. It is common in the second or third decades, although presentation at the extremes of age has also been described. It is twice as common in males as in females. The most common site of occurrence is the spine constituting 32 to 45 % of all osteoblastomas [8,24]. It has a strong predilection for the posterior elements, although uncommonly it can extend into the posterior vertebral body in more aggressive lesions. Anterior vertebral body involvement, as in this case is even less common [9-11]. They occur most often in the lumbar spine when compared to the other regions of spine.

In 1956, Jaffe and Lichtenstein differentiated osteblastoma from osteoid osteoma and identified it as a separate entity, although both are variants of the same basic lesional process of osteoblastic derivation [4-6]. Subsequently, Dorfman and Weiss introduced the entity 'aggressive osteblastoma' which was associated with a higher recurrence rate and risk of malignant transformation [7]. Osteblastoma can be classified into two subtypes; Conventional osteblastoma (stage 2 Enneking) and aggressive osteblastoma (stage 3 Enneking). Stage 2 lesions are contained within the bone and stage 3 lesions present with cortical breach and soft tissue extension. Hence, spinal deformities, neurological deficits and recurrence rates are higher in aggressive osteblastoma [8]. Boriani et al [9] reported paraparesis in 75% of patients with osteblastoma involving the thoracic spine secondary to cord compression (6 out of 8 patients). Ozaki et al [12] reported 69% incidence (9 out of 13 patients) of paraparesis in his study group of patients. The radiological findings in our patient correspond to an aggressive osteblastoma subtype with the presence of an expansile lesion with cortical breach, soft tissue mass and presence of subtle neurological findings.

Osteblastoma is a benign bone forming tumour with histology similar to osteoid osteoma. In comparison to osteoid osteoma, osteblastomas tend to be larger in size >2 cm, expansile and often present with cortical breach and a soft tissue mass. Spinal osteblastomas usually present with dull back pain. However, night pain and dramatic response of pain to NSAIDs, which are typical features of osteoid osteoma, are not commonly seen in osteblastoma. Scoliosis has also been described in osteblastoma, though less common than in osteoid osteoma [13]. Neurological symptoms are estimated to occur in greater than half of all patients with osteblastoma due to either pathological fractures or soft tissue extension producing mass effect.

Initial radiological work-up includes a plain radiograph to identify the lesion. Osteblastoma has a non-specific radiological appearance with some resembling osteoid osteoma with sclerotic

margins and others having periosteal new bone formation [14-17]. Contrast enhanced MRI is useful for evaluating the soft tissue extent of the lesion, although it can overestimate the aggressiveness of osteblastoma due to associated marked inflammation alluded by the authors as “Flare response” [18, 19]. MRI features are non-specific with typically low to isointense T1 and T2-weighted signal, decreased signal due to matrix calcification (if present) and intense enhancement representing the highly vascular nature of this lesion. CT is of immense help in delineating the site, nature and extent of bone destruction [18, 19]. Osteblastoma typically presents as a lytic expansile lesion on CT with a shell of sclerosis. Some lesions may show matrix calcification and cortical breach. Bone scintigraphy is non-specific and typically shows increased radiotracer uptake indicating increased osseous turnover [20]. Histology of osteblastoma is characterized by the presence of plump and epithelioid osteoblasts producing woven bone. Prominent nucleoli, larger trabeculae and invasion of cortical bone are seen in aggressive osteblastomas [8]. Histopathology of the intraoperative specimen in our case showed a similar picture compatible with diagnosis of osteblastoma.

Osteblastomas are usually managed surgically as this is the most effective treatment described in the literature [8, 12, 21, 22]. While marginal resection can be curative in osteblastoma, complete resection is not possible in certain locations such as the spine. In spinal osteblastomas total excision is the preferred treatment option as it has lowest chance of recurrence [23, 24]. However, some researchers have reported good results with subtotal excision or resection without wide margins [9, 25]. Intralesional curettage is done when en bloc excision is not possible. Among the postoperative adjuvant therapies, Radiotherapy is the most common modality but its role in the treatment to reduce recurrence is controversial. Some studies have found beneficial effects of Radiotherapy following subtotal excision [22], whilst others have shown no clear benefit of Radiotherapy following subtotal excision or in recurrent tumours [23,27,28]. In his review of 197 osteblastoma cases, Marsh et al., [24] concluded that

Radiotherapy does not alter the course of the disease and appears to be contraindicated. In addition, Radiotherapy is associated with the risk of post radiation sarcoma [8, 15, 23, 26]. With regard to Chemotherapy as post adjuvant therapy, there are only limited anecdotal case reports and its use is largely in patients with recurrence [28]. A systemic review from Harrop et al., states that there is only weak recommendation for Chemotherapy in osteoblastoma due to very low quality literature and may be of limited role in recurrent aggressive osteoblastoma [23].

#### **Conclusion:**

In conclusion, the differential diagnosis for vertebra plana is extensive and we add spinal osteoblastoma as one more etiology to the existing list. Diagnosis and treatment of vertebra plana involves multimodality radiological imaging, and careful histological and surgical evaluation to find out the underlying etiology.

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**Figure Legends:**

Figure 1. Radiograph depicting vertebra plana at the T9 vertebra.

Figure 2. Bone scan showing increased radionuclide tracer uptake at T9

Figure 3. a) Preoperative CT (sagittal and axial) showing an expansile, mainly lytic soft tissue replacement of the bony trabeculae throughout the body, pedicles, laminae, transverse and spinous processes

b) Preoperative Sagittal and axial MRI showing pre and paravertebral soft tissue extension, and circumferential epidural extension enveloping and compressing the cord

Figure 4. Postoperative radiograph with instrumentation in situ with “2 up – 2 down construct”

Figure 5. Histopathology showing plump osteoblasts with new osteoid and anastomosing trabeculae of woven bone suggesting Osteoblastoma

Figure 6. Postoperative CT scan (sagittal and axial) at 12 months follow-up showing reossification of the posterior vertebral body and pedicles post curettage of the lytic lesion

Figure 7. PET scan at 15 months' follow-up did not show any suspicious activity.