Telangiectatic osteosarcoma of the spine: a case report

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Abstract Telangiectatic osteosarcoma (TOS) of the spine is rare accounting for only 0.08% of all primary osteosarcomas. Though a well described radio-pathological entity it is not often thought of as a cause of paraplegia. We describe the clinical, radiological and pathological features and discuss the treatment options of telangiectatic osteosarcoma of the dorsal spine presenting in a young man. The diagnostic pitfalls are discussed emphasising the fact that the diagnosis of TOS of the spine requires not only a multi modal approach of appropriate radiological and pathological tests but also an awareness of this condition.

Keywords Telangiectatic osteosarcoma · Spine · Diagnosis · Treatment

Introduction

Telangiectatic osteosarcoma (TOS) of the spine is rare accounting for only 0.08% of all primary osteosarcomas [2]. Though a well described radio-pathological entity it is

not often thought of as a cause of paraplegia. The case of a young man with gradual onset paraplegia is presented here and in doing so we review the clinical, radiological and pathological features and discuss the treatment options. A word of caution is also made concerning the diagnostic pitfalls.

Case history

A 25-year-old farmer presented with a history of mid back pain for 3 months. A month later he developed weakness and numbness of both lower limbs which over a period of 1 week, progressed to complete paraplegia with loss of bowel and bladder function. A history of loss of weight and appetite was present but he gave no history of fever or cough. He had localised tenderness over the D₅–D₈ vertebrae. No obvious deformities or soft tissue swellings were noticed. He had complete loss of motor power and sensation below D₈ vertebra. A per rectal examination revealed an absence of sphincter tone.

His hematological parameters were unremarkable apart from a marginally increased alkaline phosphate = 130 U/L. Radiographs of the thoracic spine (Fig. 1a, b) showed destruction of D₆ vertebral body and its right pedicle. The D_{6–7} disc space was reduced on the right. A large pre and paravertebral component was seen in relation to D₆. The MRI (Fig. 2a, b) showed altered signal intensity seen within the D_{5,6} and D₇ vertebral bodies. There was complete destruction of the D₆ body (Fig. 2c) which was replaced by a large expansile lytic lesion. This showed multiple bony locules (soap bubble appearance) showing fluid–fluid levels. The locules were formed by thin expanded bone as seen on the CT sections taken prior to the biopsy (Fig 3).

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Fig. 1 X-ray AP and Lateral view showing destruction of D₆ vertebral body and its right pedicle. The D₆₋₇ disc space is reduced on the right. A large pre and paravertebral component is seen in relation to D₅₋₇ vertebra

Fig. 2 a, b MRI T₁ and T₂ weighted sagittal images shows altered signal intensity seen within the D_{5,6} and D₇ vertebral bodies. There was complete destruction of the D₆ body which was replaced by a large expansile lytic lesion. c Axial T₂ weighted image showing multiple fluid fluid levels within the lesion

A small soft tissue component was seen anterolaterally on the right. The epidural component caused severe canal narrowing compressing the cord. The expansile lesion also extended into the prevertebral region with significant mass effect on the aorta and lower trachea/carina. Large paravertebral extension, more on the right was also noted with extension to the adjacent ribs.

A CT guided needle biopsy (Fig. 3) was performed and aspiration of one of the locules revealed altered blood. Histopathological assessment of this specimen showed organizing fibrous tissue, giant cell reaction and osteoid production.

The diagnostic possibilities considered at this time were an aneurysmal bone cyst (ABC), giant cell tumour (GCT)

Fig. 3 CT Scan shows multiple locules formed by thin expanded bone and biopsy needle in situ

or a telangiectatic osteosarcoma. Since the findings were inconclusive the patient underwent an open biopsy. The intraoperative findings were suggestive of a highly vascular lesion which gave the impression of a lobulated mass with septations and bled copiously when penetrated by the probing finger. There were no definable margins to the mass. The tissue was sent for histopathological examination which revealed a tumour (Fig. 4a) showing multiple, aneurysmally dilated large blood filled septate spaces with spindle cell and osteoclast type multinucleate giant cells. Scattered markedly pleomorphic tumour cells with hyperchromatic mitotically active nuclei were present (Fig. 4b). In foci lace like osteoid admixed with tumour cells and reactive new bone were seen. A diagnosis of telangiectatic osteosarcoma was made.

Fig. 4 5× Scanner view of the tumour shows prominent blood filled septate spaces, similar to an aneurysmal bone cyst. (H&E, ×50)

Fig. 5 40× Septa contains some markedly pleomorphic cells with hyperchromatic nuclei, mitotic figure and lace like osteoid. (H&E, ×400)

The condition was explained and the patient was advised palliative chemotherapy and radiotherapy. He however, declined further treatment and was discharged. At one year a postal intimation indicated that the patient had expired.

Discussion

Many different subtypes of osteosarcoma have been described including, high-grade intramedullary, paraosteal, periosteal, intracortical, small cell, low-grade intramedullary and telangiectatic osteosarcoma [2]. Telangiectatic osteosarcoma is a well recognized, although unusual entity, representing 2.5–12% of all lesions [1–4].

Telangiectatic osteosarcoma is primarily a tumour of the long tubular bones [9, 14], the femur is involved most frequently, followed in frequency by the tibia and humerus [9], a distribution similar to that of conventional osteosarcoma. In these bones the metaphysis is the usual site of origin [9, 10, 15]. Other bones such as the sternum, scapula [6] ribs, innominate bone [6], cranial vault [15] and mandible may likewise be involved. TOS of spine is exceedingly rare [5]. The table below summarises some of the other reported cases of TOS of the spine in the English literature (Table 1).

The awareness and recognition of the unique histological and radiological features of spine TOS is crucial for making an early and accurate diagnosis [14]. It stands without saying that delaying the correct diagnosis and initiation of proper therapy may have a negative impact on survival of these patients [14].

The aggressive, expansile, osteolytic nature of this condition is the radiographic hallmark of telangiectatic

Table 1 A literature review of other cases of TOS of the spine

Author	Level (n)	Surgical treatment	Outcome
Weiss et al. [14]	Sacrum (1)	None	Not discussed
Murakami et al. [5]	Thoracic (2)	Total en bloc spondylectomy	Expired at 6 and 4 months
Ilaslan et al. [2]	Not discussed (4)	Not discussed	Not discussed
Murphey et al. [6]	Lumbar (1)	Not discussed	Not discussed
Simon and Irwin [9]	Sacrum (1)	Partial sacrectomy	Survival at 5 years
Nishida et al. [7]	Thoracic (1)	Excision, fusion	Survival at 4 years
Present	Thoracic (1)	Laminectomy, curettage	Expired within 1 year (date unknown)

osteosarcoma [2, 4, 15]. Intralésional sclerosis is infrequent and mild. In fact in the Mayo Clinic series, the presence of bone sclerosis was grounds for eliminating telangiectatic osteosarcoma from diagnostic consideration [4].

Magnetic resonance imaging may show areas of high signal intensity on T₂-weighted as well as T₁ weighted images (a feature that is unusual with other neoplasm) [6] and the presence of fluid–fluid levels. Fluid–fluid levels are suggestive of haemorrhage. The anterior portions are bright, reflecting the supernatant whereas the posterior portions are dark representing the clot [14]. Fluid–fluid levels in blood filled cavities are a characteristic although nonspecific finding in aneurysmal bone cysts [2], giant cell tumours and telangiectatic osteosarcomas. These have also been reported in other tumours or pseudotumours including chondroblastoma, osteoblastoma, myositis ossificans, fibrous dysplasia [12, 15].

The dilemmas associated with the diagnosis of TOS has been emphasized in literature [10, 14]. In particular an ABC is almost indistinguishable from TOS on radiographs [10] and hence the correct diagnosis rests on a careful histological examination [14]. The low power appearance of TOS is identical to that of ABC. TOS is characterized by multiple aneurysmal dilated blood filled cavities with high grade, sarcomatous cells in the peripheral rim and septa [6]. The distinction must ultimately rest on the finding of these obviously anaplastic cells [10], atypical mitotic figure and irregular closely packed lace like osteoid [1]. Therefore a combined modality approach, including the use of strict pathologic and radiological criteria, is crucial in diagnosing telangiectatic osteosarcoma [14]. These criteria as defined in the World Health Organization Classification [13] are: (1) predominantly lytic bone mass with minimal sclerosis on radiographs, (2) grossly cystic medullary mass with no or minimal solid or sclerotic component and (3) histological features consisting of bone forming tumour with notable blood filled spaces separate by septae lined by and/or containing malignant tumour cells with prominent nuclear atypia and limited osteoid deposition.

In the Indian setting where tuberculosis is endemic and spinal tuberculosis is by far the most common cause of

paraplegia [13] any young adult from a low socioeconomic background is considered to have spinal tuberculosis unless otherwise proved. The roentgenographic appearance in this patient may be mistaken for a central type of spinal tuberculosis where the disc spaces are by and large preserved [13]. Similarly the soft tissue shadows may appear like a tubercular abscess. With hindsight however; the predominantly right sided mass is unusual for an abscess. But perhaps the most misleading factor in the initial diagnosis of this condition was the rarity of the lesion at this site.

There have been very few reports that have described the management of spinal TOS [5, 7]. Conventionally, palliative surgery such as curettage or piecemeal excision for vertebral tumours has been commonly practiced because the anatomic peculiarity of the spine made it difficult to perform an oncologically appropriate excision of the tumour [5]. The disadvantage of this method is the high incidence of local recurrence caused by tumour-cell contamination. To avoid this mishap as far as possible, Tomita et al. [11] developed a new surgical technique of spondylectomy (vertebrectomy), termed ‘total en bloc spondylectomy’ (TES) for malignant or aggressive benign tumours of the spine [11]. The lesion is removed en bloc with attached barriers by TES. However even with this technique the survival rates of the two patients reported by Murakami et al. [5] was 4 and 6 months only. Furthermore, the authors felt that this surgical procedure was ‘overly invasive’ and that ‘deterioration of their immune response or hyper secretion of an adrenocorticotrophic hormone with these overly invasive surgeries might have a certain influence on general condition’ [5].

In addition to surgical excision, adjuvant chemotherapy plays an important role in the treatment of osteosarcoma. Huvos et al. [1] first made the observation that TOS was particularly sensitive to chemotherapy. They hypothesised that the increased vascularity of the tumour increases drug delivery to the tumour site therefore TOS may be more chemoresponsive than other osteosarcoma subtypes [8]. Nishida et al. [7] reported an 8-year-old boy with TOS of D5 who was well 4 years after excision of the mass and

posterior fusion, along with adjuvant chemotherapy and radiotherapy.

The prognosis of patients with telangiectatic osteosarcoma though initially was believed to be bad [3] no longer appears to be true in telangiectatic osteosarcoma of the appendicular skeleton [1, 14]. However, the prognosis of patients with TOS of the spine remains dismal. Long-term survival with Telangiectatic osteosarcoma of the spine has only rarely been reported [7, 9]. Our patient too expired within 1 year.

Our initial difficulty in reaching a diagnosis in this patient were two fold, first we were mislead into ‘flowing with the tide’ and given the sheer magnitude of numbers believed this to be tuberculosis and the early X-ray features of disc space reduction and contiguous vertebral involvement may have suggested an infective condition. Then the rarity of this condition prevented us from considering it in the early clinical diagnosis.

This therefore emphasises that the diagnosis of TOS of the spine requires not only a multi modal approach of appropriate radiological and pathological tests but also an awareness of this rare condition coupled with a reasonable degree of suspicion.

Conflict of interest statement None of the authors has any potential conflict of interest.

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