Malignant Transformation of Thoracic Spinal Chordoma

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ABSTRACT— ***Chordoma of the thoracic spine is an uncommon low-grade malignant tumour of bone. It can undergo transformation into a sarcoma (dedifferentiated chordoma), characterised by an aggressive, clinical course. Sarcomatous transformation is thought to stem from exposure to therapeutic radiation although recent experience suggests it can occur de-novo, or at the site of tumour recurrence. Most dedifferentiated chordomas arise from the sacrococcygeal spine and only three elderly patients were reported to suffer from this form of chordoma of the thoracic spine. This case report is of interest as the disease affected a 15-year-old-boy. He presented with progressive paraparesis. Computed tomography revealed a bone-eroding tumour at T4, characteristic of a chordoma. Despite radical excision and adjuvant radiotherapy, it was discovered he had metastasis in bone and lymph nodes five months later. The tumour had probably dedifferentiated into a sarcoma.***

Keywords— dedifferentiated chordoma; cord compression; thoracic spine; tumour recurrence

1. INTRODUCTION

Chordoma is a rare, low-grade malignant bone tumour arising from remnants of primitive notochordal rests. There is an incidence of 0.8 per one million in the population with a peak age of 50 to 60 years [1]. Less than 5% of cases occur in children and adolescents [2]. The sites of origin in descending order are the sacrococcygeal region (49%), skull base (36%) and mobile spine (15%) [3]. Chordoma of the thoracic spine is the least common with 37 cases documented in one study in 2007 [4]. A rare complication of a thoracic chordoma is sarcomatous transformation (dedifferentiated chordoma) of which only 3 adult patients have been fully described [4]. The following case is of distinct interest since the tumour affected a teenager.

2. CASE REPORT

A 15-year-old boy presented at this institution with a 3-month history of progressive weakness of his lower limbs and paraesthesia from his mid thorax downwards. Physical examination revealed altered sensation to touch and pain below the umbilicus and bilateral extensor plantar response, findings indicative of thoracic cord compression. Computed tomography (CT) showed a soft tissue tumour at T4 vertebra, eroding its left neural arch and pedicle. There is indentation on the cord. Paravertebral extension and erosion of posterior end of the left 4th rib were evident (Figure 1). Emergency de-compressive surgery to relieve cord compression was performed. Two weeks later a T4 vertebrectomy together with excisions of T3/T4 intervertebral disc, lower T3 vertebra and the tumour's paravertebral component were carried out. Histopathology revealed a mixed pattern: some cellular elements were suggestive of epithelioid sarcoma (Figure 2). However there were sheets and cords of large regular cells with eosinophilic cytoplasm and abundant mucoid matrix that favoured a chordoma although no physaliferous cells were identified. The patient made a good recovery following a course of radiation therapy 8 weeks after surgery. But within 6 months he was found to have hypercalcaemia and an osteolytic lesion in his left 6th rib as well as an enlarged lymph node in the left supraclavicular fossa. The oncology team revised their diagnosis to a sarcoma with metastases. He was treated with 5 cycles of chemotherapy. As his hypercalcaemia returned to normal, his clinical status stabilised. Four months later he experienced a return of his paraplegia that progressed rapidly. Repeat CT spine revealed a bone-eroding tumour on the left paravertebral space at T5 with destruction of the corresponding posterior rib (Figure 3), findings consistent with recurrent disease. His hypercalcaemia returned. Considered too weak to undergo surgery, he was treated with a second course of chemotherapy, consisting of Vincristin, Cyclophosphomide and Actinomycin D that proved ineffective. His poor state of health deteriorated further and he died 4 months later.

3. DISCUSSION

There are no detailed reports on dedifferentiated chordomas of the thoracic spine in paediatric and adolescent age groups although two series [5, 6] listed data of their patients' histopathological grades, treatment protocol and clinical outcome. Of the 6 with thoracic vertebral chordomas in the larger series [6], 2 male patients suffered from the dedifferentiated form. One, aged 20, was lost to follow-up after subtotal resection while the other aged 16 underwent total resection followed by radiation therapy but died of disease 40 months after initial diagnosis. Of the 12 patients in the smaller series [5] there were 2 children with chordomas in the lower thoracic spine. The first, female aged 6 had a conventional chordoma at T10/T11 while the second, female aged 11 months, had a "poorly" differentiated or sarcomatoid form of the disease at T12. She underwent a partial resection of her tumour but died of the disease a year later. Both groups of authors concluded that among patients in their first and second decades of life there is a subset of histologically "atypical" chordomas with a significantly worse prognosis than that of the conventional forms.

Vertebral destruction and cord compression by a soft tissue tumour are the recognized imaging features of a spinal chordoma [4]. Among adolescents and young adults some other spinal diseases can be mistaken for a chordoma. A prominent pre or paravertebral mass, similar to that of the present case, is a known finding in benign conditions such as aneurysmal bone cysts, giant Langerhans cell histocytosis and giant cell tumour of bone [7]. Spinal malignancies simulating chordomas include solitary secondary deposits, and less frequently Ewing's sarcoma and the osteolytic form of osteosarcoma—the latter with preferential involvement of metaphyses of long bones, can affect the adolescent spine [8].

The overall signal intensities of chordomas on MRI are either iso or hyperintense in T1-weighted images and hyperintense in T2-weighted sequences. Contrast enhancement of chordomas is variable and at best moderate [9]. Absence of contrast enhancement makes the diagnosis of a spinal chordoma more likely as most other spinal tumours of adolescent age enhance markedly and heterogeneously [7]. Moreover chordoma is the only lesion with characteristic intralesional septations in T2-weighted sequences. Recently diffusion weighted MRI has proven to be reliable in distinguishing conventional chordoma from the dedifferentiated form [10]. Increased cellularity in dedifferentiated chordomas causes diffusion restriction and low signal intensities in T2-weighted images [11] and a corresponding decrease in ADC values in comparison with that of conventional chordomas [10].

A potentially unfavourable outcome in spinal chordomas is that 60% of patients will develop distant metastasis in the course of their illness [12]. One prognostic indicator is the presence of marked prevertebral tumour extension [12] that requires en block excision, interbody fusion and stabilisation [13]. Another critical feature is the tendency of the lesion to invade intervertebral disc spaces and adjacent vertebrae [2]. This is illustrated in the present case in which vertebral and paravertebral involvement necessitated removal of the T3/T4 disc, part of T3 as well as a T4 vertebrectomy. Such aggressive surgery is advocated to prevent local recurrence that is the most important predictor of mortality [2].

In the later course of the patient's illness hypercalcaemia was a point of concern. In a setting of supraclavicular lymphadenopathy and an osteolytic rib lesion it was evident the chordoma had turned anaplastic [14]. Although metastasis from a malignant chordoma or sarcoma is not among the common lesions to cause hypercalcaemia [15], the decision to treat him with chemotherapy was justified. But when he was stricken with tumour recurrence 4 months later, his situation was dire. It is conceivable during the patient's first presentation his tumour at T4 vertebra was a chordoma at an early stage of sarcomatous transformation.

Of the 3 patients in the literature whose thoracic spinal chordomas had become sarcomatous all were given radiation therapy. The first case related to a 67-year-old female whose chordoma at T12 vertebra was treated solely with radiotherapy. Her tumour turned sarcomatous 43 months later and she succumbed to this soon after. There was an attempt to link previous radiation exposure to malignant change but the requisite time interval was insufficient for it to take place [16]. The other 2 cases were elderly males [17, 4] with mid thoracic spine chordomas that had radical surgery and adjuvant radiotherapy. These authors did not attribute the pathogenesis of sarcomatous change in their patients to previous radiation because the latent period was only 3 years. However the ultimate clinical course of both patients bore a resemblance to the present case as both died within 5 months of tumour recurrence at site of previous surgery.

4. CONCLUSION

The clinical and radiological features of the present case and the experience of other workers suggest that the prognosis for dedifferentiated chordoma of thoracic spine is poor.

5. REFERENCES

1. McMasters ML, Goldstein AM, Bromley CM, Ishibe N, Parry DM. Chordoma: incidence and survival patterns in the United States, 1973-1995. Cancer Causes Control 2001; 12(1): 1-11.

2. Walcott BP, Nahed BV, Mohyedin A, Coumans JV, Kahle KT, Ferreira MJ. Chordoma: current concepts, management, and future directions. Lancet Oncol. 2012; 13(2): e69-76.
3. Heffelfinger MJ, Dahlin DC, MacCarty CS, Beabout JW. Chordomas and cartilaginous tumors of the skull base. Cancer 1973; 32(2): 410-420.
4. Bisceglia M, D'Angelo VA, Guglielmi G, Dor DB, Pasquinelli G. Dedifferentiated chordoma of the thoracic spine with rhabdomyosarcomatous differentiation. Report of a case and review of the literature. Annals of Diagnostic Pathology 2007; 11(4): 262-273.
5. Coffin CM, Swanson PE, Wicks MR, Dehner LP. Chordoma in childhood and adolescence. A clinicopathologic analysis of 12 cases. Arch Pathol Lab Med. 1993; 117(9): 927-933.
6. Ridenour RV, Ahrens WA, Folpe AL, Miller DV. Clinical and histopathologic features of chordomas in children and young adults. Pediatric and Developmental Pathology 2010; 13(1): 9-12.
7. Rodellac MH, Feydy A, Larousserie F, Anract P, Campagna R, Babinet A, Zins M, et al. Diagnostic imaging of solitary tumors of the spine: what to do and say. Radiographics 2008; 28(4): 1019-1041.
8. Katonis P, Datsis G, Karantanas A, Kampouroglou A, Lianoudakis S, Licoudis S, Papoutsopoulou E, et al. Spinal osteosarcoma. Clinical Medicine Insights: Oncology 2013; 7: 199-208.
9. Smolders D, Wang X, Drevelengas A, Vanhoenacker F, De Schepper AM. Value of MRI in the diagnosis of non-clival, non-sacral chordoma. Skeletal Radiol. 2003; 32(6): 343-350.
10. Yoem KW, Lober RM, Mobley BC, Harsh H, Vogel H, Allagio R, Pearson M et al. Diffusion-weighted MRI: Distinction of skull base chordoma from chondrosarcoma. AJNR Am J Neuroradiol. 2013; 34(5): 1056-1061.
11. Hanna SA, Tirabosco R, Amin A, Pollock RC, Skinner JA, Cannon SR, Saifuddin A, et al. Dedifferentiated chordoma. A report of four cases arising 'de novo'. J Bone Joint Surg (Br). 2008; 90-B(5): 652-656.
12. Sunderesan N, Galicich JH, Chu FCH, Huvos AG. Spinal chordomas. J Neurosurgery 1979; 50(3): 312-319.
13. Topsakal C, Bulut S, Erol FS, Ozercan I, Yidirim H. Chordoma of the thoracic spine—case report. Neurol Med Chir (Tokyo) 2002; 42(4): 175-180.
14. Chambers PW, Schwinn CP. Chordoma. A clinicopathologic study of metastasis. Am J Clin Pathol 1979; 72(5): 765-766.
15. Clines GA, Guise TA. Hypercalcaemia of malignancy and basic research on mechanisms responsible for osteolytic and osteoblastic metastasis to bone. Endocr Relat Cancer 2005; 12(3): 549-583.
16. Fox JE, Batsakis JG, Owano LR. Unusual manifestations of chordoma. A report of two cases. J Bone Joint Surg Am. 1968; 50(8): 1618-1628.
17. Nanda A, Hirsh LF, Antoiniades K. Malignant fibrous histiocytoma in a recurrent thoracic chordoma: case report and literature review. Neurosurgery 1991; 28(4): 588-592.

6. FIGURES

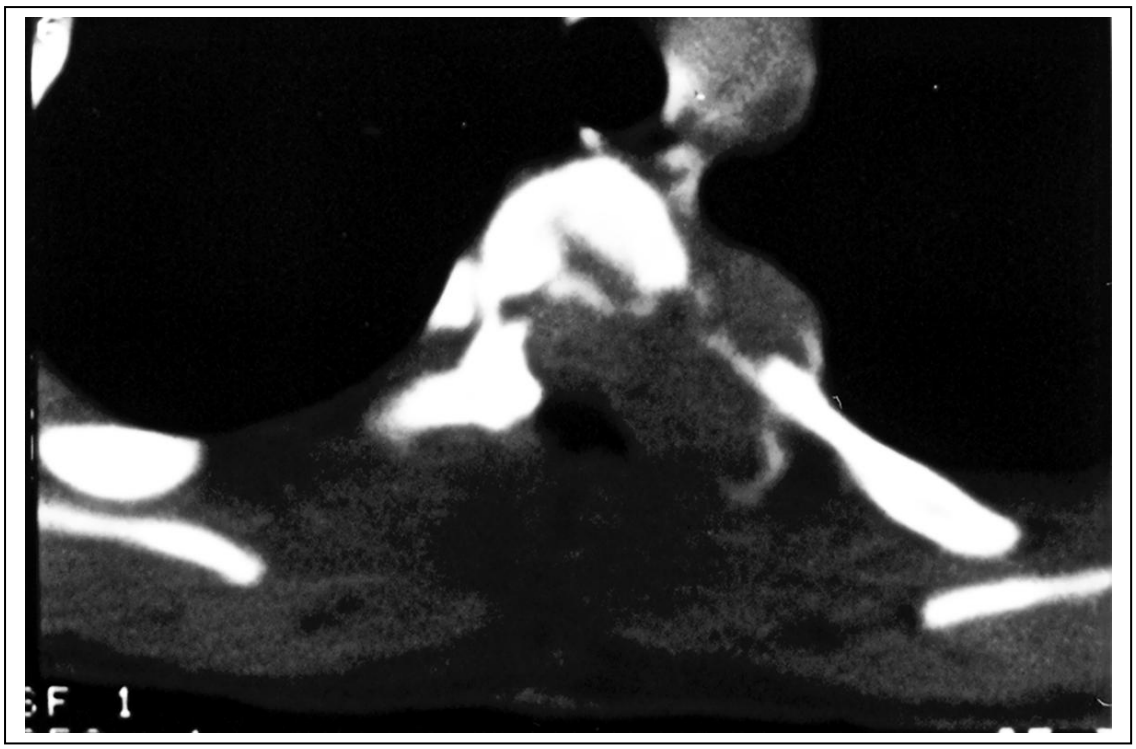

Figure 1: Axial CT through the upper level of T4 vertebra shows a soft tissue mass eroding the left pedicle with extension into the spinal canal and paravertebral space.

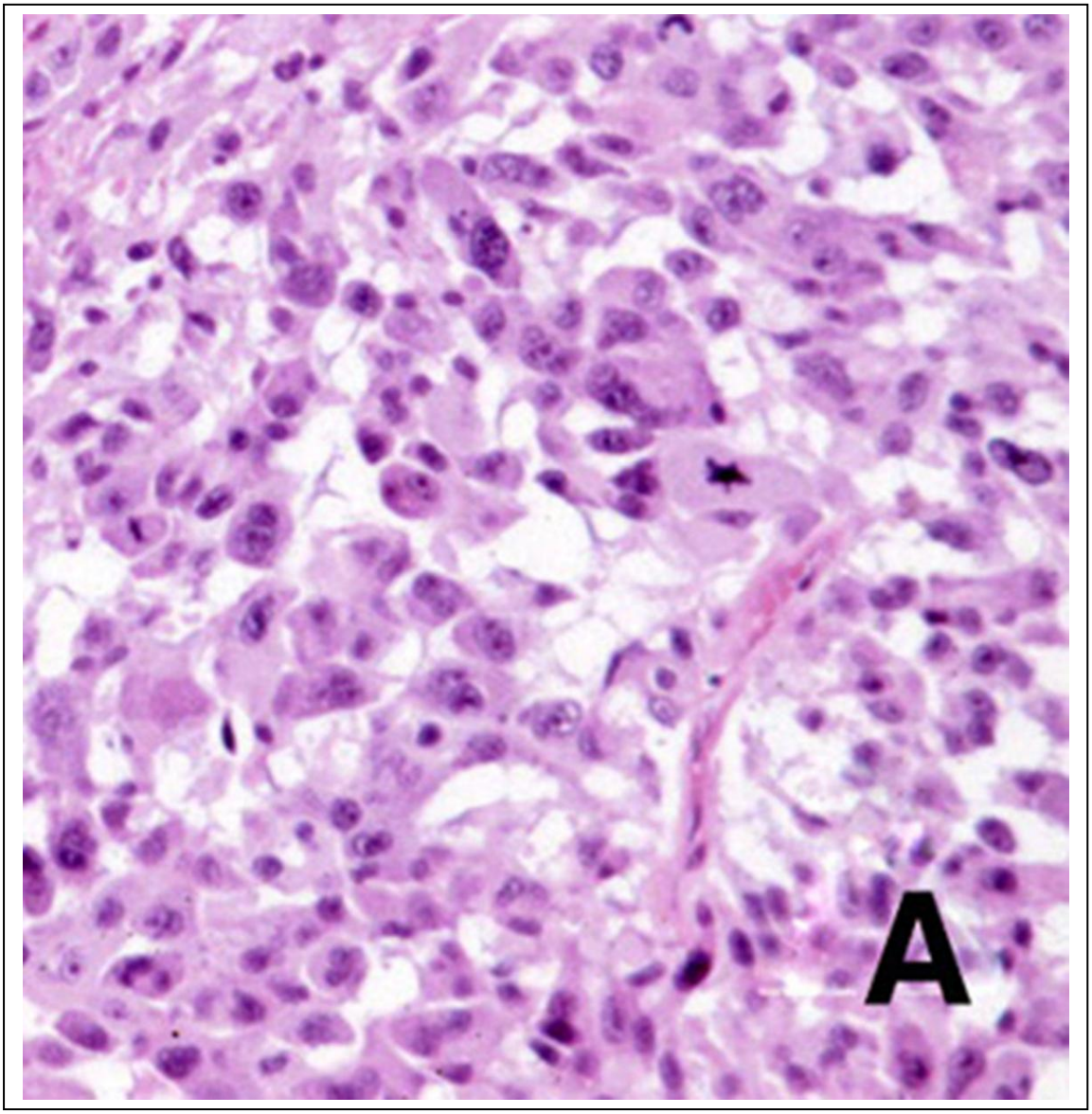

Figure 2: Histopathology reveals a mixed pattern of epithelioid sarcoma and pleomorphic chordoma. (By kind permission of Elsevier).

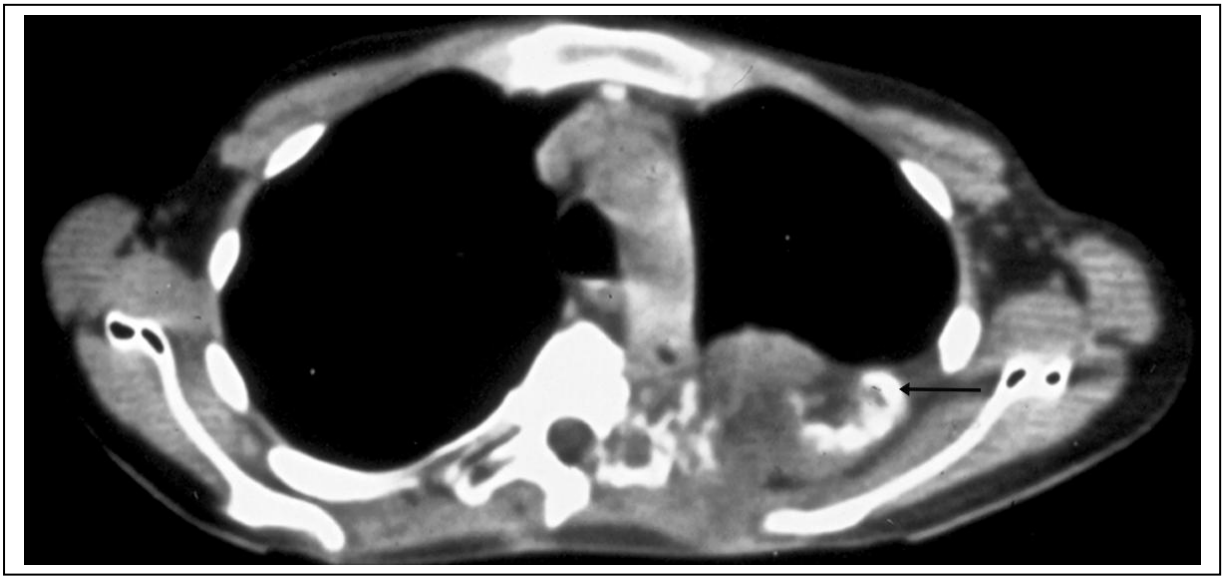

Figure 3: Axial CT through level of body of T5 vertebra showing destruction of left posterior rib by recurrent tumour mass. Note small lunar-shaped calcification at its lateral aspect (arrow). Calcification occurs typically in recurrent disease.