CASE REPORT

Giant-cell tumour of the synovium in a facet joint in the thoracic spine of a child

Giant-cell tumour of the synovium is known to affect the fingers or toes of adults. It has seldom been described in the spine and rarely in the thoracic vertebrae or in a child. The lesions of giant-cell tumour of the synovium have a classical radiological appearance, but require a high index of suspicion for correct recognition. Unlike giant-cell tumour of the synovium at other well-known sites, spinal lesions lack the characteristic papillary architecture, thereby raising other diagnostic possibilities. We describe a giant-cell tumour of the synovium of the left facet joint of a thoracic vertebra in a nine-year-old girl. The tumour was treated successfully by surgical excision.

Giant-cell tumour of the synovium is fairly common, usually affecting adults over 30 years of age and involving the fingers, wrist or knee. It occurs in the thoracic spine of a child very rarely.

We report a localized type of giant-cell tumour of the synovium involving a thoracic facet joint of a young child and presenting with sensory ataxia.

Case report

A nine-year-old girl presented with persistent back pain and reduced sensation in the lower limbs over a period of five months. The pain was localized to the seventh and eighth thoracic vertebrae (D7 and D8) and there was sensory ataxia. Magnetic resonance imaging of the spine showed an extradural mass measuring 2.9 cm × 1.5 cm × 1.1 cm in the posterior aspect of the dorsal spinal canal at the level of the D7 to D9 vertebrae. It was iso-intense with the spinal cord on T1-weighted images and markedly hypointense on T2-weighted images (Fig. 1a). The lesion was well circumscribed, solid and showed intense homogeneous post-contrast enhancement (Fig. 1b). Although it caused compression of the adjacent thecal sac, it did not extend beyond the spinal canal.

On the parasagittal MR scan, the tumour appeared to originate from the left facet joint at D8 to D9 (Fig. 2a). Abnormal intensity of the marrow-signal in the left-sided posterior elements of the D8 and D9 vertebrae (Figs 2a and 2b) prompted a CT study which confirmed that the lesion was adjacent to the left D8/D9 facet joint, and had caused osteolysis of the left posterior elements of D8 and D9 (Fig. 2c).

The mass was excised completely through a posterior approach. This gave relief from all her symptoms. On macroscopic examination, the lobulated, partially encapsulated, soft to firm specimen measured 3 cm × 1.5 cm × 1.5 cm in size and had a yellowish-white homogeneous cut surface. Histologically a uniform appearance was seen throughout (Fig. 3). It comprised intricately mixed spindle cells with pale vesicular bean-shaped nuclei and multi-

Figure 1a – Sagittal T2-weighted scan of the dorsal spine showing an extradural mass in the posterior aspect of the dorsal spinal canal. It has low T2 signal intensity and compresses the adjacent dorsal spinal cord. Figure 1b – Post-contrast T1-weighted sagittal scan showing intense homogeneous enhancement of the mass.

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Figure 2a – Left parasagittal T2-weighted scan showing abnormal intensity of the marrow signal in the left-sided posterior elements of the D8 vertebra (arrow). The lesion appears to arise from left D8 to the D9 facet joint. Figure 2b – Axial T1-weighted scan showing abnormal marrow signal intensity in the left-sided posterior elements of the D8 vertebra (arrow). Figure 2c – Axial CT scan in bone window settings confirming the presence of an osteolytic lesion in the left transverse process of the D8 vertebra (arrow).

Figure 3a – Photomicrograph of the periphery of the lesion with encapsulation (haematoxylin and eosin x 40). Figure 3b – Photomicrograph showing a uniform appearance throughout with intricately admixed spindle cells, and multinucleate osteoclast-like giant cells on a collagenised background. There is a lack of papillary or villiform architecture (haematoxylin and eosin x 100). Figure 3c – High magnification view showing pale vesicular bean-shaped nuclei of the mononuclear cells (haematoxylin and eosin x 400). Figure 3d – Photomicrograph showing scanty haemosiderin pigment (black arrow) at the periphery along with thick-walled vessels and local aggregates of lymphocytes (white arrow) (haematoxylin and eosin, x 400).
nucleate osteoclast-like giant cells on a collagenised background (Figs 3b and 3c). Thick hyalinised bands traversed the lesion, imparting a vaguely nodular appearance. There was no necrosis and the most cellular areas had a mitotic count of not more than two per high-power field. At the periphery, there was a thick fibrous capsule (Fig. 3a). Within this, a few thick-walled hyalinised vessels and focal lymphoid aggregates, scanty foam cells and some haemosiderin pigment were observed (Fig. 3d). There were no papillary or villiform structures or any cholesterol clefts. Immunostains for CD 68 highlighted the pale histiocytic cells and osteoclast-like giant cells. A diagnosis of a localised type of giant-cell tumour involving the left facet joint of the eighth thoracic vertebra was made. At the last follow-up, 18 months post-operatively, the child had no residual disease or symptoms.

Discussion

In 1941, Jaffe, Lichtenstein and Sutro\(^2\) coined the term pigmented villonodular synovitis (PVNS) to encompass all proliferative lesions of the synovium of joints, tendons and bursae. More recently, the World Health Organisation has subtyped giant-cell tumours of the tendon sheath into localised and diffuse types, based on the small size and circumcision of the former, compared with the destructive and infiltrative growth pattern of the latter which is synonymous with PVNS.\(^1,3\) The localised type (giant-cell tumour of the synovium) has a predilection for tendon sheaths in the fingers,\(^4\) whereas the diffuse type commonly involves the knee, in which lack of resistance may lead to a large size.\(^4\) The spine is an unusual site for either variety and vertebral lesions may be compact, because of the natural restrictions of the spine.\(^5\)

Of the 46 cases of spinal giant-cell tumour of the synovium/PVNS in the English literature,\(^4-25\) only eight involved the thoracic vertebra (Table I). Giant-cell tumour of the synovium is rare in children of less than ten years of age, with only one documented case to our knowledge.\(^22\) Pigmented villonodular synovitis of the spine usually presents with pain and/or nerve-root claudication\(^5\) and compression of the dorsal columns may lead to sensory ataxia, as in our case.

Radiologically, a soft-tissue mass related to a facet joint with a profoundly low T2 signal intensity is characteristic of PVNS. Such lesions typically contain areas of intermediate and/or low signal intensity on T1- and T2-weighted images which become more pronounced on the long time to repetition (TR)/echo time (TE) images because of the preferential shortening of T2 relaxation times of haemosiderin.\(^24,25\) After contrast administration, PVNS often shows marked enhancement.\(^21\) In their retrospective review of 15 cases of spinal PVNS, Motamedi et al\(^21\) showed that determination of origin from a facet by imaging was dependent on the imaging modality and the

<table>
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<th>Authors</th>
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\(^{1}\) NK, not known
\(^{2}\) GTR, gross total resection; SR, subtotal resection; RR, radical resection; en bloc resection; CT, chemotherapy; RT, radiotherapy
\(^{3}\) NED, no evidence of disease; AWD, alive with disease
size of the lesion. In our case, the relationship to the facet joint was established by CT and MRI. The mass-like nature of PVNS often appears as bony erosions on both sides of the joint. This feature along with osteolysis on CT and an abnormal marrow signal on MRI were all seen in our case.

Giant cell tumour of the synovium may lack the classical papillary/villiform architecture and haemosiderin pigment; inflammatory cells may be scanty. In a series of 15 cases of giant-cell tumour of the synovium of the spine, only one was found to have a papillary architecture. The absence of these features raises other diagnostic possibilities such as tuberculosis, Langerhan’s cell histiocytosis, aneurysmal bone cyst and giant-cell tumour.

Detailed accounts of the management and prognosis of giant-cell tumour of the synovium of the spine are lacking and complete surgical resection, as performed in our case, is considered to be the treatment of choice. The local recurrence rate of 18% of spinal giant-cell tumour of the synovium is comparable with that of similar lesions (17% to 48%) occurring elsewhere and excision appears to be curative. In the case of infiltrative lesions, which preclude complete resection, chemotherapy with imatinib mesylate has recently been described.

Giant-cell tumour of the synovium and PVNS are considered to represent the same pathological process and unification of the two entities has been suggested. Our case of an otherwise common entity is presented because of the unusually young age of the patient and the rare site of the tumour.

No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

References
2. Jaffe HL, Lichtenstein L, Sutro CJ. Pigmented villonodular synovitis, bursitis and tenosynovitis: a discussion of the synovial and bursal equivalents of the tenosynovial lesion commonly denoted as xanthoma, xanthogranuloma, giant cell tumor or myxoplaoma of the tendon sheath, with some consideration of this tendon sheath lesion itself. Arch Pathol 1941;31:731-65.