Preadolescent presentation of a lumbar chordoma: results of vertebrectomy and fibula strut graft reconstruction at 8 years

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Abstract Chordoma is a tumour of notochordal origin which usually involves the sacrum or skull base presenting in adulthood. Chordoma in a mobile spinal segment is infrequent and the authors report an extremely rare presentation of L3 chordoma in a child aged 7 years. Although a benign tumour, mobile segment chordoma is more locally aggressive, more likely to metastasise and has a poorer 5 year survival than sacral and clival lesions. Wide surgical excision and reconstruction is the treatment of choice in vertebral chordoma. This case was treated with staged vertebrectomy and fibular strut graft reconstruction and the results of clinical and radiological follow up at 8 years are presented.

Keywords Chordoma · Benign tumour · Mobile spinal segment · Vertebrectomy · Fibular strut graft reconstruction

Case report

A previously well 7-year old child presented with a 6 month history of right anterior thigh and knee pain. Examination revealed a marked list to the right, painful restriction of spine movements in all directions, an absent right knee jerk with reduced sensation in the right L3 distribution. There was no motor weakness or sphincter disturbance. Back pain restricted straight leg raise bilaterally to 30°. No history of weight loss, sys-

temic symptoms and no past medical and family history of relevance. Blood tests were within normal limits. Inflammatory markers were not elevated. Plain radiographs revealed an osteolytic lesion in the posterior body of L3. MRI of the whole spine identified an isolated lesion at L3 with right pedicle involvement and tumour extension anterior to the dural sac at L4 (Fig. 1). Isotope bone scan showed no increased uptake. A staging CT of the chest showed no evidence of metastases.

Transpedicular biopsy at L3 was performed and histology revealed fragments of remodelling bone, haemorrhage and a myxoid tissue containing strips and ribbons of cohesive, vacuolated cells. These cells had the appearance of notochordal-type tissue. Immunohistochemistry was positive for Cytokeratin S-100.

In view of the histological findings and atypical young age of presentation a radical 360° L3 vertebrectomy with pedicle screw stabilization and fusion was planned.

At operation the L3 tumour had extended through the lamina and transverse process on the right compromising the right L3 nerve root. There was both cranial and caudal extradural extension of the tumour anterior to the dural sac with tenting from L2 to L4. A wide decompression was performed with resection of the posterior elements and partial posterior vertebrectomy at L3. To ensure that the tumour within the spinal canal was completely excised it was necessary to resect the inferior lamina of L2 and most of the lamina at L4 bilaterally. Hence it was felt appropriate to extend the instrumentation inferiorly to L5.

Pedicle screw instrumentation was performed at L2 and L5 bilaterally. Free fat graft was placed over the dura and nerve roots. Bone graft was placed

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Fig. 1 Pre-operative MRI scan of lumbar spine showing the lesion involving L3 vertebra

posterolaterally from L2 to L5 (Fig. 2). An epidural infusion was used postoperatively for pain relief. Initial post operative neurological assessment was normal.

The following day deterioration in her lower limb neurological status was noted with no active movement or sensation in her lower limbs. That this was not recognised immediately was in part attributed to her epidural and to some difficulty with neurological assessment in a young child with systemic analgesia after major surgery. She was returned to theatre where a large haematoma was found and drained. Although subsequent sensory recovery was good, her motor

recovery was slow with grade 1 power in her lower limbs.

Two weeks later, once progressive neurological recovery was established, she underwent second stage anterior vertebratomy of L3 through a retroperitoneal approach. Unilateral L2, L3 and L4 segmental vessels were ligated to expose L3. A complete excision of the remaining L3 and posterior longitudinal ligament was performed in order to adequately decompress the dural sac anteriorly. Three fibular strut autografts from L2 to L4 were used to reconstruct the anterior column. Posterior iliac crest graft was used for further augmentation (Fig. 3).

Neurological recovery continued post-operatively and she had regained normal sensation and grade 5 motor power in both lower limbs by 6 months. Bladder sphincter dysfunction has persisted, requiring intermittent self catheterisation and anti-cholinergic medication. She was nocturnally continent. Bowel function remained normal throughout.

Radiographic fusion was evident at 6 months. Metal work was removed at 12 months and intra-operative biopsy showed no tumour recurrence (Fig. 4).

She remained under annual follow-up with MRI and plain radiographs. At 8 years she has no evidence of tumour recurrence. Her neurological status is unchanged. Imaging has shown excellent remodelling of the graft (Figs. 5, 6). She is skeletally matured with no lower limb malalignment and experienced no fibular donor graft site morbidity.

Fig. 2 AP and lateral X-rays following first stage of the procedure

Fig. 3 AP and lateral X-rays following second stage of the procedure showing the fibular strut grafts used

Discussion

Chordoma is a primary bone tumour arising from notochord remnants. It is a rare tumour accounting for only 1–4% of primary bone tumours [1]. Chordoma is usually a slow growing tumour, often with local recurrence and it may metastasise late in its course [1]. Typically it presents in late middle age and may occur anywhere along the spinal column. Chordoma is found most commonly in the clivus, sacrum and infrequently in the mobile vertebrae [2].

The optimum treatment for vertebral chordoma is wide surgical excision. Radiotherapy may provide short term benefit when adequate excision is not possible and in cases of local recurrence [3]. Surgical treatment has become more aggressive in recent years evolving from intralesional debulking to *en bloc* resection, as documented by Boriani et al., over a 45-year period in their treatment of 21 cases of chordoma located in the spine above the sacrum [4].

Chordomas are considered to be low grade tumours and metastases are infrequent at presentation [1, 5].

Fig. 4 AP and lateral X-rays following removal of metal work showing consolidation of the strut grafts

Fig. 5 Chronological X-rays showing graft incorporation

The prognostic value of staging is therefore diminished, compared to the value of staging in other bone tumours, because all these tumours are stage IA or IB according to the Musculoskeletal Tumor Society Staging System [6].

Mobile segment chordomas have a 50% survival rate at 5 years and a 28% survival rate at 10 years [4, 7]. Sacral chordomas on the other hand are reported as having 86% 5 year survival [3]. Treatment outcome is significantly influenced by the size and site of chordoma [1, 4]. Chordomas found in the vertebral bodies appear to be more aggressive than those arising in the clivus or the sacrum [3]. Metastases have been reported in 80% of the vertebral body chordomas, as

compared to a rate of 43% of all chordomas [3]. However, the survival rate appears to be affected more by local tumour progression than by metastases [1].

Conclusion

This case is unusual given the young age at presentation. She is the youngest patient reported in world literature. Despite the poorer published results of mobile segment chordomas, radical surgical excision and reconstruction has resulted in an 8 year disease free survival. Fibular strut graft reconstruction proved an excellent reconstructive option in the growing

Fig. 6 Eight year follow-up MRI Scan

skeleton with potential for remodelling. It is likely that the young age has contributed to the near complete neurological recovery.

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