Intraosseus schwannoma

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A 35-year-old lady presented with gradually increasing swelling over right arm for the last four years without a history of trauma. The initial painless swelling showed a sudden increase in size and warmth for the past five months. On clinical examination, there was a large, globular, smooth-surfaced, bony swelling measuring 25 x 15 cm over the anterolateral aspect of the upper right arm (Figure 1). The shoulder joint could not be examined optimally due to the sheer distortion of the joint by the mass. There was marked wasting of the biceps and triceps muscles. The tumour was deep to the deltoid muscle and had expanded the proximal humerus. There was no evidence of metastasis to the regional lymph nodes or liver.

The plain radiographs revealed a large, well-defined, lytic, expansile lesion with marginal sclerosis arising from the proximal humeral shaft (Figure 2). The matrix was clear. The cortex was intact with no periosteal reaction and no evidence of fracture. The possibilities of giant cell tumour of bone and aneurysmal bone cyst were considered. The patient underwent a needle biopsy. A diagnosis of schwannoma was made based on histology supported by S100 staining of the tumour cells. Subsequently, the patient underwent surgical excision of the tumour and fibular grafting. The tumour was found to be vascular.

Gross examination of the specimen revealed a mass measuring approximately 14 x 10 x 9 cm, arising from the medullary cavity of the proximal shaft of the right humerus. Sectioning revealed a firm, lobulated, white mass causing expansion and destruction of the metaphysis and a part of the diaphysis. Microscopy revealed cellular Antoni type A areas composed of interwoven fascicles of spindle-shaped cells with elongated wavy nuclei and indistinct cytoplasm (Figure 3). In some areas, nuclear palisading with the formation of Verocay bodies was also seen (Figure 4). At one-year follow-up, the patient did not have any evidence of recurrence, and her shoulder joint was fully functional.

Discussion

Intraosseous schwannoma is a rare primary bone tumour.1 The most common site of involvement is the mandible, with few other sites reported including humerus.2-5 Within a bone, the tumour can be located in the medulla, nutrient canal or can be extra-osseous. There are three mechanisms by which schwannomas can involve bone. The tumour can arise within the medullary cavity, within a nutrient canal or from the pe-
Riphepy with secondary bone erosion.\textsuperscript{5} When the tumour arises from the long bones, the most common location appears to be around the nutrient canal in the dia-metaphysis.\textsuperscript{5} Previously described radiological features include osteolysis with sclerotic borders, trabeculated contours, cortical erosions, and occasional central calcification.\textsuperscript{6} Pathological fractures are rare.\textsuperscript{1}

The differential diagnoses for this slow-growing, lytic expansile bony lesion were giant cell tumour and aneurysmal bone cyst. The possibility of intraosseous schwannoma was not considered due to its extreme rarity. Usually, the radiological findings are non-specific, and pre-operative diagnosis of this entity is difficult. The previously described intraosseous schwannomas of the humerus were smaller. In one case the size was 2.5 cm and another was a cortical defect.\textsuperscript{2,5} In the third case, the size of the minimally expansile tumour was not mentioned.\textsuperscript{4} The striking feature in our case is the unusually large size with significant bone expansion. The sacrum is a known site for giant intraosseous schwannoma, attributed to the large size of the sacral canal.\textsuperscript{7}

Microscopically, two types of cell arrangements, Antoni A and Antoni B, are found in schwannomas.\textsuperscript{8} In our case, predominantly Type A areas were reported, composed of closely packed, spindle cells arranged in bundles and cords. The nuclei of these cells are arranged in palisading rows, forming so-called Verocay bodies. Type B areas have also been described, which are composed of Schwann cells arranged in a haphazard fashion and separated by a loose myxoid stroma.\textsuperscript{8} All the intraosseous schwannomas reported have been benign.\textsuperscript{5,8}

Compared to the three cases of humeral intraosseous neurilemmomas previously reported in the literature, our case presented later (after four years). This simply could be due to the longer clinical course, which is a feature of giant intraosseous schwannoma reported in the sacrum.\textsuperscript{7} Surgery was the modality of treatment for all these cases. The large size of the tumour in our case makes it even more unique as a giant humeral intraosseous neurilemmoma.

\textbf{References}