We describe a schwannoma located in the mid-diaphyseal region of the fibula of a 14-year-old boy. Radiologically this was an expansile, lytic, globular and trabeculated lesion. MRI showed a narrow transition zone with a break in the cortex and adjacent tissue oedema. Differential diagnosis included schwannoma, fibrous dysplasia, giant cell tumour and aneurysmal bone cyst. The tumour was excised en bloc, with marginal resection limits, and there has been no recurrence two years after surgery. Histopathological examination confirmed the diagnosis of classic schwannoma. There were typical hypercellular Antoni A zones, less cellular Antoni B zones, and diffuse immunoreactivity to S100 protein. This is the first report of schwannoma involving a long bone in a child.

Schwannoma, also known as neurilemroma, is a benign tumour derived from Schwann cells of the nerve sheath.1 They commonly arise from cranial nerves (particularly the eighth nerve), spinal nerves, and peripheral nerves of the head and neck, with a predilection for sensory nerves.1 They are classified by type into classic (most common) and rarer variants, such as ancient, cellular, plexiform, epithelioid, glandular, pacinian, and melanotic.2 Schwannoma is primarily recognised as a neurogenic tumour although it may occur in other locations such as the skin, viscera and bone.1,3 Intra-osseous schwannoma is uncommon, there being only approximately 200 previously published cases, mostly involving the mandible, sacrum, maxilla and vertebrae.4-7 Occurrence in the long bones is particularly rare, but has been reported in adults.7-14 We describe a case of classic schwannoma involving a long bone in a child. Its distinction from other painful and radiologically benign lesions of long bones in children is also discussed.

Case report
A 14-year-old boy presented with a three-year history of pain and swelling of the right leg. The swelling was located in the posterior aspect of the middle third of the fibula and was gradually increasing in size and becoming more painful. There was no history of fever or significant night pain.

On physical examination an 8 cm × 5 cm firm, tender swelling was noted over the posterior aspect of the middle third of the right fibula. The overlying skin was intact, and there was no warmth, erythema or induration. The right knee and ankle joints had normal movement. There was no evidence of lymphadenopathy or neurovascular involvement.

Anteroposterior and lateral radiographs (Fig. 1) showed a well-defined, lytic expansile lesion with multiple trabeculations and cortical destruction involving the upper and middle thirds of the fibula. Axial CT sections showed cortical expansion and destruction and soft-tissue extension (Fig. 2). MRI showed the lesion to be homogeneous and hyperintense on...
T₂-weighted images and isointense on T₁-weighted images with surrounding soft-tissue oedema (Fig. 3). There were no fluid levels or signal abnormalities elsewhere.

A needle biopsy demonstrated a benign spindle-cell lesion suggestive of schwannoma or fibrous dysplasia and the tumour was subsequently excised en bloc with the middle third of the fibula, giving marginal resection limits.

The excised segment of bone measured 12.5 cm and was expanded in the middle by a tumour measuring 5.5 cm × 4.5 cm. The surgical clearance was 5 cm proximally and 2.5 cm distally. Sectioning of the bone revealed a lobulated lesion with a soft greyish-yellow cut surface. The intra-osseous origin was confirmed and there was extension into the soft tissues.

Detailed histopathological examination demonstrated the appearance of a classic schwannoma, comprising hypercellular (Antoni A²) areas and scattered looser and less cellular zones (Antoni B). The lesion was well circumscribed and lacked mitotic activity or cellular pleomorphism. Nuclear palisading was present (Fig. 4), as were degenerative changes and scattered haemosiderin-laden macrophages. Diffuse expression of S100 protein was observed by immunohistochemistry, confirming the diagnosis of schwannoma.
Discussion

There are no previously published reports of classic schwannoma in a skeletally immature long bone. Although there have been a few previous accounts of this tumour in the long bones of adults, only one report of schwannoma in a child exists, which was of a melanotic variety. This is histologically different from classic schwannoma and is associated with the Carney complex, an autosomally dominant disorder characterised by lentiginous facial pigmentation, cardiac myxoma and endocrine overactivity. The microscopic findings in our case were typical of classic schwannoma and were confirmed by a positive immunohistochemical reaction for S100 protein.

Intra-osseous schwannoma is an extremely uncommon tumour, accounting for less than 1% of all benign bone tumours. Its rarity has been attributed to the paucity of tumour, accounting for less than 1% of all benign bone tumours. A melanotic schwannoma in a child exists, which was of a melanotic variety. The neoplasm may be inseparable from a sensory nerve fibres in bone. The neoplasm may be intramedullary or located within the nutrient canal. When a long bone is involved the most common location appears to be around the nutrient canal in the metaphyseal-diaphyseal junction, as was seen in this patient.

The child's age, clinical presentation and radiological findings were suggestive of a benign primary bone lesion, such as a giant cell tumour or aneurysmal bone cyst. However, the absence of a fluid level and the presence of a cortical break with extension of the lesion into adjacent soft tissues was not in favour of these alternative diagnoses.

This case report demonstrates that schwannoma should be included in the differential diagnosis of a painful, radiologically benign lesion in a long bone. Other more common conditions that need to be considered in this clinical situation include non-ossifying fibroma, benign fibrous histiocytoma, desmoplastic fibroma and fibrous dysplasia. Microscopic examination is the key to differentiating these conditions. All of these lesions are composed of benign-appearing spindle cells. However, they lack Antoni A and B areas and can normally be distinguished from schwannoma without much difficulty by histological examination, provided the biopsy is of adequate size. In cases of doubt, immunochemical staining for S100 protein is extremely valuable, as schwannoma is diffusely positive for this marker.

True intra-osseous schwannomas must also be distinguished from soft-tissue schwannomas which secondarily invade adjacent bone. This sometimes difficult distinction can be made by careful radiological examination and pathological evaluation of the resected specimen.

Curettage and bone grafting is probably adequate treatment for bone schwannoma, as malignant change is extremely rare. However, large lesions may require en bloc excision, as was necessary in this case. Although recurrences have been reported in the early literature these have been attributed to incomplete excision 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