Recurrent congenital haemangiopericytoma in a child

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A five-day-old boy was referred with a soft-tissue mass in his right upper arm. Plain radiographs and ultrasound demonstrated a lesion extending from the axilla to the elbow on the posterolateral aspect of the humerus. Open biopsy confirmed the diagnosis of congenital haemangiopericytoma. After MRI and selective angiography, excision biopsy was carried out, but no adjuvant therapy was administered. At further examination, four years and ten months later, he was noted to have three small nodules at the site of the original tumour. Excision biopsy confirmed this to be a local recurrence, although the lesion was less cellular with no appreciable mitotic activity. Congenital haemangiopericytoma is a rare cause of a soft-tissue mass in children. Most tumours are benign, and recurrence is uncommon. The treatment is controversial, but most centres recommend the use of adjuvant chemotherapy, combined with complete excision. We recommend treatment with doxorubicin. Orthopaedic surgeons should be familiar with this tumour since 30% to 50% of cases occur in the limbs.

Congenital haemangiopericytoma is a rare tumour originally described as arising from the vascular pericytes of Zimmermann and differing from a glomus tumour or a haemangioma. We describe a case in a five-year-old boy in whom the tumour recurred.

Case report

A five-day-old boy was referred with a mass in his right upper arm. There had been no problems during the pregnancy or birth and he was otherwise well. Examination revealed a firm violet-coloured swelling extending from the axilla to the elbow on the lateral aspect of the arm (Fig. 1). The girth of the arm on the right was 17 cm, compared with 11 cm on the left. There was marked surface telangiectasia and focal crusting. The surface was easily indented. Visible subcutaneous veins were tender to palpation. The lesion was non-pulsatile and no bruits were heard on auscultation.

Radiographs of the lesion showed a soft-tissue mass of homogenous density around the posterolateral aspect of the humerus (Fig. 2). There was no bony erosion or periosteal reaction. The mass appeared to be slightly lobulated. Ultrasound showed a lesion 9 cm in length with an anteroposterior diameter of 5 to 6 cm and a transverse diameter of 8.5 cm. The echo pattern was diffusely mixed, with no clear demonstration of vascular loops, and no substantial vessels within or around the lesion. A thick-walled echogenic capsule was demonstrated around the lesion. Histological examination of biopsies showed a haemangiopericytoma (Figs 3 and 4). MRI showed that the mass had infiltrated the fascia surrounding the triceps muscle, but not the muscle fibres (Fig. 5); the neurovascular bundle was also unaffected. An angiogram using the right axillary artery (Fig. 6) showed the main arterial supply to be from the deep brachial artery, the supratrochlear artery and, particularly, from branches of the ulnar collateral artery. There was also evidence of a narrow band of extension into the axilla and of a possible associated retroscapular hypervascular tumour.

At excision biopsy, the tumour was excised with a margin of surrounding tissue. Care was taken to preserve the main neurovascular structures. The axillary component was excised in continuity with the main tumour. Frozen sections taken from the margins overlying the deltoid and axilla were clear of tumour. Pathological examination showed a lobulated, ulcerated, cellular tumour with a biphasic appearance. Most of the tumour was densely cellular, with proliferation of normal to oval cells, prominent vascularity, and easily identified mitotic figures. Other
lobules were less cellular and consisted of pale, myxoid, spindle-cell aggregates with obvious myofibroblastic differentiation, often associated with, or arising from, contiguous vessels. There was abundant haemosiderin, and extensive areas of necrosis and calcification compatible with infarction within this vascular tumour. No adjuvant chemotherapy or radiotherapy was given.

At further examination four years and ten months later, the patient was noted to have three nodules on the medial side of his mid-upper arm at the site of the original tumour. After further excision biopsy, pathological examination showed many similarities to the previous lesion, including intravascular cuffs of tumour cells. The recurrent lesion was, however, much less cellular with no appreciable mitotic activity, and was thought to reflect a maturation effect with time. It was felt that the lesions should be regarded as part of the same process, either representing clinical persistence or recurrence of the disease. Again the patient received no chemotherapy or radiotherapy.

At his most recent examination, his wounds had healed and there was no sign of recurrence. He remains under review.

Discussion

Haemangiopericytoma is a rare tumour with only 87 cases reported. It was originally described as arising from the vascular pericytes of Zimmermann and differing from a glomus tumour or a haemangioma. Stout and Murray did not differentiate between infantile and adult forms, but later studies make this distinction. Kauffman and Stout categorised congenital haemangiopericytoma as that appearing
in children younger than one year of age. About 10% of cases are of the infantile type and occur more often in boys. About 30% to 50% are found in the limbs with the remainder either in the head and neck or the trunk. There is seldom discolouration of the lesions in the limb in contrast to those in the head and neck. Congenital haemangiopericytoma tends to be located more superficially than that in adults.

The diagnosis is confirmed by biopsy. Plain radiography shows a soft-tissue mass, with or without calcific stippling. Ultrasound determines the size of the lesion, vascular loops, the echogenic content and whether or not a capsule is present, and angiography shows the vascular pattern and the presence of feeder vessels. Although they may not aid diagnosis, CT and MRI can provide information about the extent of the tumour, and help in preoperative planning.

The gross microscopic appearance of this lesion is usually transparent or white-grey. Feeder vessels may be prominent, and the infantile form tends to be more multilobular than in the adult. The pericyte may be round or oval in shape. The tumour tends to be densely cellular, with prominent vascular channels. Mitotic figures are usually easily identified. The tumour cells stain positively with vimentin.

The differential diagnosis of congenital haemangiopericytoma includes all tumours which present as a soft-tissue mass, including lipoma, haemangioma, and lymphangioma. Some tumours have similar histological features, such as infantile myofibromatosis, synovial sarcoma, fibrosarcoma, malignant fibrous histiocytoma, mesenchymal chondrosarcoma, and leiomyosarcoma. It is therefore important to examine multiple areas of the specimen to confirm the diagnosis.

The treatment of congenital haemangiopericytoma differs from that in adults, but there is no consensus as to the best method. Wide surgical excision is the most generally accepted treatment. Most centres use adjuvant therapy to reduce the risk of local recurrence, but the value is difficult to assess because of the rarity of these tumours. Chemotherapeutic regimens have included the use of vincristine, doxorubicin, actinomycin and cyclophosphamide. Jha et al reported success with radiotherapy when there was gross or microscopic evidence of tumour remaining after excision, and adjuvant radiotherapy was used with success by Borg and Benjamin. Other authors have reported that radiotherapy is ineffective. There is considerable concern regarding its use in the limbs of young children.

We decided not to use adjuvant therapy in this patient since the value of such treatment at the time of diagnosis had not been established. It was felt that excision had been curative since the margins of the frozen section were clear of tumour. Metastatic disease has only been reported in three cases of congenital haemangiopericytoma, although the adult form of the tumour has a rate of distant metastasis of up to 56%, mainly to the lung and skeleton. No histological criteria are available to predict which tumours are likely to metastasise. Careful follow-up is therefore required. When recurrence does occur, adjunctive chemotherapy has been used successfully. In our patient, because the recurrence of the tumour was so localised and histology showed that it was less cellular with no appreciable mitotic activity, it was decided not to administer chemotherapy.

As haemangiopericytomas are tumours of blood vessels, an orthopaedic surgeon is not always involved in the management. We were able to find only two cases reported in the orthopaedic literature. One was in an adult with a haemangiopericytoma of the knee, and the other a nine-month-old infant with a tumour above the knee. To our knowledge this is the first case of recurrent congenital haemangiopericytoma and also the first involving the upper limb. Orthopaedic surgeons should be aware of this condition when considering the differential diagnosis of a soft-tissue mass. Although there are no established criteria regarding the role of chemotherapy in the treatment of
congenital haemangiopericytoma, it is now our practice to treat these tumours with adjuvant chemotherapy, using doxorubicin. We have treated three cases in this manner subsequent to this case with no evidence of recurrence. There may also be a role for neoadjuvant chemotherapy to make very large lesions more manageable operatively.

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References