PRIMARY HAEMANGIOPERICYTOMA OF BONE

Report of Two Cases

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Haemangiopericytoma is a rare tumour of blood vessels arising from the contractile, elongated cells which surround capillaries, described by Zimmerman (1923) as pericytes. Haemangiopericytoma originating in bone is a most unusual occurrence. Marcial-Rojas (1960) has reviewed the literature on this subject.

Two tumours, one arising from a humerus and the second from a femur, are described in this paper. The natural history and management of haemangiopericytoma are discussed.

CASE REPORTS

Case 1—A woman aged forty-one presented in March 1972 with a one-month history of pain and swelling about the mid-portion of the right upper arm. Apart from slight aching in the back of the hand, together with some pain on shoulder abduction, she had not had any symptoms.

Examination revealed a tender, bony hard mass in the middle third of the right upper arm. The mass measured approximately seven centimetres by three. It was not pulsatile nor attached to the overlying skin. The skin itself felt warmer than on the other side but there were no dilated veins and no bruit was heard over the mass. Elbow function was unimpaired but she complained of pain on active abduction of the shoulder. There were no enlarged lymph glands in the axilla, and there was no other evidence of a primary tumour or of secondary deposits elsewhere. The following tests showed normal values: haemoglobin, white blood count and differential count, erythrocyte sedimentation rate, acid and alkaline phosphatases, serum proteins, serum calcium and phosphorus, electrolytes and blood urea nitrogen, T3 (serum level of 3, 5, 3 tri-iodothyronine) and T4 (serum level of thyroxine) levels, and stools and urine for occult blood. The cytology of the sputum was normal. Radiographs of the right humerus showed a destructive intraosseous lesion (Fig. 1). Otherwise the skeletal and chest radiographs were normal. Intravenous pyelography showed a horseshoe kidney.

The diagnosis was thought to be between secondary neoplastic deposit, chronic infection and reticulum-cell sarcoma. An atypical Ewing's tumour was also considered.

Biopsy was undertaken through an antero-lateral incision. The biceps brachii was retracted medially and was noted to be normal, as was the brachialis, which was split down to the humerus in the midline. Retraction of the brachialis medially and laterally revealed a pinkish-grey, grape-like tumour bulging from a large cortical defect in the middle third of the antero-medial humerus. The tumour was lobulated and very friable, although firm in consistency. It did not bleed unduly. Extension of the tumour was noted in the medullary cavity proximally and distally. A large portion of tumour was resected together with normal bone and a plate applied because much of the humeral cortex had been removed. The wound was closed in the normal way.

Representative sections of the decalcified tumour are shown in Figures 2 and 3 and indicate its vascular and, in particular, its pericytic origin. This was confirmed by Dr D. C. Dahlin of the Mayo Clinic, who thought the tumour was malignant.

Amputation was decided upon, and the right shoulder was disarticulated. The patient was discharged after an uneventful recovery and was well a year later.
Case I—Antero-posterior and lateral radiographs showing a lesion in the humerus destroying the cortex. No bony and very little periosteal reaction is seen.

Case I. Figure 2—A section showing prominent blood vessels surrounded by collars of connective tissue and mantles of pericytes which have large round or oval vesicular nuclei. (Haematoxylin and eosin, ×500.) Figure 3—A section of the tumour stained with haematoxylin and eosin. (×200.) A similar section (which is not shown) stained to show reticulin demonstrated that the tumour cells lie outside the reticulin sheaths of the blood vessels.
Case 2—A woman aged thirty-seven presented in June 1964 with a three months' history of pain in the inner and lower left thigh which had started after jumping from a height. Apart from a limp she had no other symptoms.

Examination showed a tender, bony hard mass on the anterior aspect of the lower end of the left femur. There were no other abnormalities. A radiograph of the left femur showed a lytic lesion approximately 6 centimetres in diameter in the postero-medial aspect of the distal femur with a thin cortex expanded over it (Fig. 4).

The following tests gave normal results: haemoglobin, white blood count and differential count, urinalysis, electrolytes, acid and alkaline phosphatases and serum calcium and phosphorus levels. The erythrocyte sedimentation rate was 44 millimetres in one hour. Radiographs of the chest, lumbar spine, and pelvis and hips were normal. No further investigations were done.

Biopsy was done. A medial approach through the vastus medialis revealed the thin cortex overlying the tumour which was opened with an osteotome. The tumour, a white mass the colour and consistency of fish flesh, extended into the medullary cavity proximally and distally. The histology was essentially the same as that of the first case, with whorls of blood vessels surrounded by swollen pericytes.

Four weeks after operation cobalt irradiation was given over thirty-three days with a dosage of 6,250 r. Three months after biopsy, the chest radiograph being normal, a high femoral amputation was done. The patient remained well thereafter and was last seen in August 1972, eight years after the original operation.

DISCUSSION

Haemangiopericytoma arising in soft tissues have been reported with some frequency. A comprehensive review of the literature was made by Backwinkel and Diddams (1970). Marcial-Rojas (1960) reported a case of primary clavicular haemangiopericytoma; pulmonary metastases preceded death seventeen months after clavicular resection. Pathological and clinical accounts of both the benign and malignant forms of soft-tissue tumours were given by Stout 1953.

The glomus tumour is the best known form of haemangiopericytoma. It differs from the latter by having a typical organoid pattern interspersed with large numbers of non-myelinated nerve fibres; also the glomus tumour behaves as a typically benign tumour although it may recur locally after excision.

The behaviour of haemangiopericytomata seems unrelated to their histological structure, and makes management controversial. The tumour is highly malignant but not necessarily so over a short period of time. Growth may be slow or extremely rapid, with death in a matter of months. The tumour may arise from early childhood to old age and may be painless. Backwinkel and Diddams (1970) in a review of 224 cases of soft-tissue tumours found a recurrence rate of 52.2 per cent after removal. They concluded that classification into benign...
and malignant forms was of no prognostic value. This view was shared by Marcial-Rojas (1960) in regard to the primary tumour.

Treatment of primary haemangiopericytoma has been by surgical removal, either local or radical, by radiotherapy, or by a combination of both. Ortega, Finklestein, Isaacs, Hittle and Hastings (1971) described the control of both primary tumours and osseous metastases using a combination of radiotherapy and chemotherapy; actinomycin D, cyclophosphamide and vinchristine appeared effective. Methotrexate has also been effective in the control of skeletal metastases.

The extent of the surgical excision lacks agreement. Spiro et al. (1964) advocated a very radical excision; Backwinkel and Diddams (1970) recommended wide surgical excision, and suggested that cases treated by surgery alone showed a cure rate of 53.1 per cent while radiotherapy alone gave a 13.3 per cent cure.

This discussion is based largely on the experience of soft-tissue haemangiopericytomata and until more evidence is available, it must be assumed that when the tumour occurs in bone it will behave and respond to treatment in the same way. It was for this reason that amputations were done on the patients described.

SUMMARY

1. Two cases of primary skeletal haemangiopericytoma treated by ablative surgery, one in combination with radiotherapy, are reported.
2. A discussion of the management of these tumours includes surgical excision, radiotherapy, a combination of both or a combination of radiotherapy with chemotherapeutic agents.
3. Because of the unpredictable behaviour of these tumours it seems necessary to include ablative surgery if cure is to be expected.

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REFERENCES


