Infantile haemangiopericytoma

D M D E A Gunawardene[1], M V C de Silva[2], B A D Jayawardene[3]

Sri Lanka Journal of Child Health, 2001; **30**: 44-5

(Key words: Infantile haemangiopericytoma)

Introduction

Haemangiopericytoma is an uncommon tumour arising from pericytes of blood vessels. It usually affects adults with a mean age of 45 years[1]. Occurrence of haemangiopericytoma in infants is rare1. Infantile haemangiopericytomas have a different histological picture and clinical behavior compared to the adult type. Unlike the adult type, mitotic activity and presence of focal necrosis do not indicate poor prognosis[1]. Occurrence of spontaneous regression has been reported[2]. These tumours usually follow a benign course and are cured by local excision. They should be distinguished from malignant tumours such as synovial sarcoma, mesenchymal chondrosarcoma and infantile fibrosarcoma which may contain focal areas resembling haemangiopericytoma. We report the occurrence of an infantile haemangiopericytoma in a 4 month old infant.

Case report

A 4 month infant presented with the history of a gradually enlarging swelling in the left parotid area of the face, which has been present since the age of one week. On examination, there was a cystic lump measuring 6 cm in diameter, in the left cheek, CT scan excluded involvement of the underlying mandible. The tumour was excised completely following fine needle aspiration (FNAB) and incisional biopsies. The patient was referred to the Cancer Institute Maharagama for further treatment but was subsequently lost for follow-up.

Pathology

FNAB revealed sheets of round and spindle shaped cells with scanty cytoplasm. It was reported as a low grade malignant spindle cell tumour. Histology showed sheets of cells with round and oval nuclei and moderate amounts of ill defined cytoplasm arranged around variable sized thin walled vascular spaces (Figure 1).

[1]*Demonstrator in Pathology,* [2]*Senior Lecturer in Pathology, Faculty of Medicine, Colombo,* [3]*Consultant Paediatric Surgeon, Lady Ridgeway Hospital, Colombo.*

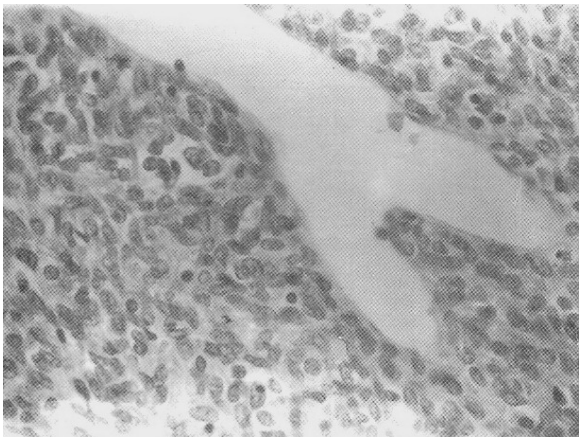

Figure 1 *Sheets of cells arranged around vascular spaces.(Haematoxylin and Eosin x 400)*

Some branching vessels were of the characteristic staghorn type. Focal areas contained spindle shaped cells. Some areas were composed of solid sheets of cells without intervening blood vessels. Reticulin preparation showed a dense reticulin meshwork surrounding individual tumour cells and vessels (Figure 2). Foci of necrosis and haemorrhage were present. Mitotic count was six per ten high power field.

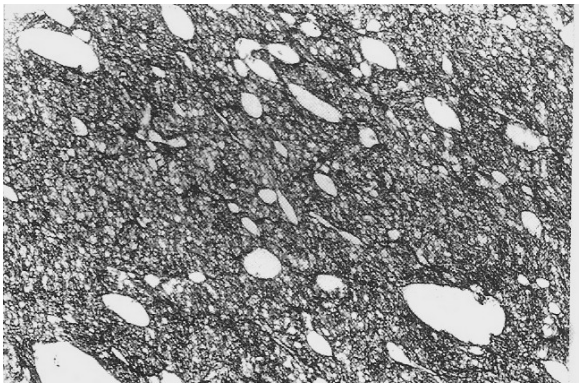

Figure 2 *Dense reticulin meshwork surrounding tumour cells and vessels (Reticulin stain x 100)*

Discussion

The histological features were compatible with those of infantile haemangiopericytoma. These tumours are multilobulated often with distinct intravascular and perivascular satellite nodules outside the main tumour. The differential diagnosis of infantile haemangiopericytoma includes synovial sarcoma, mesenchymal chondrosarcoma and infantile fibrosarcoma. Infantile haemangiopericytoma lacks the focal biphasic or glandular pattern, positive staining for cytokeratin, hyalinisation and calcification shown by synovial sarcoma[1]. The range of caliber of vessels seen in haemangiopericytoma is more variable. Presence of islets of well differentiated cartilage or less frequently bone favours the diagnosis of mesenchymal chondrosarcoma. The spindle cell areas may be confused with aggressive infantile fibromatosis and infantile fibrosarcoma. Unlike in these tumours the spindle cells in this child's tumour were not arranged in distinct bundles or fascicles. Although infantile fibrosarcomas may contain focal vascular areas resembling haemangiopericytoma, they usually do not contain regularly distributed staghorn type vessels1. Features which often leads to a misdiagnosis of malignancy include arrangement of cells in solid sheets without the characteristic vascular pattern and the presence of mitoses and necrosis. Extensive sampling will help to identify areas of typical haemangiopercytoma.

Most infantile haemangiopericytoma follow a benign course. Rarely these tumours behave aggressively with local infiltration, recurrences and even distant spread3.

References

1. Enzinger F M, Weiss S W. Soft tissue tumours. 3rd ed. St. Louis; Mosby, 1995.

2. Chen K J, Kassel S H, Medrano V A, Congenital Haemangiopericytoma. *J Surg Oncol* 1986; **31**: 127.

3. Atkinson J B, Mahour G H, Issacs H, Ortega J A, Haemangiopericytoma in infants and children: a report of six patients. *Am J Surg* 1984; **148:** 372