INFANTILE HEMANGIOPERICYTOMA (A Case Report)

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Introduction

Hemangiopericytoma is an uncommon tumour of pericyte origin, mostly in adult age group. Hemangiopericytomas which occur in children have a similar histological picture, but a more benign clinical behaviour [1]. A case of infantile hemangiopericytoma is reported to illustrate the clinical and pathologic features of this uncommon tumour.

CASE REPORT

Four-year-old male child was noticed to have swelling of left forearm at birth, which was gradually progressive. He was born at term after an uneventful pregnancy. Mass was biopsied elsewhere two months prior to reporting to this hospital. Histology was reported as infantile hemangiopericytoma. On evaluation here, he was found to be having a diffuse, bosselated, non-compressible, soft to firm mass, involving entire length of left forearm. Skin over the swelling was free and contained a 4 cm long biopsy scar over the ulnar aspect. Elbow, wrist and radio-ulnar movements and peripheral pulsations were normal. There was no regional lymphadenopathy. Radiograph of left forearm revealed soft tissue mass over medial aspect of forearm with normal underlying bones.

At surgery, the tumour was found to be partially circumscribed, infiltrating the flexor compartment muscles and was excised down to periosteum. Cut surface showed cystic, multiloculated lesion filled with hemorrhagic material. Microscopically it showed a tumour comprising of many vascular channels with plump polygonal to elongated cells, as sheets in the walls and as solid groups (Figs 1 and 2). These had a fair amount of cytoplasm, big vesicular nuclei and infrequent mitosis. Tumour extended in between the skeletal muscle fibres (Fig 3).

Child has now been followed up for 30 months and there is no local recurrence or systemic metastasis.

Fig. 1 : Sheets of round cells surrounding branching blood vessels (H&E x 100).

Fig. 2 : Staghorn pattern of blood vessels surrounded by tumour cells (Reticulin x 200).

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Fig. 3 : Tumour cells infiltrating skeletal muscle fibres (H&E x 100).

Discussion

Hemangiopericytoma is a rather uncommon neoplasm that was first described and named by Stout and Murray in 1942 [2]. In their original description they postulated that the tumour is composed mainly of pericytes, a specific cell type first identified by Zimmerman in 1923. In subsequent years, the clinical and morphological characteristics of hemangiopericytomas have been more clearly defined, and its pericytic origin has been confirmed ultrastructurally. Hemangiopericytoma is primarily a tumour of adult life. It is rare in children constituting only 3 percent of all soft tissue sarcomas in that age group [1]. Hemangiopericytomas which occur in children have similar histological picture but a benign clinical behaviour compared to their adult counterparts.

Infantile hemangiopericytomas occur in infants during first year of life and like juvenile hemangiomas, are located almost exclusively in the subcutis. Characteristically, they are multilobulated, often with distinct intravascular and perivascular satellite nodules outside the main tumour mass. Majority of the lesions are solitary [3] but multiple lesions have been described [4,5]. Tumours are most common in the lower extremity especially the thigh, the pelvic fossa and the retroperitoneum. It affects, less frequently,

the trunk and the upper extremity. Most are deep seated and the majority are found in muscle tissue. Lesion occurring in the tongue and sublingual region discovered at birth have been described [6].

Microscopically, the tumour consists of tightly packed cells around thin walled, endothelium-lined vascular channels ranging from capillary-sized vessels to large gaping sinusoidal spaces. They closely resemble the adult type except for frequent endovascular proliferation of tumour cells, increased mitotic activity and focal necrosis, features that indicate malignancy in an adult-type hemangiopericytoma. This, however, is generally not the case with the infantile form. Most of these tumours tend to follow a benign clinical course and are curable by local excision. In rare instances, however, there may be local infiltrative growth or recurrence and even metastasis [7].

The lesion reported here involved the forearm, a site less commonly affected and despite being large and infiltrating the muscles, it exhibited a benign clinical behaviour.

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