

Case Report

Infantile hemangiopericytoma with recurrence: a case report

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Received: 23 November 2015

Accepted: 17 December 2015

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ABSTRACT

Infantile hemangiopericytoma is rare vascular tumour in infant. These tumour usually solid, occur in the subcutaneous tissues of head and neck. Mainly it has a benign behaviour but recurrence and metastasis also seen. Herein, we report on one full term female child, delivered with a nodular swelling present behind the right ear. Complete surgical excision with safety margins was performed for the lesion. Histological examination of the lesion showed picture of infantile hemangiopericytoma. There is evidence of local recurrence present, nodular swelling reoccur at excision site and gradually increase in size and become 15×13×11 cm size. After twenty days child has died.

Keywords: Infantile, Hemangiopericytoma, Vascular, Recurrence

INTRODUCTION

In 1942 Stout and Murray described the hemangiopericytoma. Hemangiopericytoma is a soft tissue tumour derived from mesenchymal cells with pericytic differentiation.^{1,2} Therefore, hemangiopericytoma have a wide distribution in both soft tissue and skeletal system. Disease can either be benign or malignant. Two types have been described; (1) infantile hemangiopericytoma and; (2) adult hemangiopericytoma. Infantile hemangiopericytoma (also known as “congenital hemangiopericytoma”) is a cutaneous condition characterized by single or multiple dermal and subcutaneous nodules that may be alarmingly large at birth or grow rapidly. Infantile hemangiopericytoma represent about one third of all pediatric hemangiopericytoma.³ It arises from Ziemmermann pericytes.

Diagnosis of infantile hemangiopericytoma must be made by histology and surgical resection is the most common method of treatment. Microscopic characteristics are the so called pericytoma pattern with tightly packed cells

around ramifying thin walled and endothelial lined vascular channels ranging from small capillary sized vessels to large sinusoidal spaces.²

These solid, vascular neoplasms typically occur in the subcutaneous tissues of head and neck and follow a benign course. Clinical features and behaviour are particular, with subcutis and oral cavity locations; multifocal and metastatic forms are reported, as well as good response to chemotherapy and occasional spontaneous regressions. Prognosis is usually favourable.⁴

CASE REPORT

Two months old female child present in paediatrics outdoor with complain of nodular swelling about 2-3 cm in size present behind the right ear. It was present since birth about one cm in size, size gradually increased. Outer surface was nodular with a small ulcerated area measured 0.3×0.5 cm. Growth was variable in consistency, warm, non-tender, nonreducible and non-fluctuant. Growth was immovable, fixed to the overlying

skin and under lying structures. Growth resected and sent for histopathological examination. After fifteen days of resection, there was nodular swelling measuring 12×10×9cm, reoccur at resected site. Patient also complains of fever, nausea, vomiting and diarrhoea. Needle biopsy was taken and sent for histopathological examination. It was diagnosed as same of previous diagnosis infantile hemangiopericytoma. Child was referred to further higher centre. After twenty days of resection, swelling has become 15×13×11 cm in size and patient had died without treatment.

Pathologic findings

Gross examination shows a grey white soft tissue mass measuring 3×2.5×2.5 cm with skin attached one side. Skin flap measuring 1.5×0.5 cm with ulcerated area in centre of skin flap measured 0.3×0.5cm. Margin of ulcerated area was flat and base was haemorrhagic. Outer surface of mass was nodular and irregular. Cut surface was grey brown with haemorrhagic areas. Tissue was taken, processed and stained by H&E and PAS.

Microscopic examination

Hematoxylin and Eosin stained sections showed plump spindle shaped cells with prominent vascular pattern consisting of thin walled vessels lined by single layer of flattened endothelial cells with dilated vascular spaces, at places forming staghorn configuration (Figure 1). Moderately pleomorphic spindle cells have indistinct cell boundary with moderate amount of eosinophilic cytoplasm and oval to spindle shape nuclei. At places myxoid stroma was also present. Mitotic activity 1- 2 per 10 HPF was noted. Necrosis was not present. These tumour cells also extend up to epidermis (Figure 3).

Periodic acid Schiff (PAS) stain showed negative staining for tumour cells. Only connective tissue basement membrane showed positivity (Figure 4).

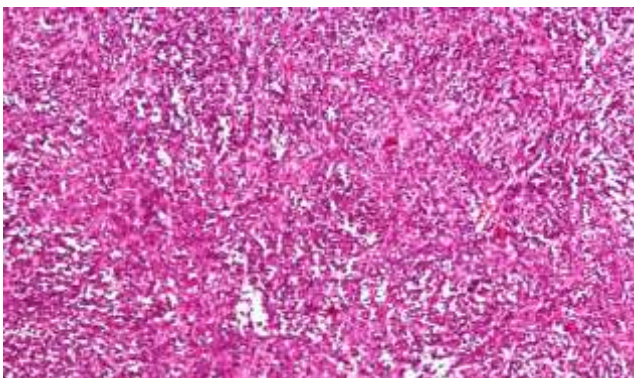


Figure 1: Hematoxylin and eosin stained section showed prominent vascular pattern consisting of thin walled branching vessel (staghorn configuration), (H and E 10x).

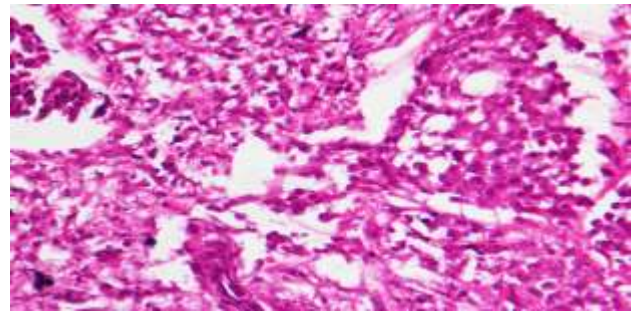


Figure 2: Hematoxylin and Eosin stained section showed plump spindle and oval to round cells with prominent vascular pattern consisting of thin walled branching (H and E 40X).

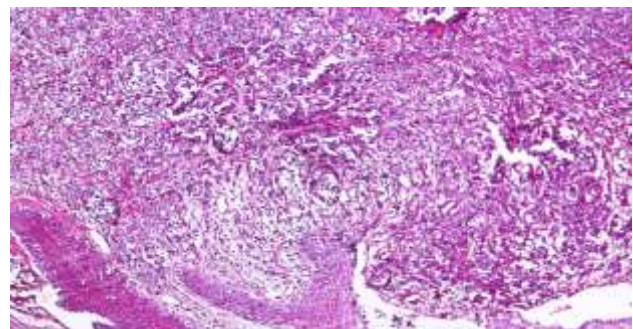


Figure 3: Hematoxylin and Eosin stained section showed tumour cells extend up to epidermis (H and E 10X).

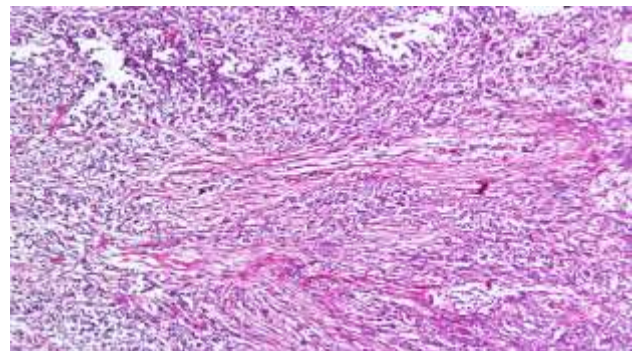


Figure 4: Periodic acid schif (PAS) stained section showed negative staining for tumour cells and positive staining for connective tissue (PAS 10X).

Needle biopsy from mass at resection site

Grossly two grey white threads like specimen measuring 1.5 cm long with attached skin at one end. Microscopically section showed same feature of previous excisional biopsy without any feature of malignancy.

DISCUSSION

Infantile hemangiopericytoma is a rare vascular tumour in children below one year of age.⁵ Hemangiopericytoma arises from the vascular pericytes of Zimmermann which

are contractile cells that spiral around capillary walls and differing from a glomus tumour or a hemangioma but they did not differentiate between infantile and adult forms. The infantile type occur in 10% of cases and in boys more than in girls.⁶⁻⁸ Approximately 30% to 50% hemangiopericytomas are found in the limbs with the remainder either in the head and neck or the trunk. There is seldom discoloration of the lesions in the limb in contrast to those in the head and neck.⁶ Infantile hemangiopericytoma tends to be located more superficially than that in adults.⁹

In our case, mass was present since birth in female child. Mass was lying subcutaneously and behind right ear. It was nodular, immobile and fixed to the skin.

The diagnosis is confirmed by excisional biopsy.⁹ The gross appearance of this lesion is usually transparent or white-grey.⁶ Feeder vessels may be prominent, and the infantile form tends to be more multilobular than in the adult. Microscopically; the pericytes 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.^{6,9}

In our case the final diagnosis was achieved by excisional biopsy. The histological features were consisting with infantile hemangiopericytoma. The morphological criteria suggested by Enzinger and Smith were used to distinguish a benign from a malignant tumour. The criteria were: large size (>5 cms), increased mitotic rate (4 or more per 10 HPF), high degree of cellularity, immature and pleomorphic tumour cells, foci of hemorrhage and necrosis.¹⁰ In accordance with Enzinger and Smith's criteria, our case was histologically benign. There was no evidence of malignancy at the time of examination. After recurrence, mitotic figures were two per ten HPF and there was no necrosis however tumour size was more.

The clinical differential diagnosis of congenital hemangiopericytoma includes all tumours which present as a soft tissue mass, including lipoma, hemangioma, and lymphangioma. Histological differential diagnosis includes infantile myofibromatosis, synovial sarcoma, fibrosarcoma, malignant fibrous histiocytoma, mesenchymal chondrosarcoma, and leiomyosarcoma because of the shared hemangiopericytoma-like vascularity of all these tumours.¹¹⁻¹⁴ It is therefore important to examine multiple areas of the specimen to confirm the diagnosis.⁹

The management of infantile hemangiopericytoma differs from that in adults, but there is no consensus as to the best method.⁹ Wide surgical excision with safety margin is the most accepted method of treatment.^{6,15,16} This method was used in management of our case. Chemotherapeutic regimens have included the use of vincristine, doxorubicin, actinomycin and

cyclophosphamide. Success with radiotherapy when there was gross or microscopic evidence of tumour remaining after excision, and adjuvant radiotherapy was used with success.^{17,18}

Metastatic disease has only been reported in three cases of congenital hemangiopericytoma although the adult form of the tumour has a rate of distant metastasis of up to 56%, mainly to the lung and skeleton.^{5,8,16,19,20} Careful follow up is therefore required. When recurrence does occur, adjunctive chemotherapy has been used successfully.¹⁶ Our case was free from metastasis at the time of diagnosis.

Hemangiopericytoma is an innocent-looking tumour, it has a high recurrence rate; Stout found it to be 28% and Backwinkel 52.2%.^{21,22} In our case, recurrence was occur at resection site. After fifteen days of resection which was nodular, rapidly growing and measurement about 15×13×11 cm. Patient had died without treatment.

CONCLUSION

Infantile hemangiopericytoma is rare neoplasm which usually behaves benign course. But local infiltration, recurrences and even distant metastases may occur. Therefore, wide local excision and careful follow up is therefore essential.

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: Not required

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Cite this article as: Jaipal D, Kulhari S, Gupta M, Gupta N. Infantile hemangiopericytoma with recurrence: a case report. *Int J Contemp Pediatr* 2016;3:277-80.