

Congenital Haemangiopericytoma of the Chest Wall

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Abstract : Hemangiopericytoma is a rare vascular tumor that usually occurs in adults. The occurrence of these tumors in infants, known as congenital or infantile hemangiopericytoma, is even rarer and their behavior may be more benign than the adult type. We describe a 10 days old male neonate with congenital hemangiopericytoma, presenting with a mass on the right anterior chest wall at birth. The anterior chest wall location is an unusual site for a congenital hemangiopericytoma. Tumor excision was performed and morphologic and immunohistochemical results confirmed the diagnosis of infantile hemangiopericytoma. There was no tumor recurrence during 11 months of follow-up.

Key Words: Congenital haemangiopericytoma, chest wall, CD 34, immunohistochemistry

Introduction

Hemangiopericytoma is a rare vascular tumor which was first recognized as pathological entity in 1942 by Murray and Stout.¹ It derives from pericapillary cells or Zimmerman's pericytes, the cells located around vessels and capillaries, which presumably provide vascular stability and regulate luminal size. Most cases of hemangiopericytoma are benign, but they have a definitive malignant counterpart.² This rare tumor most commonly occurs in subcutaneous tissue and lower limb muscles.³ Hemangiopericytoma is very uncommon in childhood and comprises two different clinical entities, the adult type and infantile type, occurring in first year of life.⁴ Fewer than 10% of cases are seen in children, and congenital tumors are found in only 3% of cases. Recently it was reported that hemangiopericytoma in children older than 1 year behave similarly to those seen in adults where cases involving the infantile variant have a more benign clinical course.⁵

Case report

A 10 days old newborn baby boy with a birth weight appropriate to gestational age presented with a well defined, subcutaneous, non-tender, firm mass on the right side of chest wall measuring 6x5cm. After routine investigations, complete surgical excision was performed after 2 days. The postoperative course was uneventful, and the patient was discharged from the hospital after 4 days. On gross examination the mass was well circumscribed, soft to firm in consistency and

measuring 6x5x5 cm. The cut surface was grey brown without the areas of hemorrhage and necrosis.

Microscopic examination revealed a vascular neoplasm composed of oval to spindle shaped cells with pale staining nuclei which are arranged around vascular channels exhibiting a characteristic well-developed "staghorn" branching pattern (Fig:1A). Reticulin staining demonstrated vessels lined by a single layer of endothelial cells and enclosure of single tumor cells. (Fig: 1B). Mesenchymal tumor cells were stained positive for CD34 (Fig: 1 C&D). In the neoplasm, the mitotic rate was 1 per 10 high power fields. No necrosis was observed in the tumor. The tumor was pathologically diagnosed as a hemangiopericytoma. Regular follow up examinations after discharge showed no recurrence at the site of resection and no remote metastasis up to 11 months of age.

Discussion

Hemangiopericytoma (HPC) is a rare soft-tissue neoplasm. It can occur anywhere vascular capillaries are found. The tumor most commonly occurs in the musculature of extremities, pelvis (uterus, ovary and urinary bladder), head and neck and lungs⁶. Most commonly it is seen in adults; only 5-10% of cases occur in children.⁷ In children HPC comprises two different clinical entities. Forty percent cases occur during first year of life and are considered to be congenital or infantile HPC and are less aggressive.

Those occurring in children older than one year behave in a manner similar to adult HPC and are more

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aggressive.^{5, 8}The etiology of HPC remains unknown.⁸

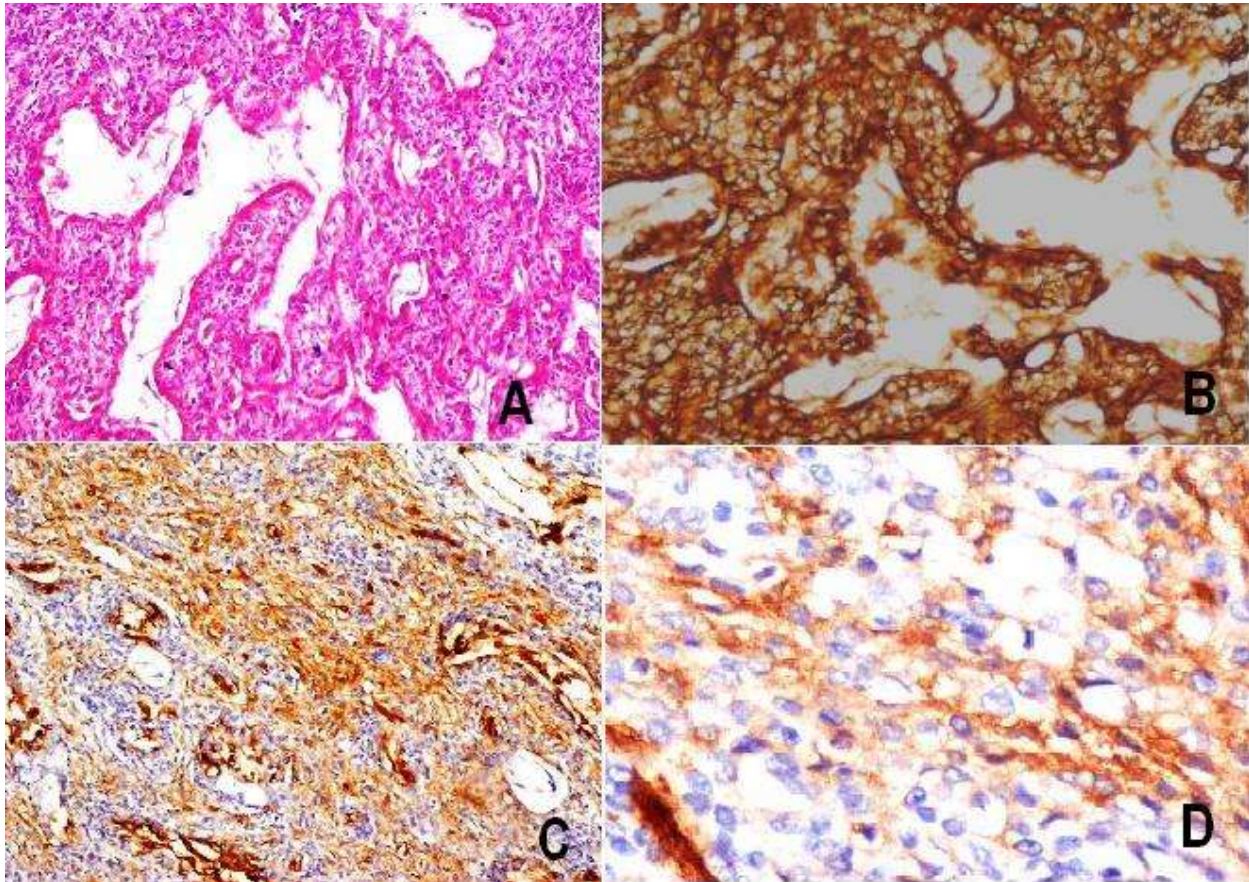


Fig:1- Microscopic features of the congenital haemangiopericytoma A, Spindle shaped pericytes surrounding the endothelial-lined branching vascular spaces (H&E x 200). B, Stag-horn vessels lined by a single layer of endothelial cells and enclosure of single tumor cells. (Reticulin stain, x 400). C, CD34 positive tumor cells. (CD34 stain x 200). D, tumor containing CD 34 positive neoplastic cells (CD34 stain x400).

We diagnosed a hemangiopericytoma following an examination of the structural features of the mass. Spindle-shaped cells surrounding the endothelial-lined vascular spaces were observed by hematoxylin and eosin staining and immunohistochemically, these were positive for CD 34.

The consideration of congenital HPC in the newborn infant with a vascular mass should always be advocated.⁹ Congenital HPC is characterized by a better clinical behavior than the adult type, which requires an aggressive multimodality therapy. Chemoresponsiveness and spontaneous regression have been reported in children younger than 1 year, suggesting that a more conservative surgical approach should be used.¹⁰ Congenital hemangiopericytoma is unresponsive to steroid therapy, unlike other vascular malformations; resection is the treatment of choice. Long-term postop-

erative follow-up is essential for the early detection of metastases⁹

Conclusion

This report presented a rare case of a congenital hemangiopericytoma on the chest wall. Surgical resection is the mainstay of treatment. Infantile HPC is characterized by a better clinical behavior than the adult type.

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