Malignant hemangiopericytoma of the retroperitoneum: A Case Report And Review Of Literature

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Citation

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Abstract

Hemangiopericytomas, forming 1% of vascular tumors, are soft-tissue tumors arising from pericytes. These tumors arise in skin, soft tissues, extremity muscles, retroperitoneum, sinonasal tract etc. A rare case of haemangiopericytoma of the retroperitoneum is presented in this article where complete resection could be achieved and high vascularity of the tumor was observed during resection.

INTRODUCTION

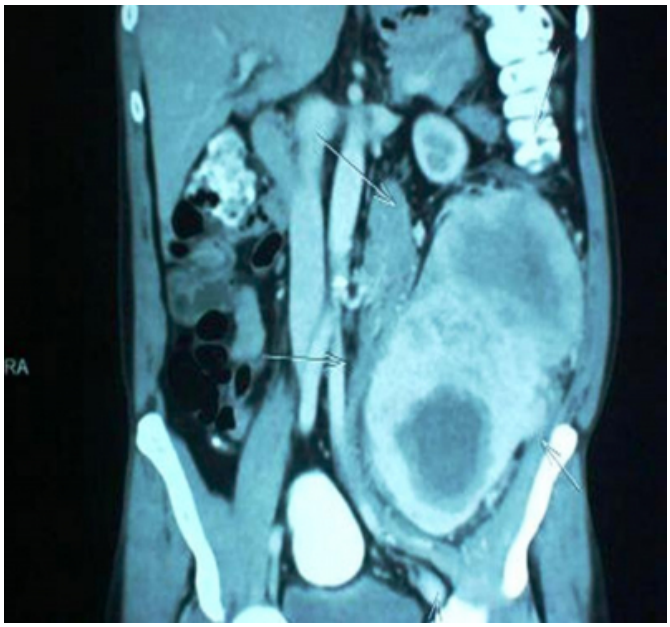
Hemangiopericytoma was initially described by Stout and Murray in 1942 as a soft-tissue tumor derived from pericytes of Zimmerman, which surround the capillaries. [1] It is a rare mesenchymal tumor accounting for 1 % of vascular tumors. [2] This rare tumor of adult life (fifth decade) is uncommon in children. Herein, a case of retroperitoneal hemangiopericytoma is being presented for its rarity and high vascularity. When the tumor in the retroperitoneum is large, more vascularity is expected. But in this case the neovascularisation was so extensive that it resulted in massive blood loss during resection and the tumor was found to invade the underlying ilium histologically

CASE REPORT

A 58-year-old man presented with pain radiating to the left lower limb and with an abdominal lump of three months duration. Clinical examination revealed a large retroperitoneal lump extending to the pelvis. Computerized tomography (CT scan) of the abdomen revealed a heterodense mass in the retroperitoneum, posterior to the left psoas major, extending to the pelvis and displacing the left iliac vessels (Fig. 1).

Figure 1

Figure 1: CECT of the abdomen, coronal section: Heterodense mass displacing iliac vessels



The left kidney was free of tumor radiologically.But the left ureter could not be traced in its entirety. Metastatic work-up like X-ray of the chest and CT scan of the chest were within normal limits. On laparotomy, the tumor was found located in the left lumbar region behind the left psoas major and extending beneath the iliacus. Left ureter and iliac vessels were isolated and resection of the tumor was done with resection of left psoas major, part of iliac bone and perinephric fat as clearance. There was a lot of neovascularisation which leaded to significant blood loss during surgery. An interesting thing noted were large veins

draining the tumor in the posterior aspect and through the iliacus muscle. These opened up during sharp dissection. The bleeding was from both the patient and tumor side which was difficult to control. Hence, the tumor was resected and removed rapidly so that bleeding could be controlled with packing. The patient had an uneventful postoperative recovery. Postoperative histopathology revealed a 17x15x10cm tumor with features of hemangiopericytoma invading the bone (Fig. 2). Immunohistochemistry for CD34 and Vimentin were positive (Fig. 3&4).

Figure 2

Figure 2: Spindle cells admixed with vascular spaces

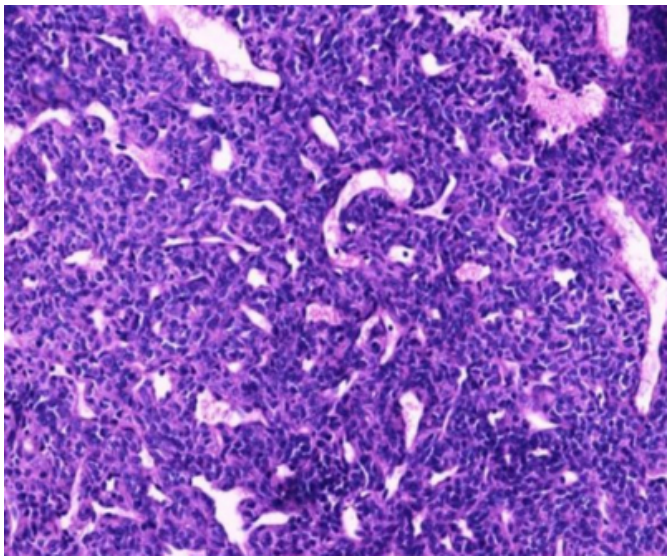


Figure 3

Figure 3: IHC for vimentin - tumor cells strongly positive

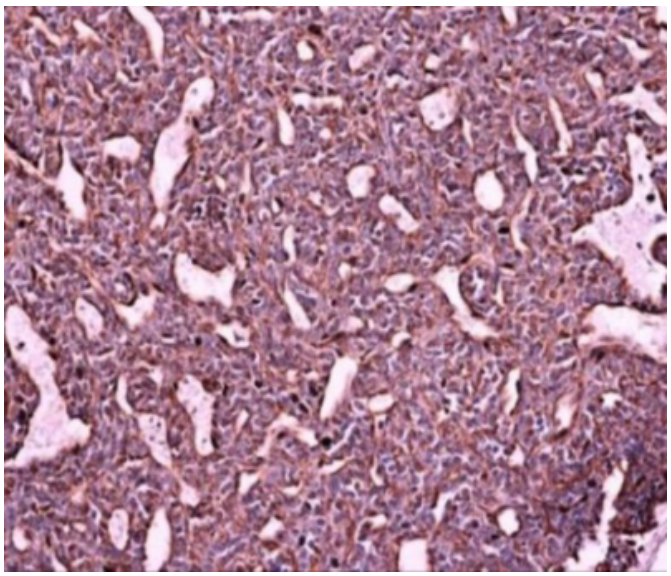
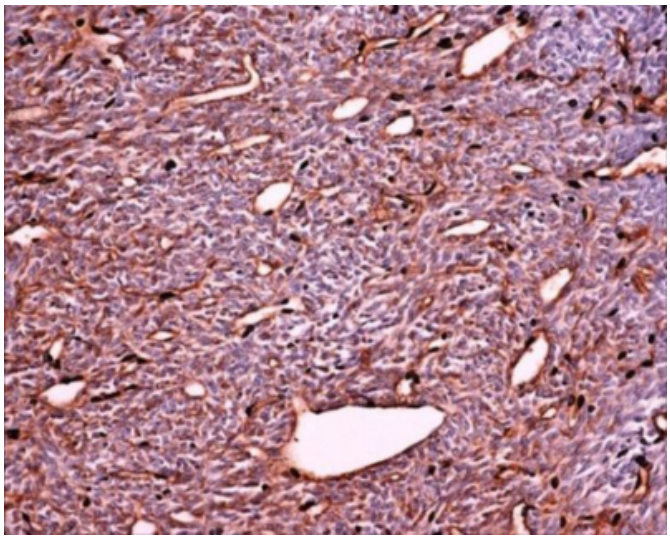


Figure 4

Figure 4: IHC for CD34 - tumor cells strongly positive



DISCUSSION

Hemangiopericytoma is a vascular tumor arising from pericytes that surround the basement membrane of capillaries. Pericytes are cells with long processes that surround the capillaries and serve to alter the caliber of the capillary lumen. Hemangiopericytomas can either be benign or malignant. Two types, infantile and adult type hemangiopericytoma, have been described. Microscopically, hemangiocytomas are characterized by a so-called pericytoma pattern with tightly packed spindle cells and prominent vascular channels. No clinical or histological feature predicts biologic aggressiveness [3] . Malignant hemangiopericytoma is recognized by cellularity, increased mitotic rate, hemorrhage and necrosis. Immunohistochemically, hemangiopericytomas demonstrate positivity for CD34, vimentin and type IV collagen and are negative for S-100 protein, neuron specific enolase, and factor VIII related antigen, carcinoembryonic antigen, desmin, laminin, and cytokeratin. [3]

Soft-tissue hemangiopericytoma is a controversial pathologic entity owing to nonspecificity of characteristic branching capillary pattern, cytological features of the constituent cells and lack of distinct immunohistochemical profile. This has led to uncertainty and absence of consensus regarding these tumors. These tumors are often confused with synovial sarcoma, fibrous histiocytoma, solitary fibrous tumor and mesenchymal chondrosarcoma as they all share hemangiopericytoma-like vascularity.

Hemangiopericytoma is a rare tumor of adult life. Pediatric cases account for less than 10% of all hemangiopericytomas

and approximately 3% of all soft-tissue sarcomas in this age group. [4] The tumors occur anywhere in the body, the most common locations being lower extremities, pelvis and the head and neck. They are deep-seated masses and found in muscle tissue. Dermal and subcutaneous locations are less common. Most tumors present as slow-growing tumors and symptoms are non-specific. Pain is a late symptom associated with an enlarging mass. Cases presenting with paraneoplastic hypoglycemia, hypertension or gynecomastia due to secretion of insulin-like growth factor and renin have been reported. [5,6] Radiological findings are non-specific.

Two distinct clinical entities have been described: the infantile type occurring in the first year of life and the adult type in adults and children older than 1 year. Most infantile hemangiopericytomas are congenital. Clinical features and biological behavior are unique, mostly located in subcutis and oral cavity locations, multifocal and metastatic forms are reported. They respond well to chemotherapy and have favorable prognosis. [7]

Adult types usually have a worse prognosis. Ten-year survival has been reported from 47% to 70%. [8,9] Malignant hemangiopericytoma is capable of local recurrence and distant metastases. The most common sites of metastases are lungs and bone.

Surgical radical excision is the mainstay of treatment. Preoperative embolization has been tried for reducing intraoperative blood loss. [10] For these vascular tumors in the retroperitoneum, rapid resection helps to reduce the blood loss. It may be more troublesome if attempts to control bleeding by blind clamping are made. Radiotherapy has been demonstrated to have a role in reducing local recurrence in incomplete surgical resection. Radiotherapy has been studied in adjuvant setting; surgery and postoperative radiotherapy with 50Gy or more resulted in significantly improved local control compared to surgery alone. Infantile hemangiopericytomas have been found to be highly chemoresponsive. Although surgical resection is the mainstay of treatment, chemotherapy has been used in unresectable and life-threatening tumors.

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