■ *Case Report*

Hemangiopericytoma in the retroperitoneal space

A Bajracharya[1], R Gupta[1], C S Agrawal[1], S Karki[2]
Department of Surgery[1], Department of Pathology[2]
B P Koirala Institute of Health Sciences, Dharan, Nepal

Abstract

Introduction: A hemangiopericytoma is a rare, soft-tissue tumor of vascular origin derived from a pericyte of Zimmerman, which is a modified smooth muscle cell that surrounds the small blood vessels. Hemangiopericytomas can occur wherever there are vascular capillaries. However, there were no previous reports of a hemangiopericytoma in the retroperitoneal space. **Case presentation:** We describe the first reported case of a hemangiopericytoma found in the retroperitoneal space. A 25-year-old man presented with a palpable tumor on the left abdomen. Preoperative imaging indicated that the tumor was in the retroperitoneal space without invasion of other organs. A complete resection was performed via a midline incision. The histological and immunohistochemical staining patterns supported the diagnosis of a hemangiopericytoma. **Conclusion:** A complete resection without piecemeal excision is the best way to treat a hemangiopericytoma. Recognizing the presence of a hemangiopericytoma in the retroperitoneal space requires appropriate surgery.

Introduction

A hemangiopericytoma is a rare, soft-tissue tumor of vascular origin derived from a pericyte of Zimmerman, which is a modified smooth muscle cell that surrounds the small blood vessels. This type of tumor was first described by Stout and Murray in 1942[1]. It represents approximately 5% of all sarcomatous tumors, and can occur anywhere, but more usually in the musculature of the extremities, sacrococcygeal space, pelvis (uterus, ovary, and urinary bladder), head, neck and lungs[2]. There are no reports of a hemangiopericytoma in the retroperitoneum space. Since the recommended treatment for a hemangiopericytoma is wide excision, due to high local recurrence[3,4], it is important to recognize the presence of this malignant tumor in this area, where various tumors occur. We describe a rare case of a hemangiopericytoma in the retroperitoneal space

Address for correspondence
Dr. Amir Bajracharya (MS), Assistant Professor
Department of Surgery
B. P. Koirala Institute of Health Sciences
Email: dr_amirb@hotmail.com

Case presentation

A 25-year man presented with a palpable painful lump on the left side of abdomen for last five months. The lump was firm with a smooth surface. Computed tomography (CT) scan showed a mass in the

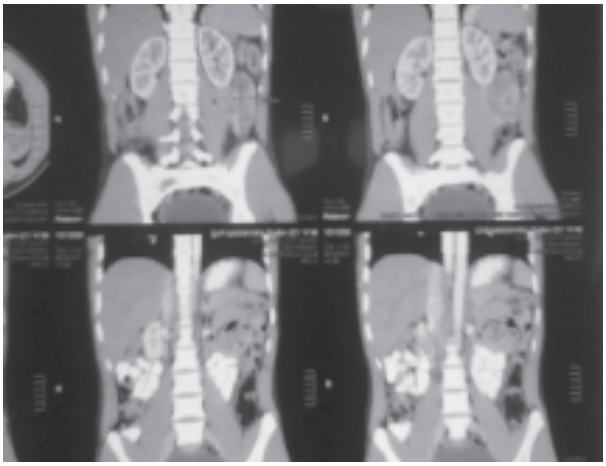

Figure 1: shows a huge mass size of 8.5x6x4cm adjacent to lower pole of left kidney free from bowel.

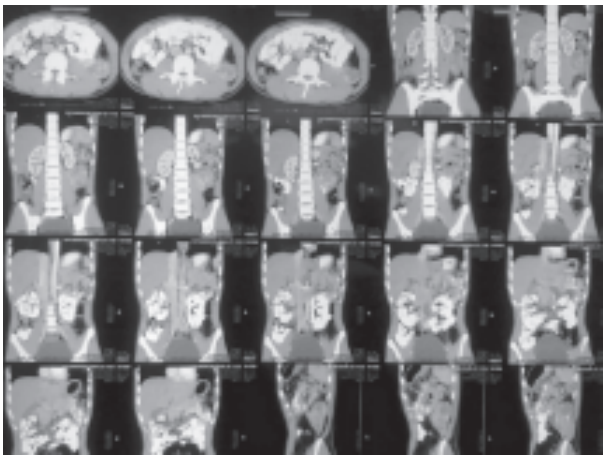

Figure 2: Retroperitoneal mass free from adjacent structure.

retroperitoneum space adjacent to lower pole of left kidney. (Figure:1).

The pre-operative images indicated that the tumor was derived from the retroperitoneum. Because of its vascularity, the pre-operative diagnosis was a soft-tissue tumor such as a solitary fibrous tumor, fibrous histiocytoma, synovial sarcoma, mesenchymal chondrosarcoma, or hemangiosarcoma. A biopsy was avoided due to the risk of needle track seeding. The patient underwent a tumorectomy via midline incision.The tumor was completely removed (Figure 3 & 4).

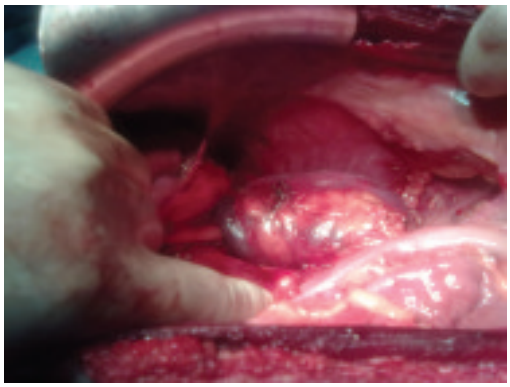

Figure 3: Intraoperative tumor localization

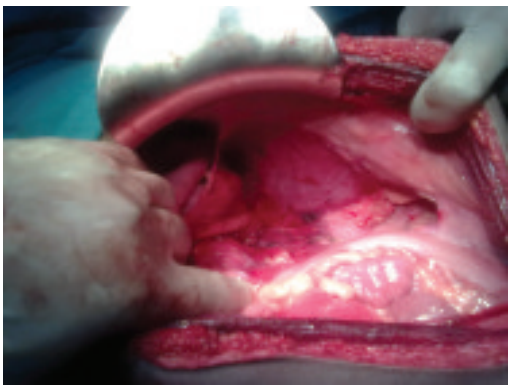

Figure 4: Complete excision of tumor.

The excised tumor was 8.5x6x4cm in diameter with a capsule. Its cut surface was mostly grayish-white and partially reddish (Figure 5). Histopathological features of the hematoxylin and eosin staining revealed that the tumor contained spindle-shaped cells surrounding the endothelial-lined vascular spaces, which is consistent with the histology of hemangiopericytoma (Figure 6). Argyrophil fibers were seen on silver impregnation surrounding the tumor cells. We performed an immunohistological analysis to obtain a diagnosis of the type of mesenchymal tumor. The mesenchymal tumor cells in our patient stained positive for CD34 and reticulin preparation revels enclosure of single cells. The mitotic rate was 1 per 10 high-power fields. No necrotic lesion was observed in our patient's tumor.

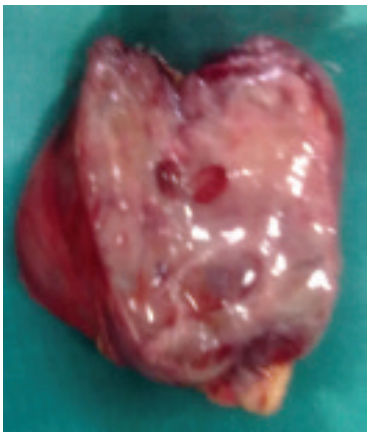

Figure 5 : Tumor cut section (8.5x6x4cm in diameter)

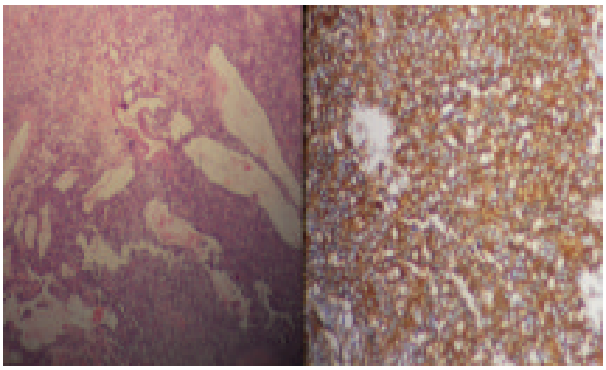

Figure 6a: Hematoxylin and eosin staining revealing spindle-shaped cells surrounding the endothelial-lined vascular spaces.

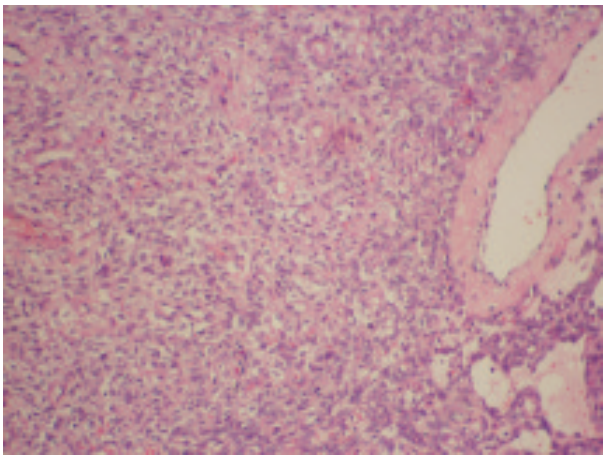

Figure 6b: Immunohistochemistry demonstrating CD34 positive tumor cells.

The tumor was pathologically diagnosed as a hemangiopericytoma.

Discussion

Little has been published about hemangiopericytoma, a rare, soft-tissue tumor. It can occur anywhere vascular capillaries are found. The tumors most commonly occurs in the musculature of the extremities, pelvis (uterus, ovary, and urinary bladder), head, neck and lungs [3,4].

The pathological diagnosis of a hemangiopericytoma, in comparison to other mesenchymal tumors such as solitary fibrous tumors, can be difficult[6]. Bcl-2 and CD99 immunohistochemistry were used to distinguish a hemangiopericytoma from a solitary fibrous tumor because a solitary fibrous tumor is positive for Bcl-2[7] and CD99[8]. 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 the mass was positive for CD34 and vimentin and negative for Bcl-2, CD99, c-kit, factor VIII, desmin, alpha-smooth muscle actin, S-100 protein, epithelial membrane antigen, and keratin upon immunohistochemical analysis. Making a differential diagnosis between a solitary fibrous tumor and a hemangiopericytoma is particularly difficult and controversial[9], and a novel molecular marker for distinguishing between the two close entities is required.

Radiotherapy and chemotherapy are not generally effective for the treatment of a hemangiopericytoma[10]. Some have advocated the use of adjuvant radiotherapy in response to the locally aggressive nature of hemangiopericytomas but, due to tumor radioresistance, no differences in local disease control were observed between treatment with and without adjuvant radiotherapy[11]. Spitz *et al.* reported that hemangiopericytomas showed a poor response to chemotherapy. They treated six patients with pre-operative chemotherapy, and only one of them responded to anthracycline-based chemotherapy[3]. Therefore, complete surgical resection is the only effective therapy for hemangiopericytoma.

Spitz *et al.* also reported that 5-year and 10-year survival rates of patients with a hemangiopericytoma were 71% and 54%, respectively. In addition, they noted that the survival rate differed between a curative and a non-curative resection. The 5-year survival rate in patients treated with curative resection and non-curative resection was 79% and 50%, respectively[3]. These data indicate that a complete resection is necessary to improve patients' survival rates.

Conclusion

This report presented a rare case of a hemangiopericytoma in the retroperitoneal space. Many diseases are associated with retroperitoneum, therefore a thorough differential diagnosis and complete resection without piecemeal excision must always be performed in the management of this type of malignant tumor.

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