Retroperitoneal Hemangiopericytoma in a young woman. Case report and literature review

Andrea Balla, Stefano Mancini, Marco Catarci, Annarita Costantino, Giovanni Battista Grassi

Department of Oncology, General and Oncologic Surgery, “San Filippo Neri” Hospital, Rome, Italy

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Hemangiopericytoma (HPC) is a primary tumor with mesenchymal vascular origin that represents 1% of all vascular neoplasms. HPC develops from the Zimmerman's pericytes around capillaries venules and it is possible to observe it frequently in the extremities, pelvis, retroperitoneum, head, neck and meninges. The only definitive parameter of malignancy is the development of recurrence or distant metastases. This report describes a case of symptomatic retroperitoneal HPC in a young female patient treated by surgical complete removal of the mass, and literature review. Despite the relatively simple surgical management of retroperitoneal Hemangiopericytoma, its diagnosis still remains difficult and often is incidentally. Patients should undergo a close long-term follow up, by periodic CT scan, due to the high probability of local recurrence or distant metastases that can occur also many years after surgery.

KEY WORDS: Hemangiopericytoma (HPC), Retroperitoneum, Surgery

Introduction

Stout and Murray in 1942, for the first time, described Hemangiopericytoma (HPC) as a primary tumor with mesenchymal vascular origin that represents 1% of all vascular neoplasms. HPC develops from the Zimmerman's pericytes around capillaries venules and it is possible to observe it frequently in the extremities, pelvis, retroperitoneum, head, neck and meninges. Other studies, however, suggest that abdominal or retroperitoneal HPC are often malignant tumors although the only definitive parameter of malignancy is the development of recurrence or distant metastases. In most of cases, HPC is an asymptomatic tumor, but in literature is associated with palpable mass, pain or hypoglycemia. This report describes a case of symptomatic HPC in a young female patient whose HPC was completely removed, and literature review.

Case Report

A 46-years-old woman (Body Mass Index 20.2), was admitted in authors’ center for severe anemia. During diagnostic work up a mass in right side under the liver was incidentally discovered. Tumor markers' assay was: carbohydrate antigen 19.9 (CA 19.9) 12 IU/ml (range 0-37), carcinoma antigen 125 (CA 125) 27 U/ml (range 0-35) and alfa feto-protein 3.5 ng/ml (range 0.0-13.4). After a negative clinical examination, the patient underwent a Total Body Computed Tomography (CT) scan, which showed, in the retroperitoneum, a contrast-
Fig. 1. Bulky mass under the liver (8x4.3 cm) with fluid collection and internal septum inside, vascularized and with contrast-enhancement. Cleavage plane with kidney and liver preserved at CT scan (A, B). Bulky mass at MRI (C, D).

Fig. 2. A: Area with sustained proliferative index Ki67 x40 hpf. B: Stromal positive elements to specific color immunohistochemistry, Smooth muscle actin x20 hpf. C: Vascular positive elements to specific color immunohistochemistry CD34 x10 hpf. D: Stromal positive elements to specific color immunohistochemistry vimentin x10 hpf.
enhancement bulky mass (8 x 4.3 cm) with fluid collection and internal septum inside, and vascularized (Fig. 1). Abdominal magnetic resonance imaging (MRI) confirmed the presence of the mass (8x5x3.5 cm) by contrast-enhancement (Fig. 1).

Considering the clinical case and after multidisciplinary discussion with surgeons, radiologists, oncologists and radiotherapists, the patient underwent surgical excision of the mass with a right subcostal incision. Intra-operatively 3 vascular pedicles were detected, deriving from the intercostal vessels. Definitive pathology showed: low grade mesenchymal tumor Hemangiopericytoma like, BCL-2+, CD34+, CD31+, vimentin+, smooth muscle actin, HBME-1, CD99, desmin, calretinin, AE1/AE3, CD68 and S-100 protein. Positive Ki67 in 7% of tumoral cells (Fig. 2).

Postoperative course was uneventful and patient was discharged in the fourth postoperative day. After oncological counseling, the patient was enrolled in a close follow up program.

Discussion

Hemangiopericytoma is a rare tumor with same distribution in both gender and a predominance in the fifth decade, such as in the present report. Diagnosis is frequent incidentally during exam for other pathologies, anyway HPC is often encapsulated by a pseudocapsule without infiltration in other organs. Typically it is a large and lobulated, hypervascular soft mass, and for this reason biopsy is contraindicated and pathological diagnosis is established only after surgery. The hypervascularization is an important feature to make the diagnosis in particular by angiography and MRI. Surgical treatment is still the main treatment, but tumor embolization is also proposed in literature before surgery. Moreover, due to radio-sensibility of HPC, it is possible to treat this tumor by adjuvant radiotherapy. However, even if the real impact of adjuvant therapy is difficult to evaluate, HPC is a potential malignant tumor, but definitive criteria or specific marker have not yet been developed.

Enzinger et al. described malignant form of HPC as an increased cellularity, foci of necrosis, hemorrhage and increased mitotic activity, but still now the only definitive parameter of malignancy is the development of local recurrence or distant metastases.

In literature, the development of local recurrence or distant metastases ranged from 13 and 23% of cases and the treatment of choice for both is a wide surgical excision, when possible. Metastases occur by hematogenous and lymphogenous course and most frequently were observed in lung, pleura, liver, bones and regional lymph-nodes. Anyway, after complete surgical excision, a 10-year survivals rate ranging from 47 to 86% is reported.

Conclusion

Despite the relatively simple surgical management of retroperitoneal Hemangiopericytoma, its diagnosis still remains difficult and often it is incidentally detected. Patients should undergo a close long-term follow up, by periodic CT scan, due to the high probability of local recurrence or distant metastases that can occur also many years after surgery.

References
