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CASE REPORTS

Pelvic hemangiopericytoma. An unusual location of a vascular tumor

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ABSTRACT

Background: The hemangiopericytoma is an uncommon vascular tumour. We report a case of interest because of its rarity, size and location.

Case report: We present the case of a 63-year-old woman who consulted for abdominal pain. TAC, MRI and arteriography showed a pelvic mass occupying Douglas' space, displacing the uterus, bladder and sigma, with vascularisation relative to the inferior mesenteric artery and both hypogastric arteries. The vascularity of the tumour itself was selectively embolised before the mass was resected. There were no intra- or postoperative complications. Pathology confirmed the diagnosis of hemangiopericitoma. The patient is being monitored as an outpatient, with no signs of recurrence to date.

Discussion: The hemangiopericytoma is a tumour of the pericyte cells so it can occur in any location. The pelvic location is exceptional. The tumour may appear as nonspecific abdominal pain, show signs of compression of adjacent organs or occasionally be associated with paraneoplastic syndromes. The diagnosis is suspected via CT and angiography findings, but confirmation is only made by analysing the surgical specimen. The treatment of choice is surgery, in some cases after preoperative embolisation of the vascularisation of the mass. There is no agreement on chemo/radiotherapy as the primary treatment for hemangiopericytoma, although adjuvant radiation therapy has been found to improve local control and reduce recurrences. The prognosis is good if complete resection is achieved, with five- and 10-year survival rates between 70 and 80%, depending on the series.

Key words: Hemangiopericytoma. Vascular. Pelvis.

INTRODUCTION

The hemangiopericytoma is a rare and potentially malignant vascular tumor, which includes 1% of vascular tumors (1). It originates from the mesenchymal cells of the pericyte (1,2). We report a case of interest because of its rarity, size and location.

CASE REPORT

This is a 63-year-old woman who consulted for abdominal pain. The CT and MRI showed a solid cystic mass of 12 x 9 x 9.4 cm with punctate calcifications and a rich vascularity occupying almost the entire Douglas space, displacing the uterus (U), bladder (V) and the right side of sigmoid colon, but without infiltrating them. The study was completed with an arteriography that showed vascular contributions relative to the inferior mesenteric artery (IMA) and both hypogastric arteries (AH) (Fig. 1).

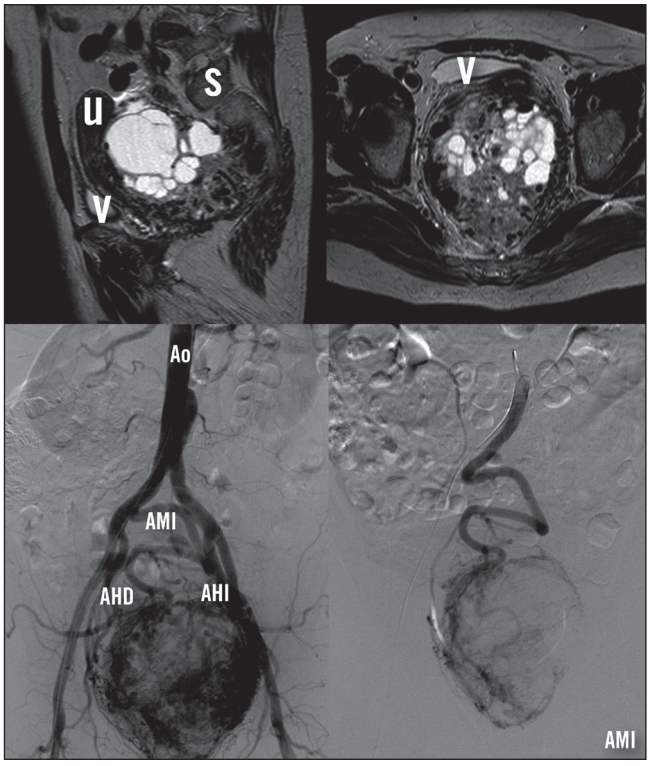

Fig. 1. The MRI showed a big pelvic tumor displacing the uterus (U), bladder (V) and sigmoid colon. The arteriography showed vascular contributions relative to the inferior mesenteric artery (IMA) and both hypogastric arteries (AH).

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Prior to the surgery, a selective embolization of the tumor vasculature was performed to reduce the risk of intraoperative bleeding, and then, the pelvic mass was successfully removed without intra- or postoperative complications. Histology confirmed the diagnosis of pelvic hemangiopericytoma with free resection edges. The patient is currently being monitored as an outpatient with no clinical or radiological signs of recurrence to date.

DISCUSSION

Hemangiopericytoma is a rare tumor of the pericyte that surrounds capillaries and venules, so it can occur at any location (1,3). The most common locations are the lower limbs, followed by the retroperitoneal area. Pelvic location is exceptional.

Clinically, the tumor usually appears as a painless mass, but it may also show signs of compression of neighboring organs or the development of paraneoplastic syndromes (1,2,4,5). CT and angiography confirm the diagnosis. Percutaneous biopsy is not recommended because of the high risk of bleeding. The definitive diagnosis is anatomopathological after analyzing the surgical specimen (6). The distinction between benign or malignant hemangiopericitoma is based on clinical and pathological criteria (7). It is considered to be malignant if it invades nearby structures, develops metastasis or recurs during follow-up (7). Although there is no histological pattern of malignancy, it has been shown that size, high mitotic index, high cellularity and the presence of necrosis or hemorrhage can promote the development of recurrence and/or metastasis (2,7). The treatment of choice is complete resection, sometimes preceded by embolization of the vascularization of the mass to decrease the risk of bleeding (1,7,8). There is disagreement about the effectiveness of chemo/radiotherapy as the primary treatment. The use of adjuvant radiation therapy has been described for improving local control and reducing recurrences, although it is considered to be a relatively radio-resistant tumor (1,2,9,10). The prognosis is conditional on achieving a complete resection, with five- and ten-year survival rates between 70 and 80%, depending on the series (6). The recurrence rate ranges between 20 and 50% after resection. Long-term monitoring is recommended because recurrence may appear at a later time (2).

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