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CASE REPORT

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Primary retroperitoneal cavernous hemangioma: An exceptional disease in adulthood

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Abstract

Primary retroperitoneal cavernous hemangioma is an extremely rare disease in clinical practice. It is classified as a vascular tumor. Only three cases have been reported in the literature. The diagnosis is uncommon due to the lack of specific radiological features. It becomes symptomatic as a consequence of its enormous size or complications like rupture or compression. We herein report a unique case of primary retroperitoneal cavernous hemangioma treated with conventional surgery in a 35-year-old male patient admitted to our department for chronic abdominal pain. Retroperitoneal cavernous hemangioma is an extremely rare vascular tumor in adulthood. Confirmation is made by histopathological examination after total surgical resection.

KEYWORDS

cavernous hemangioma, hemangiomas, primary retroperitoneal tumors, retroperitoneal tumors

1 | INTRODUCTION

Cavernous hemangioma is a vascular tumor containing proliferative vascular endothelial cells with an angiogenic phenomenon.[1] This type of tumor is usually reported in the orbital, skin, mucosal areas, and liver.[2] It is exceptionally found within the retroperitoneal cavity.[3,4]

In the literature since 1950, fewer than 30 cases of adult retroperitoneal hemangioma have been reported in the literature.[5,6] The majority of retroperitoneal hemangiomas originate from the kidney, pancreas, and adrenal glands. Primary retroperitoneal cavernous hemangioma is even rarer and is unique in that it is separated from the surrounding organs. In the literature, only three cases have been reported as primary retroperitoneal hemangiomas.[3,7,8] To our knowledge, we are reporting the fourth case of primary retroperitoneal cavernous hemangioma and also the first case in Tunisia and Africa.

2 | CASE PRESENTATION

A 35-year-old male patient, without any medical history, was admitted to our department for chronic vague abdominal pain. The physical examination was normal, without any masses. There were no signs of other disorders, including fever, jaundice, vomiting, or hematuria. Laboratory studies were normal.

A subsequent CT (computed tomography) scan revealed a well-circumscribed, cyst-like mass measuring

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45*27*22 mm, adjacent to the inferior vena cava and the third part of the duodenum. There was no enhancement of the wall and consistent hypodensity of the mass during arterial and portal phases, with the mass effect showing a contour deformation of the inferior vena cava. There was no evidence of feeding arteries from surrounding organs or adenopathy, and there was no obstruction of the right upper urinary tract. (Figure 1).

Tumor markers were within normal ranges.

Given that the tumor was localized and there was no evidence of invasion or metastasis to organs, the diagnosis of a benign tumor was suspected, such as lymphangioma cyst, GIST (gastrointestinal stromal tumor), or primitive retroperitoneal benign tumor.

A vertical midline incision and the Kocher maneuver were used for the surgery. A tumor measuring 4 cm was found localized in the inferior portion of the third part of the duodenum, anterior to the inferior vena cave, and medial to the urinary tract. (Figure 2).

There was no evidence of invasion of the surrounding organs. Total resection of the tumor was performed. (Figure 3).

The postoperative went uneventfully, and the patient was discharged after 4 days.

Pathological examination revealed a benign vascular proliferation composed of venular and capillary structures. CD34 and CD31 immunostaining exhibited diffuse positivity on the endothelium of these vascular channels, confirming their endothelial origin and the diagnosis of cavernous hemangioma. DOG1 and CD117 were negative, allowing us to eliminate a GIST. There were no signs of malignancy. (Figure 4).

The patient did not present any recurrence of pain or symptoms at 12 months of follow-up.

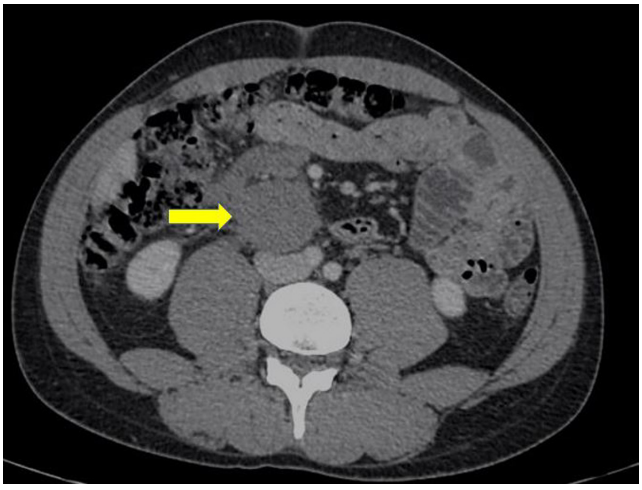

FIGURE 1 Axial view of the abdominal computed tomography scan showing a retroperitoneal mass. (Yellow arrow)

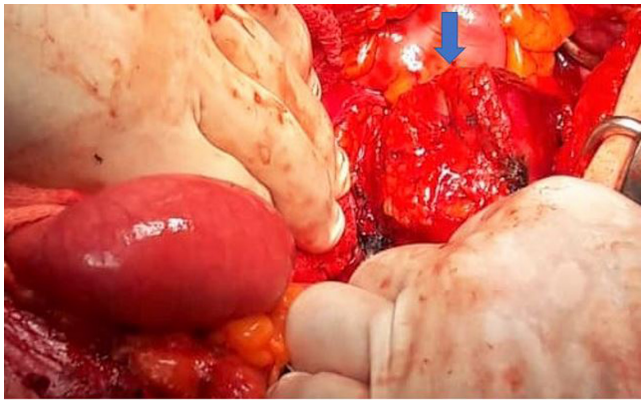

FIGURE 2 Intraoperative findings: after duodenal mobilization, a 4-cm retroperitoneal tumor without any specific communication. (Blue arrow)

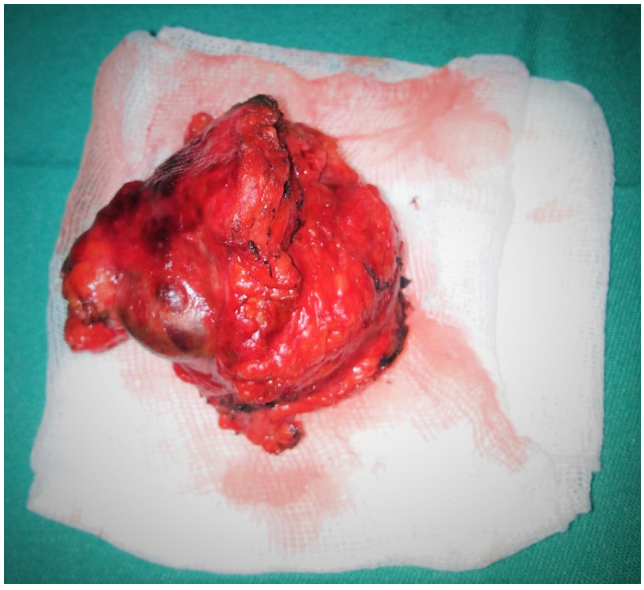

FIGURE 3 The operative specimen after total resection

3 | DISCUSSION

Retroperitoneal tumors account for <0.2% of all tumor types. Among malignant tumors located in the retroperitoneal space, liposarcomas and leiomyosarcomas are the most frequent, while teratomas, cysts, and neurinomas are common benign masses. Cavernous hemangioma is extremely rare.[6,9]

Retroperitoneal hemangioma is very rare and is only confirmed in <5% of all retroperitoneal tumors in adults.[9] It is considered a congenital abnormality and is commonly seen in infants originating from residual embryonic angioblasts.[6] This pathology is associated in some cases with the Kasabach–Merritt phenomenon.[1] It is characterized by spontaneous involution and regression after childhood.[1,3] It seems it can affect both sexes similarly.[1,3]

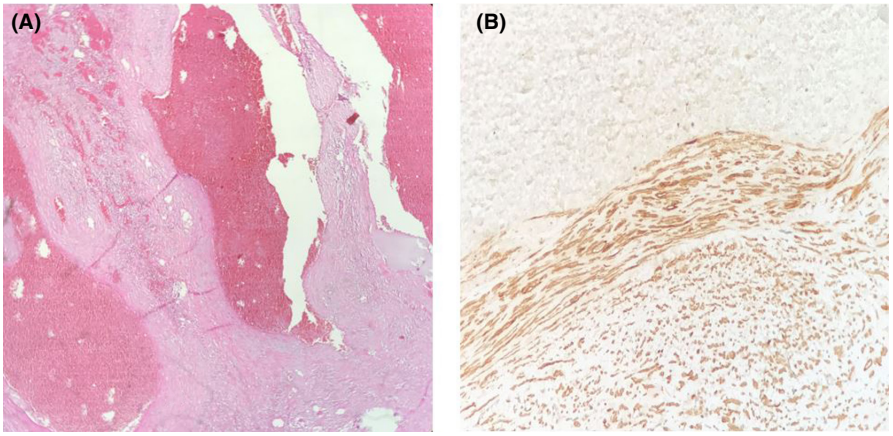

FIGURE 4 (A) vascular proliferation composed of venular and capillary structures (X 100), (B) CD31 Immunostaining positivity on the endothelium of the vascular channels (X 100)

But it is an uncommon disease in adulthood. In the literature, approximately, <30 cases have been reported since 1950, including 19 cases in Japan.[1,5,10] This rare tumor often originates from the kidney, urinary tract, pancreas, and adrenal glands. Primary retroperitoneal cavernous hemangioma is even rarer and is unique in that it is separated from the surrounding organs. Till today, only three cases have been reported as primary retroperitoneal hemangiomas.[3,7,8]

The mode of establishing the diagnosis is very variable. There are no specific symptoms for retroperitoneal localization. Cavernous hemangioma is usually asymptomatic, especially in the early stages and in small sizes. However, compression or the invasion of the adjacent anatomical structures may induce non-specific symptoms such as vague abdominal pain, hematuria, renal vein thrombosis, hydronephrosis, and upper ureterectasis.[11] Meanwhile, some were incidentally diagnosed by histopathological examination after resection of suspected masses identified by radiological images.[3] Bleeding with hemorrhagic shock has been reported as a complication of the cavernous hemangioma, signifying the potential danger represented by this tumor in case of rupture or bleeding.[12]

Radiological images are not specific. The CT scan revealed a cystic mass with minor contrast enhancement, and it may also be present, like in our case, as a homogeneous hypodense component tumor without enhancement during the artery or portal phase.[5,13] The reasons for atypical findings might be neovascularity, thrombosis, or hemorrhage.[10] MRI (Magnetic resonance imaging) may help with the diagnosis. On T1-weighted imaging, it usually shows low signal intensity, whereas in T2-weighted imaging, hemorrhage can be interpreted with high intensity.[5,6,14]

The retroperitoneal cavernous hemangioma is very difficult to diagnose before surgery. The differential diagnosis included GIST, lymphangioma cyst, or primary retroperitoneal tumors.

There is no gold standard therapeutic option to follow. But it seems that surgical management is the best option, as publications agreed, for two reasons. The first reason is the histopathological confirmation of the mass, excluding any malignancy. The second reason is the therapeutic goal and avoiding complications such as bleeding, invasion, and compression.[12]

The laparotomy (median or subcostal incision) was mainly performed. In some selective cases, robotic surgery was performed.[15] Resection of the mass is usually recommended. Combined resection of the mass with surrounding organs seems rational to avoid local recurrence. Pathologically, the inner components of the tumor consisted of variously sized vascular spaces lined by a single layer of flattened cells, which stained positive for CD34 and CD31.[5]

4 | CONCLUSION

Retroperitoneal cavernous hemangioma is an extremely rare vascular tumor in adulthood. The tumor was separated from the surrounding organs without any specific feeding artery, so it was diagnosed as a primary retroperitoneal cavernous hemangioma. It is difficult to make a clinical or radiological diagnosis. Surgical removal is the only curative treatment to avoid complications. The diagnosis is confirmed by histopathological examination.

AUTHOR CONTRIBUTION

Debaibi M conceived the idea for the document and contributed to the writing of the manuscript. Sghair Asma contributed to the writing and editing of the manuscript. Sahnoun Moez contributed to the acquisition and conception of the manuscript. Essid Rime reviewed and edited the manuscript. Zouari Rami reviewed and edited the manuscript. Kchaou Majdi contributed to the literature review and manuscript writing. Dhaoui Amen contributed to

supervision and edited the manuscript. Chouchen Adnen approved the final manuscript and supervision.

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Published with the consent of the patient

CONFLICTS OF INTEREST

None.

DATA AVAILABILITY STATEMENT

Personal data of the patient were respected. No data are available for this submission.

ETHICAL APPROVAL

Personal data have been respected. Published with the consent of the patient.

CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

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