

Focal, Extranodal Primary Polymorph Hemangioendothelioma Treated With Endovascular Embolization Plus Open Surgery Resection: Rare Case Report and Review

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Case report

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Abstract

Background: We report the rare case of an adult with a primary extranodal polymorphous hemangioendothelioma at dorsum, treated with endovascular coiling followed by open surgery resection. Clinical history, findings of spinal arteriography, histopathological findings and combined surgical procedure are reported.

Case report: A 22-year-old male patient complained of a dorsal mass that has been increasing in size, painful, soft, no mobile and without neurological symptoms. A vascular tumor was suspected and an endovascular followed by an open surgical approach was performed. Histopathological diagnosis of an extranodal polymorph hemangioendothelioma was documented and a total resection was confirmed by free-tumor resection margin. The postoperative course was uneventful.

Conclusion: Polymorph hemangioendothelioma is a rare vascular tumor. given the high vascular features of the lesion and, therefore, the high rate of bleeding during surgery.

Introduction

The hemangioendothelioma is a rare vascular tumor that was first described as it in 1982 (20). It has been suggested to be a tumor with biological borderline behavior (16), with an intermediary pathology between benign hemangioma and malignant angiosarcoma (1). Also, it can be divided in different subtypes depending on the cellular features. Between these subtypes are epithelioid, papillary intralymphatic, retiform, kaposiform, pseudomyogenic, composite and polymorphous. The last, is one of the most rare variants.

The first case of polymorphous hemangioendothelioma (PH) was described in 1992 in an analysis of 39 cases of primary vascular lymph node tumors where 3 of them were described as PH (4). Until 1999, 6 cases were described (15). Up to date, less than 10 cases of properly called PH can be found in the literature (4, 5, 7, 8, 13, 15, 17, 19), most of them from the lymph nodes and a rare case of the maxillary soft tissue PH (17). Only a few cases of primary extranodal presentation have been described in the literature (8, 13).

Given the small number of cases found, the natural history of the disease has not been elucidated. However, the potential of PH to metastasize and recur rate have been described (17) and, therefore, total resection has been proposed as the first line of treatment (1). Radiation (1) and immunotherapy (6) has been proposed without sufficient evidence. Also, it has not been described the utility of endovascular embolization combined with open resection in the context of a vascular malignancy with high bleeding rate during open surgery to achieve total resection.

Case Description

A 22-year-old male patient comes to the office due to a “lump” in his dorsal back. He said that it has been increasing in size. He has not lost weight in the last months, no fever or neurologic deficit. The patient consulted previously to a different hospital where a biopsy was performed two months ago. The surgical procedure presented significant blood loss and had to be suspended. The pathology result describes a lesion compatible with hemangioma. Since then, the patient has noted a significant size increase. The patient has no other medical conditions and takes no medication. Physical examination shows a 10 cm mass, no mobile, slightly painful, no erythema or secretions were documented. The rest of the examination was unremarkable.

The patient had a previous thoracic spine contrast-enhancing magnetic resonance imaging that revealed a contrast enhancement of a dorsal mass at T8 level, compromising only soft tissues. The mass does not involve the spine or medullary canal. No lymph node enhancement is documented. Surgical resection is planned, however, given the past history of difficult blood control during surgical procedure a spinal arteriography is indicated (**Fig 1 panel A to B.**). The spinal arteriography was performed with a Cobra catheter through the right femoral artery, ascending from T12 up to T5 radicular arteries. Artery blood supply to the dorsal mass was registered mainly from T10 segmentary arteries and, in less degree, by segmentary arteries of T11. A selective catheterization of T10-radiculomedullary branch is done, Adamkiewicz artery is not present at this level (confirmed by enhance injection), and coil-embolization is done. Neurologic monitory is continuously performed without abnormalities. In contrast, T11-radiculomedullary embolization could not be done because multiple branches directing to the spinal cord are documented. A reduction of arterial supply is registered up to 60%.

Posteriorly, the patient is taken to surgical resection. Dissection through the tumor plane was performed, and the vascular supply was coagulated with bipolar. Complete resection of the lesion was achieved (**Fig 2**) and confirmed with histopathological (**Fig 3 panel A to D**) report of free-tumor borders. Patient recovered without complications and was discharged 24 hours later.

Discussion

The hemangiothelioma is a borderline vascular tumor between benign hemangioma and malignant sarcoma. Seven different subtypes of properly hemangioendothelioma has been described (16): epithelioid variant characterized by round, oval, and polygonal cells, with abundant pale eosinophilic cytoplasm embedded in a fibromyxoid or sclerotic stroma; papillary intralymphatic (*Dabska tumor*) characterized by central hyaline core lined by hobnail-like endothelial cells protruding into the lumina; retiform subtype with elongated arborizing vessels arranged in anastomosing pattern resembling rete testis; kaposiform subtype characterized by several solid poorly circumscribed nodules, each constituted by a mixture of small capillaries and solid lobules of endothelial cells arranged in glomeruloid pattern; pseudomyogenic hemangioendothelioma (sarcoma-like hemangioendothelioma) characterized by a poorly circumscribed, fascicular lesion with infiltrative borders composed of round or oval neoplastic cells; composite hemangioendothelioma, a locally aggressive vascular neoplasms of low-grade malignancy with varying combinations of benign, low-grade malignant, and high-grade malignant

vascular components (16); the polymorphous hemangioendothelioma (PH) is characterized by spindled cell areas, polygonal epitheloid areas and variable vascular density (1). This last subtype is a rare vascular tumor, commonly arising from lymph nodes and soft tissues within the thoracic cavity as the second most common location (15) and, also, clinical syndromes associated with coagulopathy, thrombocytopenia and hemolytic anemia has been described (1).

The natural history, rate of recurrence and potential to metastasize is not known up to date given the limited available literature of PH. Likewise, the first line treatment has not been elucidated, but total resection is recommended. Additionally, immunotherapy and radiotherapy has been proposed as adjuvant treatment (1, 16).

Given the vascular etiology of the PH, operative bleeding can be of concern and should be addressed. This case was approached with preoperative arteriography that revealed irrigation from radicular T10 left artery and, in the absence of Adamkiewicz artery, coil embolization was indicated and performed without complication. Afterwards, open surgical resection was performed with low operative bleeding and complete resection with tumor-free edges was obtained. This is the first case of extranodal, primary PH treated with endovascular coiling plus open resection, and showed exceptional results. Thus, this is an approach that should be taken into account.

Conclusions

Polymorph hemangioendothelioma is a rare vascular tumor. The classification and natural history depend on localization (nodal vs extranodal). Few cases had been described in the literature. Here has been described a rare case of focal, extranodal primary polymorph hemangioendothelioma treated with a combined neurosurgical approach consisting in endovascular coil-embolization plus open surgery, which can enhance total resection given the high vascular features of the lesion and, therefore, the high rate of bleeding during surgery.

Abbreviations

PH: Polymorph Hemangioendothelioma

Declarations

Ethics approval and consent to participate: not applicable

Consent for publication: the patient has consented the submission of this case report.

Availability of data and material: “Data sharing not applicable to this article as no datasets were generated or analysed during the current study”

Competing interests: The authors declare that they have no competing interest.

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Authors' contributions: ERF carried out the recollection of data, literature review and redaction of the paper. FPP carried out recollection of data. SDMN carried out the analysis of histopathological samples. LAOB carried out a substantial revision of the paper. WMRC carried out recollection of data and literature review.

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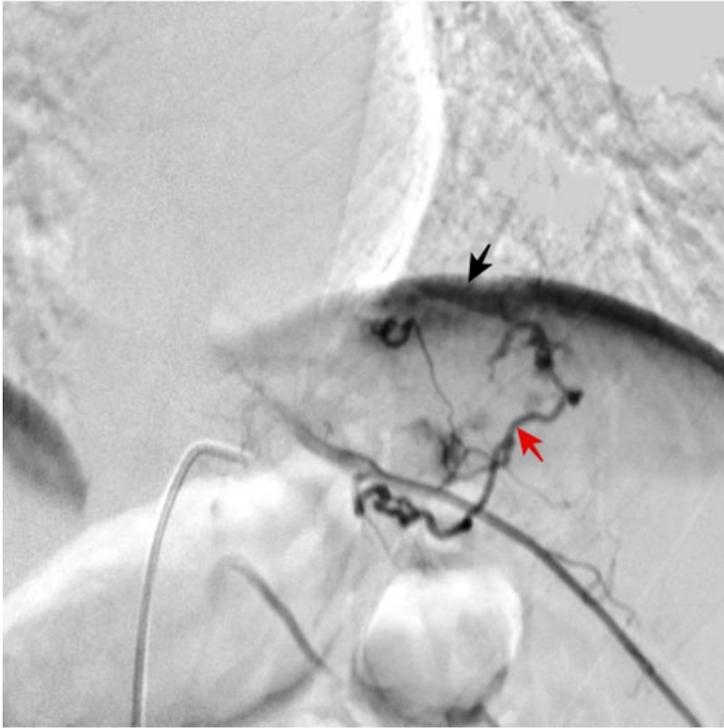
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Figures

Panel A



Panel B



Figure 1

Spinal arteriography. Panel A) arterial supply through T10 (black arrow) and T11 (red arrow) radiculomedullary arteries, was reported. Panel B) Embolization of T10 radiculo-medullary artery is performed (black arrow), T11 radiculo-medullary arteries embolization was not performed because the multiple branches visualized and remain permeable (red arrow).

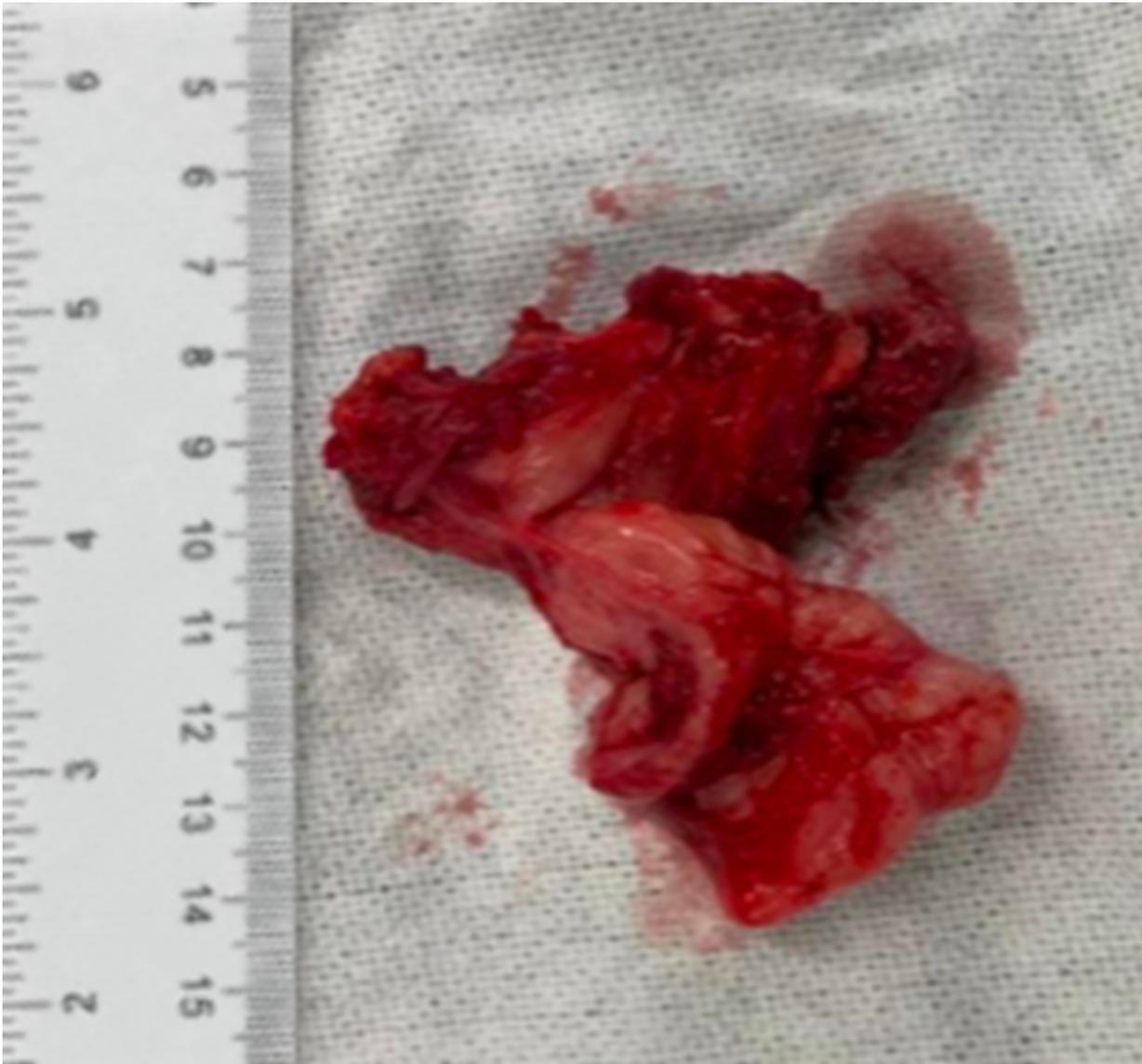
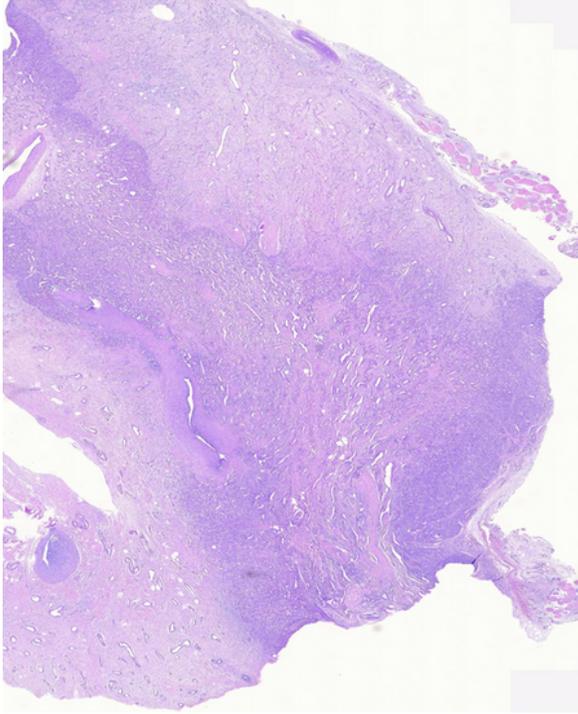


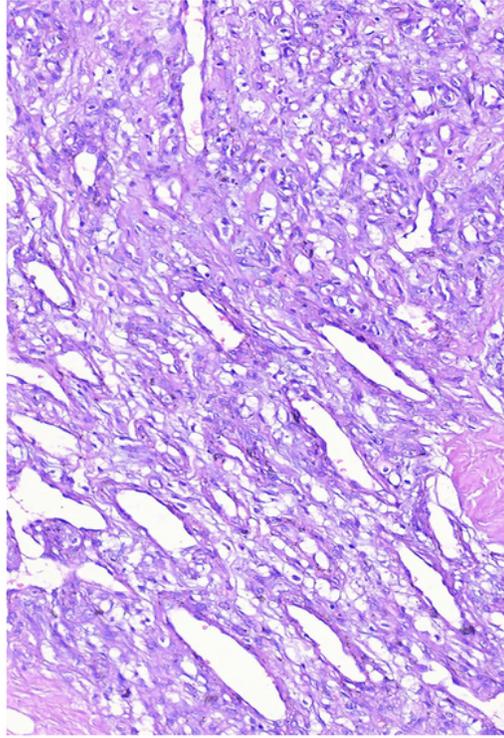
Figure 2

En bloc resection of dorsal specimen.

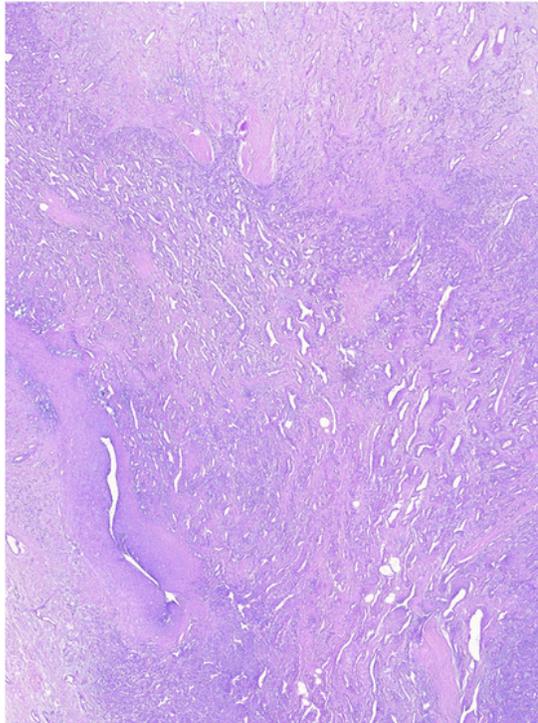
Panel A



Panel C



Panel B



Panel D

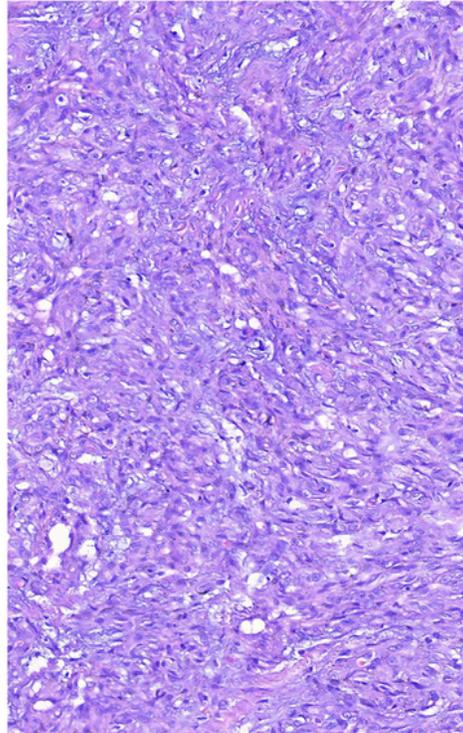


Figure 3

Panel A) Histology of the free-tumor borders mass. Panel B and C) Tumor area with angiomatous and retiform growth pattern. Panel D) Angiomatous areas composed of ovoid, polygonal, and spinal cells, with low mitotic activity.