EPITHELIOID HEMANGIOENDOTHELIOMA OF THE INTERNAL JUGULAR VEIN

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Abstract

Ephitelioid Hemangioendothelioma (EHE) is a rare type of tumor with vascular and sarcomatous components. There's only another case published of an internal jugular vein (IJV) EHE. A case of a 50 years-old woman with a palpable and pulsatile mass on the left cervical area is reported. Doppler ultrasound and magnetic resonance imaging showed an IVJ' 4 cm mass. Cytology was inconclusive. Surgical treatment was therefore decided and during surgery a mass inside the left IJV, with local nonsuspicious lymph nodes, was confirmed. The mass was resected including a segmental resection of the IJV and one affected tributary vessel. Lymphadenectomy of the adjacent cervical levels was performed. Histologic examination depicted an EHE without metastatic lymph nodes. Tumor was staged as pT1bN0M0 and a multidisciplinary sarcoma group proposed surveillance. Patient remained well, without evidence of disease and without complications in a twenty-four months follow-up period.

INTRODUCTION

The diagnosis of vascular tumors is challenging, particularly epithelioid types due to the cells' morphology.[1] Sarcomas showing endothelial differentiation represent < 1% of all sarcomas' diagnosis.[2] Hemangioendotelioma epithelioids' (EHE) clinical severity is intermediate between hemangioma and high-grade angiosarcoma.[2] Most EHEs are indolent, however 20-30% can metastasize and 15% mortality is reported.[2]

METHODS

The authors report a clinical case and present a literature review using PubMed with the key terms "epithelioid hemangioendothelioma" and "internal jugular vein".

CASE REPORT

A case of a 50 years-old caucasian woman, asymptomatic, with a left cervical mass, and relevant past medical history of multinodular goiter, referred to Vascular Surgery after positive findings in a routine cervical ultrasound (US). Upon physical examination, the cervical mass was located at II/III left levels, 4 cm long, well delimited, painless, pulsatile, without palpable cervical lymph nodes. The cervical doppler US showed a 42 mm mass in the left internal jugular vein (IJV) lumen, vascularized. A magnetic resonance imaging (MRI) showed a heterogeneous lesion in the left IJV path, measuring 46 mm, resembling a paraganglioma (Figure 1). The patient was referred to a surgical oncology referral center. Citology examination, after guided fine needle biopsy, showed a vascular proliferation, but wasn't able to define the lesions' malignant potential. Byogenic amines screening showed normal range values. In this context, surgical excision was proposed and a left lateral cervicotomy was performed showing a 4 cm nodular lesion in the left IJV extending to a thyro-facial tributary and local nonsuspicious adenomegalies (Figure 2). The mass was resected including a segmental resection of the left IJV and the affected tributary vessel (R0). Lymphadenectomy of the adjacent cervical levels was performed. Histologic examination documented a malignant angiocentric vascular neoplasm, obliterating the lumen and spreading into surrounding soft tissue, measuring 45 mm. Tumor was characterized by anastomosing cords and small nests of epithelioid cells, with abundant eosinophilic cytoplasm, containing vacuoles, some with fragmented erithrocytes;

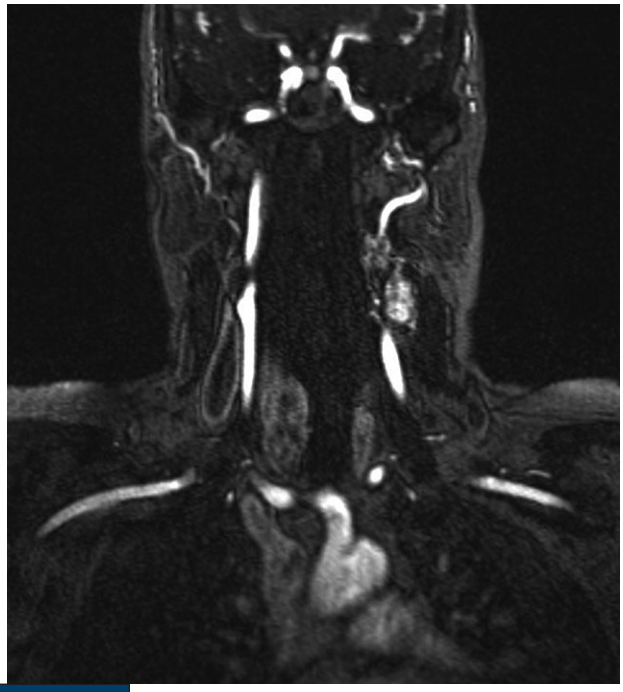

Figure 1 *MRI showing a heterogenous lesion in the left internal jugular veins' path, resembling a paraganglioma.*

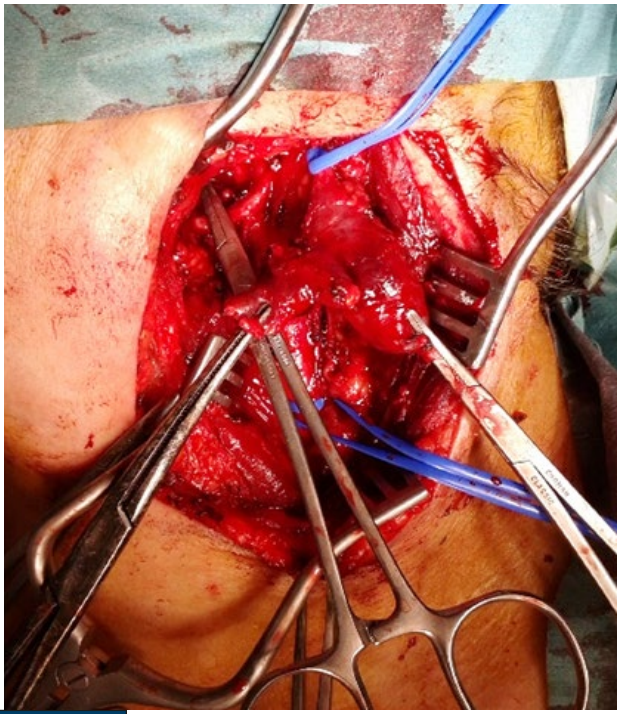

Figure 2 *Intra-operative image showing blue vascular references in distal and proximal control of the internal jugular vein (IJV); an IJV' mass; needle-holder under a tumor affected tributary vein.*

nuclei are round and may be indented, with mild to moderate pleomorphism. Minimal mitotic activity and no necrosis, lymphovascular or perineural invasion was also documented (Figure 3). The surgical margins weren't available, due to fragmentation. The eleven retrieved lymph

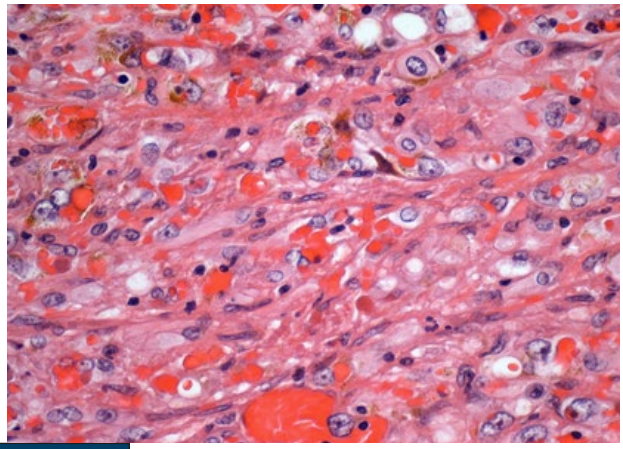

Figure 3 *Hemangioendotelioma. Malignant vascular neoplasm composed by chains and cords of epithelioid endotelial cells with abundant eosinophilic cytoplasm, containing vacuoles, some with fragmented erithrocytes; nuclei are round and may be indented, with mild to moderate pleomorphism. There are scattered inflammatory cells.*

nodes show no evidence of tumor. Epithelioid tumor cells expressed strongly endothelial markers CD31 (Figure 4) and CD34, confirming endothelial differentiation and epithelial antigens (pan-cytokeratins). The EHE was staged as grade 1 pT1bN0M0, stage Ia. A Multidisciplinary Sarcoma Group proposed surveillance, no other adjuvant treatments. Over the course of 24 months the patient remained asymptomatic, without evidence of disease (local, regional or systemic), and no complications were reported.

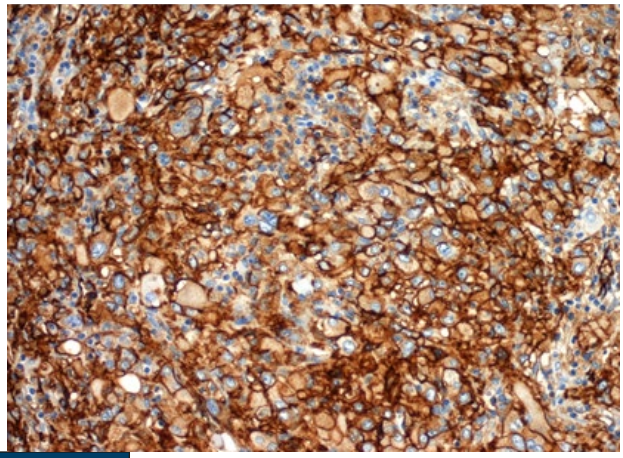

Figure 4 *Epithelioid cells express strongly endothelial marker CD31, confirming endothelial differentiation.*

DISCUSSION

To the best of our knowledge, this is the second case published of EHE of the IJV.[4] EHE has a peak incidence in the 4th-5th decade and women seem to be more affected.[5] Most of the EHE are asymptomatic, although edema or thrombosis can develop.[6] The differential diagnosis of intravascular EHE is usually difficult and often misdiagnosed as thrombosis[7] or other tumors. EHE is defined (World Health Organization) as an angiocentric vascular

tumor with metastatic potential, composed of epithelioid endothelial cells arranged in short cords and nests set in a distinctive myxohialine stroma. Immunohistological tests confirm the diagnoses, CD31 being the most sensitive and specific marker.[4] When feasible, the adequate treatment is surgical resection[5,8] and in-line prosthetic or autogenous venous reconstruction, to restore patency.[8] With veins, revascularization is seldom needed due to redundancy of the venous circulation. A complete local excision, with or without local lymphadenectomy, is positively related to long-term survival.[9] Radiation therapy should be considered in cases of high-risk features or when complete resection is not feasible.[9] Low response rates to chemotherapy makes it unattractive, used more frequently in the metastatic setting.[7] Mitotic activity and tumor size can stratify tumors into low and high-risk groups: tumors > 3 cm and > 3 mitosis/50HPFs have a 5-year disease-specific survival of 59% in contrast to 100% in those that lacked these features.[2] Other authors consider necrosis, atypia, and increased mitotic activity has prognostic factors.[10]

In conclusion, EHE is a rare form of vascular sarcoma. Literature available is mainly as case reports. This is the second case published of an IJV EHE. The adequate treatment is surgical resection. Diagnosis is often obtained after excision. Adjuvant treatments and patient' follow-up relies on prognostic factors. Despite the known prognostic factors, EHE behavior remains unpredictable.

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