Subclavian vein epithelioid hemangioendothelioma: Multidisciplinary surgical approach

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ABSTRACT

Epithelioid hemangioendothelioma (EHE) is a rare primary vascular tumour, usually malignant. We present the case of a 60-year-old woman who presented with hoarseness of voice and a persistent cough. During evaluation, a left supraclavicular mass was discovered. Initial assessments revealed a 30-mm left supraclavicular mass. Computed tomography angiogram and venogram confirmed an enhancing mass arising directly from the proximal left subclavian vein. After discussion in the joint multidisciplinary team, it was decided to resect the tumour owing to its malignant potential. Histopathology after resection confirmed a completely excised EHE. EHE is a rare vascular sarcoma requiring a multidisciplinary approach. Its main challenge is its unpredictable behaviour. (J Vasc Surg Cases Innov Tech 2024;10:101473.)

Keywords: Subclavian vein; Hemangioendothelioma; Vascular tumor; Vein resection; Trap-door incision

Epithelioid hemangioendothelioma (EHE) is a rare vascular cancer, mostly linked to two morphological and molecular variants (*CAMTA1* and *TFE3* related).[1] The prevalence of EHE is exceedingly low, with less than one case per million people.[2] This malignant vascular tumor typically affects middle-aged individuals, showing a male-to-female predilection of 2:1. The age range of patients documented with this condition varies from 7 to 83 years.[3]

Symptoms of EHE vary depending on the tumor location. This case report discusses EHE in the left subclavian vein, treated with complete surgical resection via modified trap-door sternotomy. Written informed consent was obtained from the patient for the publication of this case report.

CASE REPORT

A 60-year-old woman presented to the ENT clinic with hoarseness of voice and a persistent cough. Physical examination revealed a hard left supraclavicular mass. There were no signs of dilated veins in the upper chest or swelling in the upper limb. She had no significant medical history, and an initial assessment by computed tomography (CT) scan revealed a 30-mm left supraclavicular nodule and a 14-mm breast nodule. A biopsy of the mass indicated epithelioid hemangioendothelioma. Breast examination and ultrasound were normal. CT scans of the neck, chest, abdomen, and pelvis, along with a bone scan, revealed no additional tumors. The positron emission tomography CT scan showed increased activity in the left supraclavicular fossa, but no other sites. Two weeks after the initial scan, a CT angiogram and venogram conducted for a detailed assessment of the mass's relationship to the major vessels in the region confirmed an enhancing mass arising from the proximal left subclavian vein, measuring 16 × 20 mm. The mass blocked the immediately adjacent left subclavian vein and left internal jugular vein, with no evidence of arterial invasion (Fig 1).

After a joint sarcoma/vascular surgery multidisciplinary team meeting, the consensus was to proceed with tumor resection owing to its malignant potential. The preoperative evaluation confirmed the patient was fit for surgery, with a thorough assessment showing normal heart and lung function, adequate exercise capacity, and no abnormalities on routine blood tests.

The surgical approach for the retroclavicular tumor focused on three key aspects: precise access to the tumor using a modified trap-door incision (Fig 2), complete en bloc resection with clear margins (Fig 3), and safeguarding nearby neurovascular structures. This required a multidisciplinary surgical team involving vascular, thoracic, and cardiac specialists. The procedure began with exposing and controlling supraclavicular structures (Fig 2), including the internal jugular vein, common carotid artery, and vagus nerve (cranial nerve X). The incision was extended laterally to the left second intercostal space to access the lower part of the tumor and proximal neurovascular structures. The internal mammary artery overlying the tumor was ligated and removed. The tumor was resected completely with a 4-cm length of proximal and distal vein cuff and ligation of the SCV proximally and distally without reconstruction, leading to a successful recovery and discharge within 5 days without complications (Fig 4).

Histopathological examination and immunohistochemistry of the resected tumor confirmed the diagnosis of epithelioid hemangioendothelioma, with focal CK7 expression and weak GATA-3 staining, strong ERG and CD34 expression, and a low MIB-1 proliferation index (<5%), with negative resection margins.

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The prognosis was considered low-risk based on tumor size and mitotic activity (<3 per 50 high power fields).

Four weeks after the operation, the case was reviewed in a sarcoma multidisciplinary team meeting, leading to a decision for follow-up by a medical oncologist and annual surveillance imaging with a CT scan. At a 6-week postoperative clinic visit, the patient had recovered well with mild arm swelling; hoarseness of voice had decreased substantially.

DISCUSSION

EHE is a rare vascular sarcoma characterized by low malignancy but potential for systemic involvement. Treatment options vary globally, leading to uncertainty and suboptimal outcomes for patients.[4] EHE can originate in various parts of the body, presenting in different ways. These include solitary lesions (a single tumor), regional metastases (multiple lesions within a single organ or anatomical area), or widespread metastases (affecting multiple organs).[5] In 50% of cases, an origin from a blood vessel can be demonstrated.[5,6]

Clinical presentation upon diagnosis varies considerably. Some cases are incidental, found in asymptomatic patients, whereas others, like angiocentric EHE, may manifest with symptoms related to venous obstruction.[4] In this case, the presentation was likely due to compression of the recurrent laryngeal or phrenic nerves, rather than venous obstruction. A similar case, where EHE originated from the internal jugular vein, was initially misdiagnosed as cervical metastatic lymphadenopathy because of its presentation as a supraclavicular mass, until histopathology results revealed its true nature.[7]

Vascular EHEs usually have a venous origin, varying in location from lower limb veins to the inferior vena cava, superior vena cava, and azygos vein, with common sites being the femoral or iliac vein.[8,9] Arterial EHE cases are rare and can lead to complications like distal blue toe syndrome or form painless oval masses in the forearm, mimicking arterial occlusive disease or venous thrombosis.[10] In larger vessels, the tumor may not present as a visible mass, complicating differentiation from common occlusive conditions. In this case, standard vascular assessment could not reveal its true nature.

Owing to the rarity of EHE multidisciplinary teams with experience in sarcoma management will facilitate optimal therapy. In our case, discussions occurred in both the sarcoma multidisciplinary team and a joint vascular, thoracic, and sarcoma surgery setting. The preferred treatment for confirmed unifocal EHE is surgical intervention,[11,12] aiming for complete resection with microscopic negative (R0) margins, offering a 70% to 80% cure rate.[6] In this case, negative margins were achieved, and the low metastatic risk indicates a good prognosis.

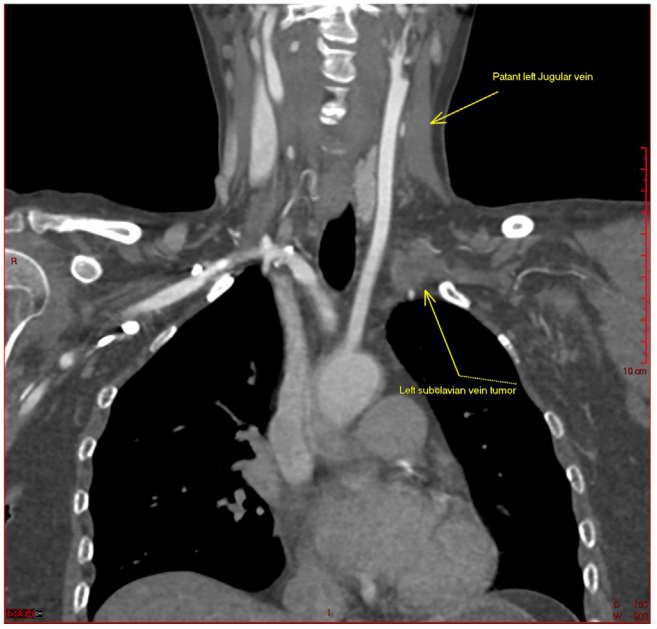

Fig 1. A 16 × 20-mm enhancing mass arising from the proximal left subclavian vein. Resulting occlusion of the proximal left subclavian vein and proximal left internal jugular vein (IJV) (patent distal IJV). No arterial invasion.

This complex case involved a tumor near the thoracic inlet, impacting vital structures like nerves and arteries. The modified trap-door approach was chosen for better exposure and control, avoiding the drawbacks of clavicular lectomy or a larger trap-door approach.

Adjunctive treatments like chemotherapy and radiotherapy remain debatable for EHE.[13] Local recurrence risk after surgery is approximately 10% to 15%, highlighting the need for long-term surveillance. Postoperative follow-up guidelines for patients with EHE vary between institutions and lack standardization.[4]

The rarity of EHEs likely precludes the possibility of conducting controlled clinical trials, suggesting that the establishment of patient registries will be crucial for gaining insights into the origins, pathophysiology, diagnosis, therapy, follow-up, and prognosis of EHE.

DISCLOSURES

None.

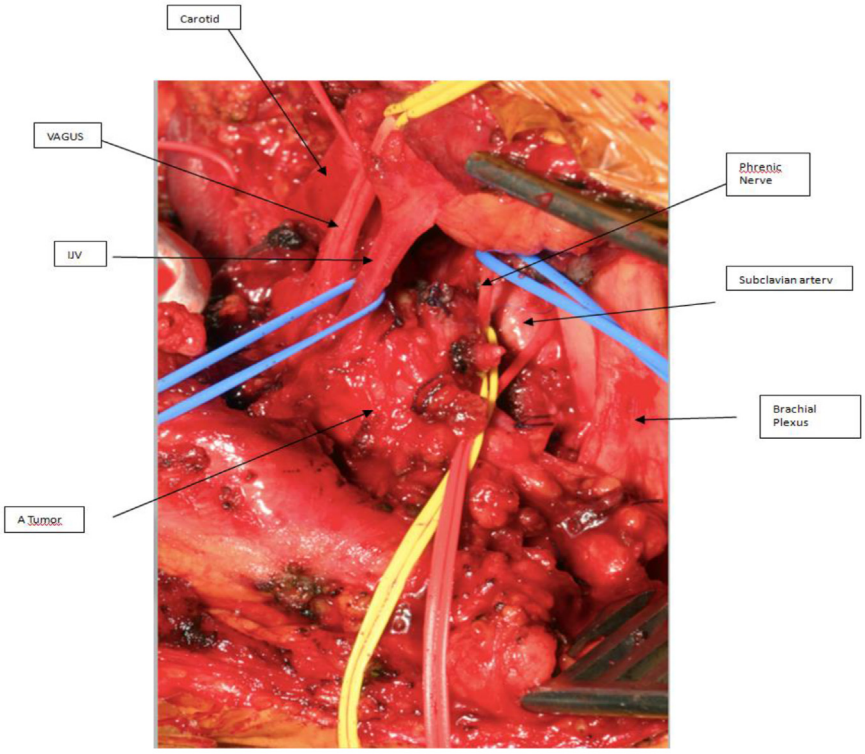

Fig 2. Complete exposure of the tumor and other vessels through trap-door exposure. *IJV*, internal jugular vein.

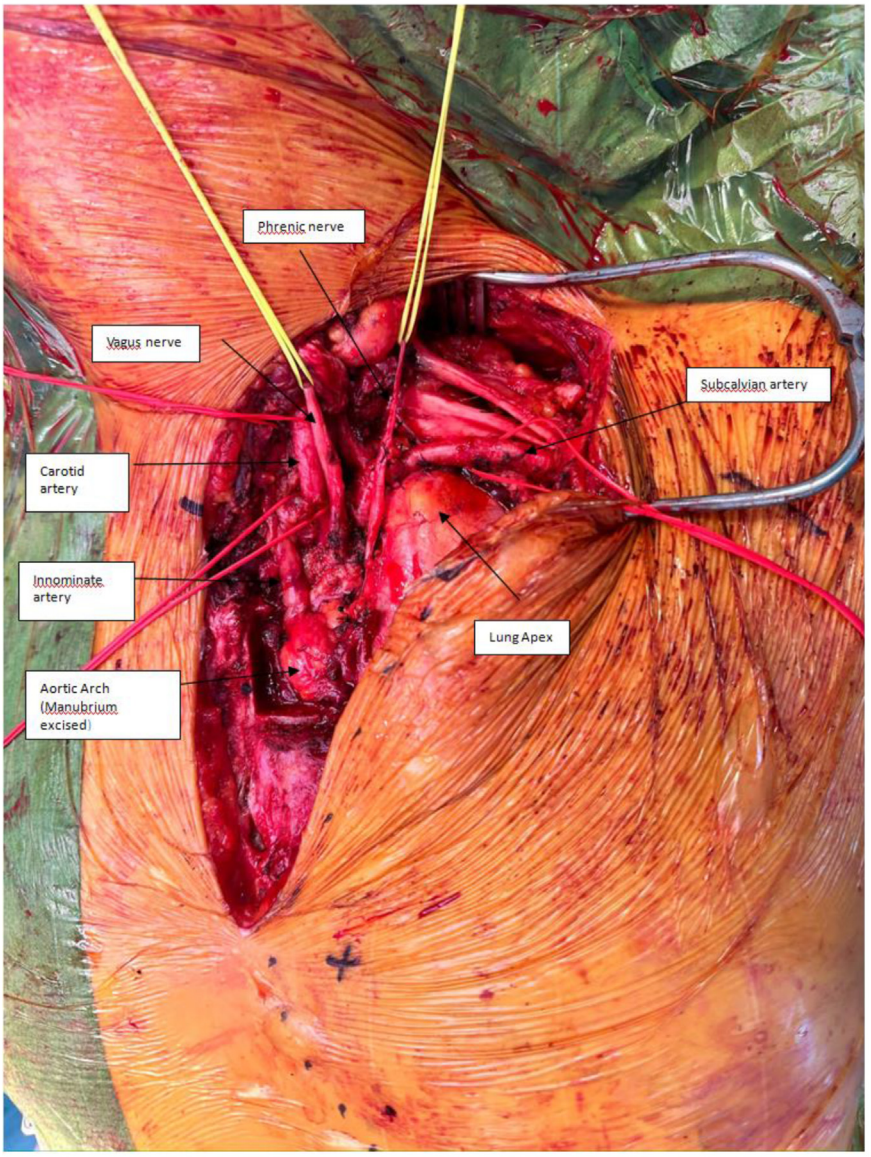

Fig 3. Postresection appearance.

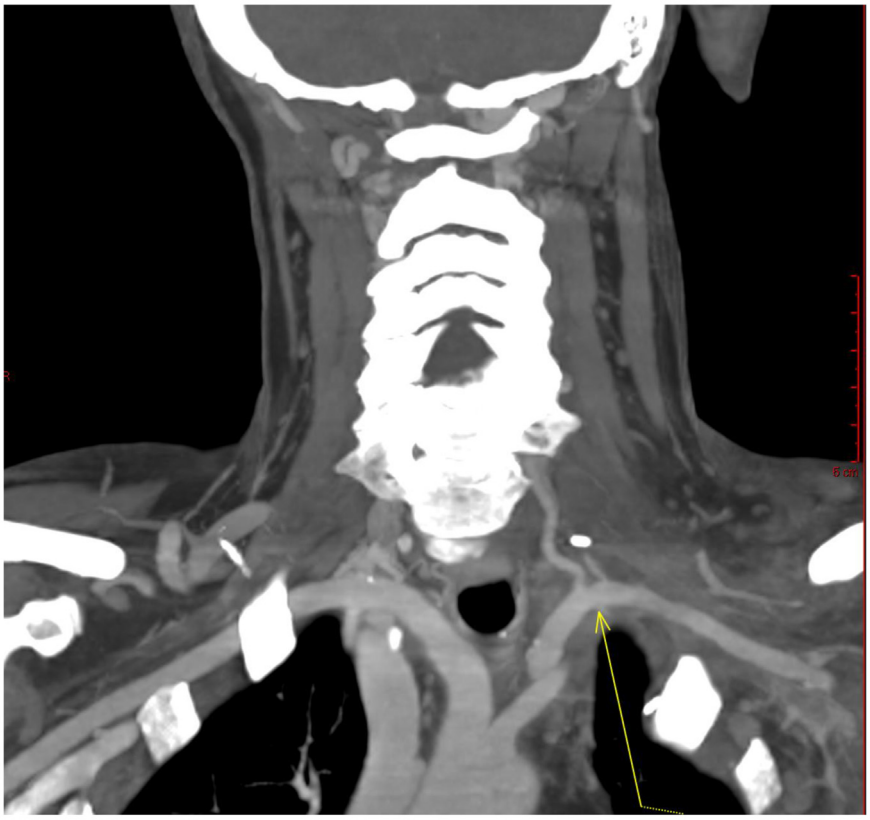

Fig 4. Postoperative surgical site after complete excision of the tumor.

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