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Open surgical treatment for rare epithelioid hemangioendothelioma of the common femoral vein

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ABSTRACT

Epithelioid hemangioendothelioma is a very rare disease affecting big blood vessels, especially veins. There have been very few articles describing the disease. We hereby present the case of a 56-year-old woman presenting with lower limb edema, who was initially being treated for residual thrombus in the common femoral vein but was eventually diagnosed with epithelioid hemangioendothelioma (EHE). The common femoral vein was resected and reconstructed using the external jugular vein. No additional therapy was administered. In this article, previous literature about EHE has been reviewed and oncologic principles have been discussed.

Introduction

It is a known fact that malignant tumors originating from large vessels are rare. These vascular tumors are mostly primary venous leiomyosarcomas while some of them are Epithelioid Hemangioendotheliomas (EHE), which arise from endothelial cells.[1] Intravascular EHE has more often been described in veins rather than in arteries. However, in reviewing English literature over the past few years, there has only been approximately 30 reports about this particular disease.[2] Most surgeons may never encounter this pathology throughout their career. Therefore, it is a clear challenge to make correct diagnostic, treatment, and management decisions pertaining to the disease. We hereby report and discuss a case of epithelioid hemangioendothelioma of the common femoral vein, where the patient presented with lower extremity edema.

Case report

A 56-year-old woman presenting with left extremity edema for six months prior to diagnosis was transferred to our department from another hospital. The edema was gradually progressive with diffuse leg pain and fatigue, which was more apparent during exercise. Ultrasonography did not detect anything but a focal non-compressible lesion in the common femoral vein. Ascending phlebography was then performed to exclude the possibility of deep vein thrombosis, and in the process, a localized endoluminal gap was found in the left femoral vein at the level of the inguinal ligament (Fig. 1). However, venous flow was not markedly affected by the filling defect. Computed tomography angiography (CTA) revealed a 2-cm long intraluminal mass, adherent to the dorsolateral vein wall at the proximity of the common femoral vein bifurcation. At first, it was considered to be a residual parietal thrombus formed after partial recanalization of venous thrombosis. Hence, an open surgical procedure was performed, and the mass was resected; the mass looked like mulberry grits, and since it was not fully adherent to the venous wall, the mass could easily be entirely dislodged from the endothelium. The walls of the common femoral vein and the great saphenous vein were thickened. There was no evident lymph node metastasis, and the common femoral artery was not involved. Since the frozen section diagnosis was not conclusive, the common femoral venous wall was closed, and low-molecular-weight heparin (LMWH) was initiated postoperatively on a daily basis. After the surgery, there was prompt relief in the swelling of the ipsilateral extremity.

One week later, permanent sections identified the mass as an EHE. Immunohistochemical stains confirmed the presence of CD34, CD31, and Ki-67 (2%,+) and the absence of CK, SMA, and CD68. Thus, a second surgical procedure was performed to resect the common femoral and great saphenous veins at a distance of 2 cm from the original position of the mass and replace them with a reversed external jugular vein graft (Fig. 2). Both resection margins were indicated by examination of the frozen section to ensure tumor-free margins, while the permanent section confirmed the absence of tumor cells in the margins of the resected area. Post-operatively, the patient was maintained on LMWH on a daily basis until the adequate therapeutic level of anticoagulants (warfarin) was

Abbreviations: EHE, Epithelioid Hemangioendothelioma.

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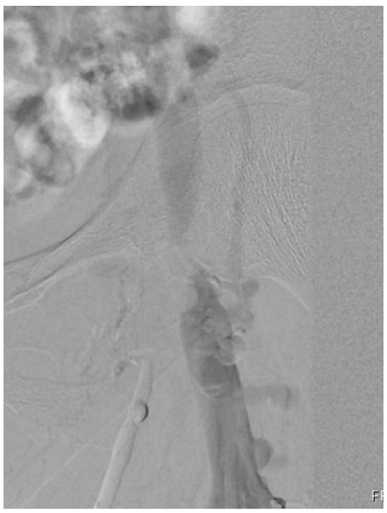

Fig. 1. Preoperative DSA found a severe stenosis with one big collateral of the left common femoral vein at the level of the inguinal ligament.

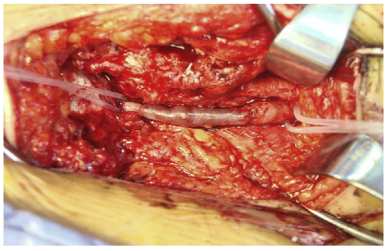

Fig. 2. Reconstruction of the CFV with external jugular vein graft.

reached. The patient was discharged with no visible edema in the left extremity, and oral anticoagulants (warfarin) and compression stockings were prescribed. No additional treatment such as radiation or chemotherapy was considered, and post-operative follow-up was maintained. Duplex study and ascending phlebography were conducted at the one-month, three-month, and six-month post-discharge follow-ups (Fig. 3), and the respective examinations displayed normal blood flow with no sign of restenosis at the site of anastomosis. A head-to-toe PET-CT scan was also performed one month after the surgery to exclude any sign of residual or recurrent tumor or metastasis.

Discussion

Epidemiologically, primary tumors of major blood vessels, mostly involving large veins, are distinctly uncommon.[1,3] Most of them are of smooth muscle origin, whereas only a few of them are of endothelial

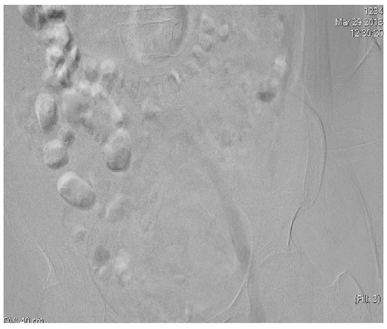

Fig. 3. DSA shows no stenosis and collaterals disappear at third month of follow-up.

origin. Leiomyomas represent nearly all reported cases of endothelial tumor while EHE, as a rare disease, accounts for a very small portion of those.[4] Hence, it is a big challenge for physicians to make correct diagnostic, treatment, and management decisions of the disease. Furthermore, it is impossible to differentiate between benign and malignant tumors on the operation table. As demonstrated in our case, frozen sections can prove to be uncertain when not examined or assessed by an experienced multidisciplinary pathologist. The histologic nuances of these extremely rare tumors frequently delay the final pathologic diagnosis and the radical tumor operation.

The term EHE was defined first by Weiss in 1982 as a soft tissue vascular tumor of endothelial origin, manifesting as a borderline or low-grade malignancy.[5] EHE is believed to be in the middle of the spectrum of endothelial tumors, located between the benign hemangioendothelioma and the malignant epithelioid angiosarcoma.[1–3] The tumor is usually more venous than arterial in origin (most frequently occurring in the femoral or iliac vein). EHE does not demonstrate any difference in the gender-specific trends and occurs in all age groups with a male to female sex predilection of 2:1, with the exception of the pediatric age group.[6]

The location of the lesions varies greatly from lower limb vein and inferior vena cava to superior vena cava.[2,7,8] It seldom occurs in the arterial system, e.g., in the popliteal artery presenting with distal blue toe syndrome, or in the radial artery presenting with a small, painless, oval mass in the right forearm.[3,9] Tumors localized in the proximity of larger vessels may produce symptoms of arterial occlusive disease or venous thrombosis due to compression of the larger vessels.[1] Thus, symptoms alter with respect to the location of the lesion. Symptoms such as extremity edema, leg fatigue, superior vena cava syndrome (SVCS), localized mass, and limb ischemia have been reported in past articles.[2–4,7–9]

Big vessel EHE, as a vascular tumor originating from the wall of larger vessels, protrudes intracellularly and does not manifest as a tumor mass. Conversely, it tends to get easily confused with other ordinary occlusive diseases such as venous thrombosis or arteriosclerotic occlusion. Therefore, upon clinical examination, big vessel EHE is not easily recognizable, and eventually this leads to failure in the identification of the true nature of the lesion when using classical vascular evaluations such as sonography, angiography, or even modern diagnostic techniques. However, the chronic medical history and unordinary location of the lesion, combined with the duplex findings as well as the normal architecture of the rest of the lower limb veins on ascending phlebography, can eventually indicate the possibility of a primary arterial tumor, even though the diagnosis is extremely rare.

Due to the borderline or low-grade malignant biological behavior of EHE, treatment options are usually frustrating. Currently, a consensus has been reached on the procedure for the tumor excision with wide surgical margins and tumor-free margins.[2,10,11] As for regional lymph node dissection, it is not included in the routine surgical plan. Schröder et al. did not recommend an extended lymph node dissection when they had observed micro-metastasis in one inguinal lymph node for two years, and there was no sign of local recurrence or metastasis.[1] Furthermore, John's most careful follow-up exams over an average of 48 months revealed a 13% local recurrence rate and distant metastasis in 31% of 46 patients.[10] The distant metastases were to regional lymph nodes, lung, liver, and bone. Less than half of all patients with metastases died of their disease, yet it was noticed that disease progression was easily controlled with local excision.[10] Other adjuvant therapies, such as chemotherapy and radiotherapy, have not proven to be beneficial.[2,12] Follow-up information on 31 cases, referred by Weiss et al. indicates that irrespective of the nature of the surgical procedure, whether it is simple excision or wide excision followed by radiation, there was no correlation with the clinical behavior of the disease.[5]

Moreover, there should be total awareness of the possibility that metastasis may occur 10 or more years after the first surgical procedure. Therefore, in addition to surgery, close follow-up care for the patient is a pre-requisite.

Declaration of competing interest

No Conflicts of Interest.

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