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Case Report

Chest wall epithelioid hemangioendothelioma unmasked by a hematoma: case report

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ABSTRACT

Epithelioid hemangioendothelioma (EHE) is a rare neoplasm originating from vascular endothelial cells, which can arise in varied tissue sites. We describe a case of EHE presented with submammary mass of the left chest wall in a 25-year-old male, and his chest CT scan revealed a hematoma. The patient underwent a resection of the mass and the pathologic examination confirmed the diagnosis of epithelioid hemangioendothelioma. Five months later, He developed a firm nodule at his surgical scar. He had a second surgery consisting of excision of the local recurrence and partial resection of the sixth rib, including the intercostal space of the chest wall. To cover the remaining defect, we used polypropylene mesh, with muscle advancement and primary skin closure. The patient subsequently received radiotherapy and remains under active surveillance. This case highlights the challenges of preoperative diagnosis and that EHE should be considered in the differential diagnosis of chest wall masses.

Keywords: epithelioid hemangioendothelioma, chest wall tumor, vascular tumor, soft tissues, surgical treatment

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Introduction

Epithelioid hemangioendothelioma (EHE) is an exceptionally rare vascular malignancy, representing less than 1% of all vascular tumors [1]. According to the 2020 World Health Organization (WHO) Classification of Soft Tissue Tumors, EHE is categorized among vascular sarcomas and exhibits highly heterogeneous clinical presentations, arising from sites such as bone, liver, soft tissues, or lung [2]. Chest wall involvement is extremely uncommon. We describe a case of chest wall EHE that developed local recurrence after R0 resection involving the ribs, managed with surgical excision followed by radiotherapy.

Case Report

A 25-year-old man presented with a left submammary mass that had been evolving for approximately one year. He underwent ultrasound-guided biopsies, but the histopathological findings were inconclusive. The patient denied any history of trauma and had no significant medical history, including no tobacco or alcohol use. On physical examination, a 5 cm mass was palpated in the left submammary region of the anterior chest wall. Preoperative thoracic computed tomography (CT) revealed a hematoma-like collection measuring 50×64×36 mm, with peripheral enhancement after contrast injection (Figure 1). Following multidisciplinary discussion, surgical management was decided, and the patient underwent en bloc resection including the mass and the pectoralis major muscle (without costal invasion). Histological examination of the surgical specimen showed a proliferation of endothelial cells arranged in small vascular channels. The tumor cells appeared large and epithelioid, with abundant cytoplasm, prominent nucleoli, and marked nuclear atypia. Mitotic activity was estimated at 6 per 10 high-power fields, with no necrosis identified. Immunohistochemistry demonstrated positive staining for CD31, EMA, and ERG, Ki-67 (approximately 20%), while CD34 was negative (Figure 2). These findings supported the diagnosis of epithelioid hemangioendothelioma with negative microscopic (R0) margins. As part of the metastatic workup, computed tomography (CT) scans of the head, neck, chest, abdomen, and pelvis were performed and showed no abnormalities.

Five months later, the patient developed a small, firm, fixed subcutaneous nodule on his surgical scar. A second surgery was performed consisting of excision of the tumor and partial resection of the sixth rib, including the intercostal chest wall, for definitive diagnosis and

treatment. The tumor was a smooth, soft mass located primarily within the intercostal soft tissues and adherent to the sixth rib, but without lung involvement. The chest wall defect was reconstructed using polypropylene mesh, and the wound was closed by dissecting and advancing by three muscles, the latissimus dorsi, pectoralis minor, and rectus abdominis. Given the good skin elasticity, primary closure of the incision was achieved (Figure 3). The postoperative course was uneventful, and the patient was discharged on postoperative day 3. Pathological examination revealed local recurrence with aggressive features, including focal tumor necrosis and a mitotic count of 14 per 10 high-power fields. As the recurrence developed five months after the initial surgery, the patient underwent radiotherapy and remains under active surveillance. The patients gave written informed consent to collect, analyze, and publish his medical data in this study.



Figure 1. A computed tomographic (CT) scan of the chest showing a hematoma-like collection with peripheral contrast enhancement.

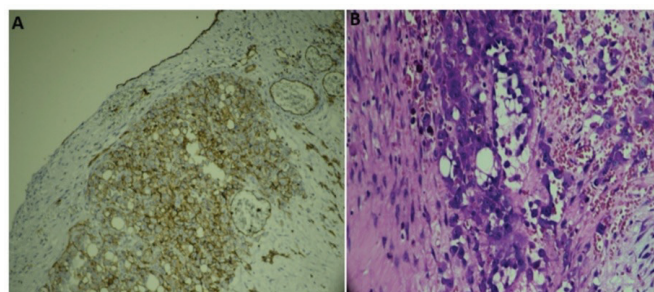


Figure 2. Histopathological features of epithelioid hemangioendothelioma: Proliferation of vascular structures and large epithelioid cell sheets with marked anisokaryosis and vesicular nuclei (A), nuclear staining of tumor cells with anti-CD31 antibody (magnification ×20) (B).

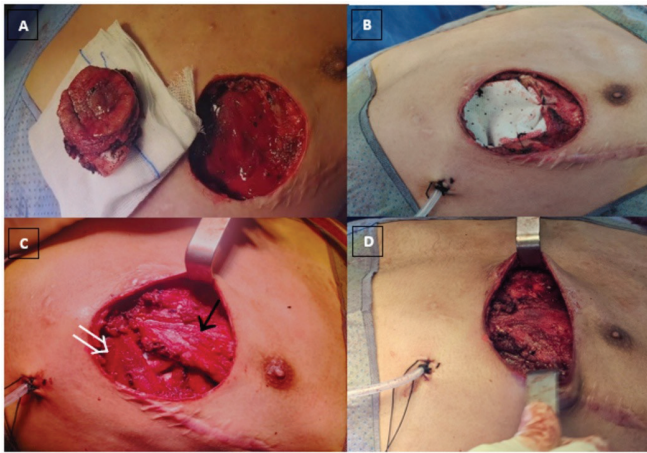


Figure 3. Clinical images of the representative case. Defect after excision of the local recurrence with the mass resected (A), intraoperative view after placement of a polypropylene mesh to close the defect (B), closure achieved by dissecting and advancing three muscles: the latissimus dorsi (black arrow), pectoralis minor, and rectus abdominis (white arrow) (C), final appearance after reconstruction and closure of the defect (D).

Discussion

Epithelioid hemangioendothelioma (EHE) is an ultra-rare vascular sarcoma with two distinct morphological and molecular variants (CAMTA1-related and TFE3-related). Its rarity, with an incidence of 0.038 per 100,000 per year, accounts for the limited literature, which is primarily composed of case reports and small case series showing variations in demographic characteristics [3]. The median age at diagnosis is typically in the fourth to fifth decades of life [1-4]. However, in our case, the patient was young.

EHE can occur in any part of the body and may present as a unifocal lesion, with locoregional metastases, or with systemic metastases [5]. Furthermore, intrathoracic EHE usually presents as bilateral pulmonary nodules, and to our knowledge, this case represents the second reported case of chest wall EHE. The clinical presentation at diagnosis is highly variable; patients may exhibit respiratory symptoms such as pleuritic chest pain, pleural effusion, or hemoptysis, although some cases may remain asymptomatic [6]. A review of the literature reveals that chest wall involvement is very uncommon [7]. In the present case, we initially suspected a spontaneous hematoma because of the negative result of the needle biopsy.

Epithelioid hemangioendothelioma of the soft tissues may arise in either supra- or sub-aponeurotic compart-

ments. These tumors frequently develop in proximity to blood vessels, reported in 50–70% of cases, and can sometimes cause vascular lumen obstruction. Magnetic resonance imaging (MRI) is recommended for the primary soft tissue disease, it presents as heterogeneous mass close, particularly following the administration of contrast agents, and may include calcifications, spontaneous hemorrhages, peripheral edema, or bone erosions [8].

EHE is diagnosed based on histological, immunohistochemical, and molecular features [5]. Histologically, the tumor is composed of nests and cords of epithelioid endothelial cells, occasionally admixed with spindle cells. Cytological features of EHE include cells with moderate to abundant cytoplasm and pleomorphic nuclei with frequent intranuclear pseudoinclusions [5]. Approximately 10% of cases demonstrate marked nuclear atypia with prominent nucleoli, areas of necrosis, and elevated mitotic activity (>2 mitoses per 10 high-power fields); these features are associated with more aggressive clinical behavior, which was the case in our patient and may explain the recurrence despite an R0 resection at the first operation [1]. Immunohistochemically, EHE typically expresses vascular markers such as Fli-1, CD31, ERG, and CD34 [5].

The treatment of choice for confirmed unifocal epithelioid hemangioendothelioma (EHE) is complete surgical excision. Resection should be performed in specialized referral centers with expertise in sarcoma surgery [7]. The tumor must be resected en bloc, including the biopsy tract, with microscopic negative (R0) margins, achieving a cure rate of approximately 70–80%. The risk of local recurrence remains in the range of 10–15%. The tumor is considered relatively radiosensitive, although the role of radiotherapy (RT) is not well established. Adjuvant RT is recommended in selected cases, particularly when margins are close or positive and the risk of local recurrence is significant [1], which was indicated in our case. Systemic treatment is not routinely indicated for localized disease. In contrast, in metastatic settings with unequivocal progression, symptom aggravation, or organ impairment, systemic therapy may be considered, although no established standard of care currently exists [1].

In conclusion, chest wall epithelioid hemangioendothelioma represents a rare entity with unpredictable behavior. Complete surgical resection remains the cornerstone of treatment when feasible, even in the setting

of local recurrence, which highlights the crucial role of long-term surveillance.

Declaration of conflicting interests

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Authors' contribution

I.A.: Conceptualization, data curation, investigation, software, project administration, visualization, writing - original draft, writing - review & editing, H.H.: Conceptualization, formal analysis, methodology, project administration, visualization, writing - original draft, writing - review & editing, M.L.: Funding acquisition, resources, supervision, validation, writing - review & editing, Y.O.: Funding acquisition, resources, supervision, validation, writing - review & editing, M.S.: Funding acquisition, supervision, validation, writing - review & editing.

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