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Epithelioid Hemangioendothelioma of the Inguinoscrotal Soft Tissue: A Rare Case Report

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Abstract

Epithelioid hemangioendothelioma (EHE) is a rare vascular neoplasm of endothelial origin with intermediate malignant potential and variable clinical behavior. Soft tissue involvement of the inguinoscrotal region is exceptionally uncommon and may clinically resemble benign surgical conditions, leading to diagnostic difficulty. We report a case of a 61-year-old male presenting with a right-sided inguinoscrotal swelling clinically suspected to be a non-reducible inguinoscrotal hernia. Histopathological examination revealed epithelioid endothelial cells arranged in cords and sheets within a fibrovascular stroma. Immunohistochemistry showed diffuse CD34 positivity along with strong nuclear TFE3 expression, supporting the diagnosis of YAP1–TFE3-associated EHE. The case highlights the importance of considering uncommon neoplasms in atypical inguinoscrotal masses and emphasizes the role of histopathology and immunohistochemistry in accurate diagnosis. Complete surgical excision and long-term follow-up remain essential because of the unpredictable biological behavior of this tumor.

Keywords: Epithelioid Hemangioendothelioma, Inguinoscrotal Soft Tissue Tumor, Vascular Neoplasm, TFE3-Positive Tumor, Immunohistochemistry

Introduction

Epithelioid hemangioendothelioma (EHE) is a rare malignant vascular neoplasm arising from endothelial cells and accounting for less than 1% of all vascular tumors [1,2]. It is classified as a tumor of intermediate malignancy [1,3] because of its biologic behavior, which lies between benign hemangioma and high-grade angiosarcoma. The tumor can occur over a wide age range but is more frequently reported in middle-aged adults. Although EHE may involve almost any anatomical site, the liver, lungs, bone, and deep soft tissues are the most commonly affected organs [3,12].

Occurrence of soft tissue EHE in the inguinoscrotal region is extremely uncommon. Such unusual anatomical presentations may clinically mimic common benign conditions such as inguinal hernia, hydrocele, epididymal cyst, lipoma, or paratesticular tumors, thereby delaying accurate diagnosis. Imaging studies often provide nonspecific findings, making histopathological

examination and immunohistochemistry indispensable for definitive diagnosis [2,10].

Recent advances in molecular pathology have improved the understanding of EHE. Most conventional EHEs demonstrate a characteristic WWTR1–CAMTA1 gene fusion [4], whereas a smaller subset shows YAP1–TFE3 fusion [5]. The TFE3-positive subset tends to display distinct histological features and may possess different biological behavior [5].

Due to the rarity of the tumor and its variable clinical course, the diagnosis and management of EHE remain challenging. We present a rare case of epithelioid hemangioendothelioma occurring in the inguinoscrotal soft tissue of a 61-year-old male, initially diagnosed clinically as an inguinoscrotal hernia.

Case History

Clinical Presentation

A 61-year-old male presented to the surgical outpatient department with complaints of swelling in the right scrotal region for one month. The swelling was insidious in onset and progressively increased in size. There was no associated history of pain, fever, trauma, weight loss, or constitutional symptoms. The patient did not report urinary complaints or previous similar swellings.

On clinical examination, a right-sided inguinoscrotal swelling was noted. The swelling was firm in consistency and non-reducible. Initial clinical suspicion favored an inguinoscrotal hernia. Ultrasonography of the scrotum revealed a large soft tissue lesion resembling omental tissue with internal vascularity, suggestive of a non-reducible omental inguinoscrotal hernia.

The patient subsequently underwent surgical excision of the mass. Gross and microscopic examination along with immunohistochemistry were performed for definitive diagnosis.

Pathological Findings

Gross Examination: Received was a single encapsulated globular soft tissue mass measuring 9 × 7 × 4 cm. The external surface was smooth with intact capsule and no areas of capsular breach. Cut section showed a predominantly solid yellowish tumor with soft to firm consistency and focal hemorrhagic areas. No obvious necrosis or cystic degeneration was identified.

Figure 1: Gross photograph showing a well encapsulated mass with yellow to brown, solid cut surface with focal hemorrhagic areas

Microscopy: Histopathological examination revealed a neoplasm composed of epithelioid to oval spindle-shaped cells arranged in cords, nests, and diffuse sheets embedded within a fibrovascular stroma. Tumor cells possessed moderate eosinophilic cytoplasm, vesicular chromatin, and inconspicuous nucleoli. Areas of hemorrhage and entrapped adipose tissue were seen. No significant pleomorphism, tumor necrosis, or vascular invasion was identified in the examined sections. Mitotic activity was low.

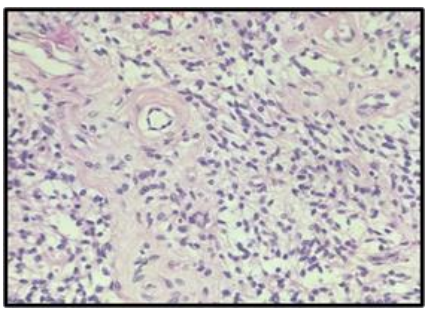

Figure 2: Microphotography showing epithelioid to oval spindle cells arranged in diffuse sheets embedded within a fibrovascular stroma (H&E 10x)

Epithelioid to oval cells with
Vesicular chromatin and moderate
eosinophilic cytoplasm.

Immunohistochemistry: Immunohistochemical analysis demonstrated strong positivity for CD34, confirming endothelial differentiation. Tumor cells also showed nuclear positivity for TFE3, supporting the diagnosis of the YAP1–TFE3 subset of epithelioid hemangioendothelioma. The tumor cells were negative for PanCK, STAT6, and HMB45, thereby excluding epithelial tumors, solitary fibrous tumor, and melanocytic neoplasms respectively.

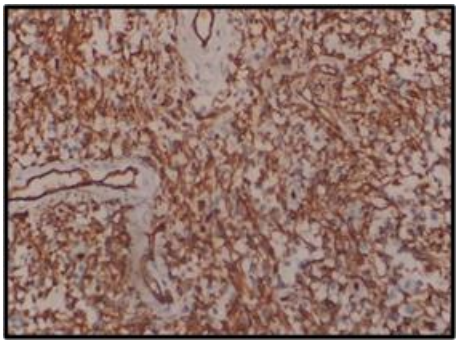

Figure 3: CD34 immunostain showing strong nuclear positivity in the tumor cells.

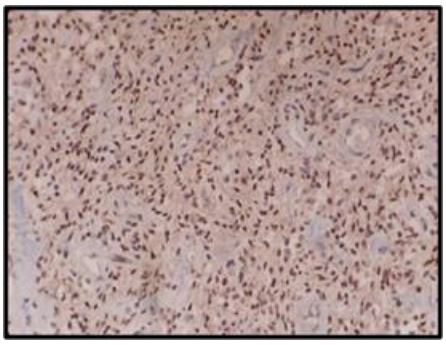

Figure 4: TFE3 immunostain showing strong nuclear positivity in the tumor cells.

Discussion

Epithelioid hemangioendothelioma (EHE) is a rare endothelial neoplasm characterized by an intermediate clinical behavior between benign hemangioma and aggressive angiosarcoma [1,2]. Since its original description by Weiss and Enzinger, EHE has remained diagnostically challenging because of its rarity, broad anatomical distribution, and overlapping histomorphological features with other epithelioid tumors [2]. The estimated incidence is less than one case per million population, with the liver, lungs, bone, and deep soft tissue representing the most common sites of involvement[3].

Involvement of the inguinoscrotal soft tissue is exceptionally uncommon, and only limited cases have been described in literature. Such tumors often mimic benign surgical conditions including inguinal hernia, hydrocele, lipoma, epididymal cyst, or paratesticular lesions. In the present case, the lesion was clinically and radiologically interpreted as a non-reducible inguinoscrotal hernia because of its soft tissue appearance and internal vascularity on ultrasonography. This highlights the nonspecific radiological features of EHE and the importance of histopathological examination for definitive diagnosis.

Histologically, EHE is composed of epithelioid endothelial cells arranged in nests, cords, or short strands embedded in a myxohyaline or fibrous stroma [1]. Intracytoplasmic vacuoles corresponding to primitive vascular lumina may occasionally contain erythrocytes and are considered a useful diagnostic clue. In our case, the tumor demonstrated epithelioid to spindle-shaped cells with eosinophilic cytoplasm and vesicular nuclei arranged in cords and sheets within a fibrovascular stroma. Absence of marked pleomorphism, necrosis, and brisk mitotic activity favored a lower-grade lesion. The differential diagnosis of inguinoscrotal EHE is extensive and includes both benign and malignant neoplasms. Adenomatoid tumor, the most common benign paratesticular tumor, may show epithelioid morphology but usually expresses mesothelial markers such as calretinin and WT1. Angiosarcoma demonstrates infiltrative growth, marked cytologic atypia, frequent

mitoses, and extensive necrosis. Solitary fibrous tumor may show spindle-cell morphology but typically exhibits STAT6 positivity due to NAB2–STAT6 fusion. Metastatic carcinoma and melanoma also enter the differential diagnosis in epithelioid tumors of unusual locations. In the present case, negativity for PanCK, HMB45, and STAT6 helped exclude these entities. Immunohistochemistry plays a pivotal role in establishing the diagnosis of EHE. Endothelial markers including CD31, CD34, ERG, and FLI1 are typically expressed [1,3]. CD31 is considered highly specific for endothelial differentiation, whereas CD34 is highly sensitive. Our case showed diffuse CD34 positivity, confirming vascular endothelial origin. Strong nuclear positivity for TFE3 further supported the diagnosis of the YAP1–TFE3 molecular subtype of EHE[5].

Recent molecular studies have significantly improved the understanding of EHE pathogenesis. Most conventional EHEs harbor the WWTR1–CAMTA1 fusion resulting from t(1;3)(p36;q25) translocation [4]. A smaller subset demonstrates YAP1–TFE3 fusion, which is associated with strong TFE3 immunoreactivity [5]. TFE3-positive EHEs often occur in younger patients and tend to display solid architecture, abundant eosinophilic cytoplasm, and more obvious vasoformative channels. Some reports suggest that this subset may exhibit a relatively favorable prognosis compared with conventional EHE, although further studies are required because of the rarity of the tumor.

The biological behavior of EHE remains unpredictable [6]. While some lesions pursue an indolent clinical course, others demonstrate aggressive behavior with recurrence or distant metastasis. Adverse prognostic factors include tumor size greater than 3 cm, high mitotic activity, multifocal disease, marked nuclear atypia, and tumor necrosis [6]. Although the tumor in our case measured more than 3 cm, the absence of marked atypia, necrosis, and increased mitotic activity may indicate a comparatively favorable prognosis.

Surgical excision with negative margins remains the treatment of choice for localized disease [6]. Due to the rarity of EHE, standardized treatment protocols are lacking. Chemotherapy and radiotherapy have shown variable response rates, whereas targeted and anti-angiogenic therapies are currently under investigation for advanced or metastatic disease [12]. Long-term follow-up is essential because recurrence and metastasis may occur years after initial treatment[6].

This case highlights the importance of considering rare vascular neoplasms in atypical inguinoscrotal masses. Awareness among clinicians, radiologists, surgeons, and pathologists is essential to prevent diagnostic delay and ensure appropriate management.

Conclusion

Epithelioid hemangioendothelioma is a rare vascular neoplasm with intermediate malignant potential [1,2] and unpredictable behavior. Inguinoscrotal soft tissue involvement is exceptionally uncommon and may mimic benign surgical conditions, resulting in diagnostic difficulty. Histopathology supported by immunohistochemistry is essential for accurate diagnosis. Complete surgical excision and long-term follow-up remain important because of the possibility of recurrence or metastasis.

References

1. WHO Classification of Tumours Editorial Board. Soft Tissue and Bone Tumours. 5th Edition. Lyon: IARC Press; 2020.
2. Sardaro A, Bardoscia L, Petruzzelli MF, Portaluri M. Epithelioid hemangioendothelioma: an overview and update on a rare vascular tumor. Oncol Rev. 2014;8(2):259.

3. Weiss SW, Goldblum JR. Enzinger and Weiss's Soft Tissue Tumors. 7th Edition. Elsevier; 2020.
4. Errani C, Zhang L, Sung YS, et al. A novel WWTR1–CAMTA1 gene fusion is a consistent abnormality in epithelioid hemangioendothelioma of different anatomic sites. Genes Chromosomes Cancer. 2011;50(8):644–653.
5. Antonescu CR, Le Loarer F, Mosquera JM, et al. Novel YAP1–TFE3 fusion defines a distinct subset of epithelioid hemangioendothelioma. Genes Chromosomes Cancer. 2013;52(8):775–784.
6. Deyrup AT, Tighiouart M, Montag AG, Weiss SW. Epithelioid hemangioendothelioma of soft tissue: a proposal for risk stratification based on 49 cases. Am J Surg Pathol. 2008;32(6):924–927.
7. Weiss SW, Enzinger FM. Epithelioid hemangioendothelioma: a vascular tumor often mistaken for a carcinoma. Cancer. 1982;50(5):970–981.
8. Fletcher CDM, Bridge JA, Hogendoorn P, Mertens F. WHO Classification of Tumours of Soft Tissue and Bone. 4th Edition. IARC Press; 2013.
9. Rosenberg A. WHO Classification of Soft Tissue and Bone, fourth edition: summary and commentary. Curr Opin Oncol. 2013;25(5):571–573.
10. Shon W, Billings SD. Cutaneous vascular tumors: update on classification and diagnosis. Surg Pathol Clin. 2017;10(2):327–362.
11. Mentzel T, Beham A, Calonje E, Katenkamp D, Fletcher CD. Epithelioid hemangioendothelioma of skin and soft tissues: clinicopathologic and immunohistochemical study. Am J Dermatopathol. 1997;19(6):527–533.
12. Lau K, Massad M, Pollak C, et al. Clinical patterns and outcome in epithelioid hemangioendothelioma with or without pulmonary involvement. Chest. 2011;140(5):1312–1318.