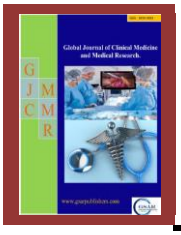

Global Journal of Clinical Medicine and Medical Research [GJCMMR]
ISSN: 2583-987X (Online)
Abbreviated key title: Glob.J.Clinic.Medici.Medica.Res.
Frequency: Monthly
Published By GSAR Publishers
Journal Homepage Link- https://gsarpublishers.com/journal-gjcmmr-home/

Epithelioid Hemangio Endothelioma, an ultra-rare tumor.

By

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Article History

Received: 05/06/2025
Accepted: 14/06/2025
Published: 16/06/2025

Vol – 3 Issue – 6

PP: -10-11

Abstract

Epithelioid hemangioendothelioma (EHE) is a very rare vascular tumor. It exhibits intermediate malignancy between hemangioma and angiosarcoma and originates from endothelial cells lining of the blood vessels. EHE is notable for its distinctive epithelioid endothelial cell morphology. Its mainly present in soft tissues or any organ.The disease commonly presents in young adults with a slight female predominance. Symptoms vary, mostly asymptomatic, but can present in aggressive forms with metastasis, often manifesting as pain or unexplained weight loss. WWTR1(TAZ)-CAMTA1 gene fusion is seen in over 90% of cases. Diagnosis is challenging due to its rarity and requires histological and immunohistochemical analysis, including markers like CD31, ERG, and CD34. There is no standardized treatment, with surgery, targeted therapy, chemotherapy, and radiation being used variably. Prognosis depends on disease spread, with survival ranging from months to decades. Though various chemo regimens have been tried post-surgery in literature, for her we are using VEGF inhibitor Bevacizumab as the tumor is of endothelial origin. A post-chemotherapy status with a one-year follow-up will reveal its efficacy.

Keywords: *WWTR1(TAZ)-CAMTA1, Bevacizumab, VEGF INHIBITOR, CD31, CD34*

Introduction

Epithelioid hemangioendothelioma (EHE) is a rare vascular tumor with only one in every one million people diagnosed with this cancer worldwide[(1)]. It is characterized by an intermediate malignancy between hemangioma and angiosarcoma [(2)] arising from cells lining the blood vessels. It is more likely to affect soft tissues, but can also involve organs such as the liver, lungs, and bones[(3)] . EHE is notable for its distinctive epithelioid endothelial cell morphology and a tendency to present in young adults, with a slight female predominance. Regarding signs and symptoms, it varies widely, ranging from asymptomatic lesions to more aggressive disease with metastasis. EHE is mostly asymptomatic. Pain is the most common presenting symptom. A mass beneath the skin or unexplained weight loss can also be the presenting feature[(10)] . Researchers have discovered that EHE is caused by a unique genetic mutation, which is TAZ dysregulation [(4)]. EHE has a *WWTR1*(TAZ)-*CAMTA1* gene fusion in >90% of cases, 45% of which have no other genetic alterations [(5)]. Due to its rarity and variable behavior, diagnosis can be challenging, often requiring histological examination and immune-histochemical analysis [(6)]. EHE consistently expresses endothelial differentiation markers such as CD31, ERG, CD34, and FLI-1[(7)]. EHE is a very rare tumor, there are no standard treatment guidelines. Surgical intervention usually is the primary treatment modality, and targeted therapy may be of some benefit, chemotherapy and radiation therapy also can be used in certain situations [(8)] .Regarding the prognosis of the disease, Although localized EHE can be surgically resected, there is no effective therapy for systemic disease, and mortality from cancer ranges between 13% and 18% when confined to soft tissue, but life expectancy in metastatic cases is unpredictable and ranges from a couple of months to 15 to 20 years[(9)].

Case description

A 67-year-old female patient presented to the clinic with right wrist and hand pain for two months duration. The pain was burning in nature, gradual in onset, persistent, and with no relieving or aggravating factors. There were no associated skin color changes. The joint movement was normal. She had no history of smoking and denied any use of chronic medication. On examination, she looked well. Elbow and

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wrist joints moved freely and the pulses were normal. There was a horizontal surgical scar on the volar surface of the right wrist (about 3 cm in length), as her condition was mistaken as carpel tunnel syndrome. She had carpel tunnel release procedure without any improvement. CT scan and PET scan were done which revealed ill-defined hypodense changes with heterogeneous FDG uptake (SUVmax 5.9) noted in right lower arm muscular plane. The patient was referred for surgery underwent right arm soft tissue tumor excision and sent for histopathology. Histopathology results revealed a tumor composed of sheets and cords of large atypical cells with epitheloid morphology, polygonal cell outlines, round vesicular nuclei, and prominent nucleoli. The tumor was infiltrating surrounding soft tissue, blood vessels, and nerve bundles . Features suggestive of Epithelioid hemangioendothelioma. The IHC results were positive for CK, CD34, ERG, and CD31 with Ki67 of 35-40%. A plan of chemotherapy was done for her with Intravenous "Bevacizumab" and she had started with the first dose (Bevacizumab 700 mg iv every three weeks) waiting for further assessment.

Conclusion:

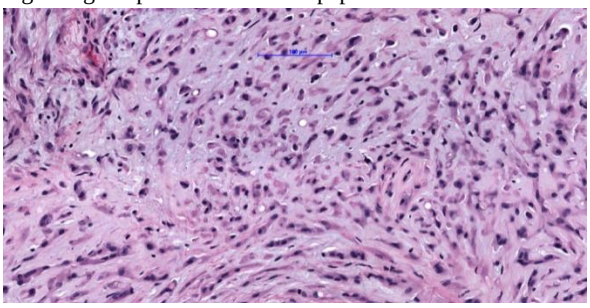
EHE is an ultra rare malignancy with incidence of one person in a population of one million .Though various chemo regimens have been tried post surgery in literature , for her we are using VEGF inhibitor Bevacizumab as the tumour is of endothelial origin. A post chemotherapy status with a one year follow up will reveal its efficacy.

Disclosure:

This study was conducted at Al-Thaqalyn Hospital Cancer Institute, Basra, Iraq.

Conflicts of Interest:

The authors declare that there is no conflict of interest regarding the publication of this paper



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