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Case Report

Hemangioendothelioma of the Forehead – A Rare Case Report

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Abstract

Hemangioendothelioma is the term used to name those vascular neoplasms that show a borderline biological behavior, intermediate between entirely benign hemangiomas and highly malignant angiosarcomas. Although originally spindle cell hemangioendothelioma was proposed as a specific clinicopathologic variant of hemangioendothelioma, currently, it is considered as an entirely benign lesion, and thus, the name spindle cell hemangioma seems to be the most accurate for this lesion. Authentic hemangioendotheliomas involving the skin and soft tissues include Papillary intralymphatic angioendothelioma (also known as Dabska tumor), Retiform hemangioendothelioma, Kaposiform hemangioendothelioma, Epithelioid hemangioendothelioma, Pseudomyogenic hemangioendothelioma (also known as epithelioid sarcoma-like hemangioendothelioma), and Composite hemangioendothelioma. Each of these neoplasms exhibit characteristic histopathologic features.

Keywords: Hemangioendothelioma, rare, vascular, soft tissue.

INTRODUCTION

Hemangioendothelioma is a rare vascular tumor of endothelial nature that occupies an intermediate position between benign hemangiomas and highly malignant angiosarcoma, characterized by proliferating neoplastic endothelial cells [1].

Hemangioendothelioma commonly occurs in the superficial or deep soft tissue of the extremities, lungs, liver, bone and lymph nodes, with oral cavity being a rare location. It is usually benign but can show variable grades of malignancy. The tumor cells may form small intracellular lumen, which may be seen as clear spaces, or vacuoles, that distort (or blister) the cell. Lesions that arise from vessels may expand the vessel, usually preserving its architecture and extend centrifugally from the lumen to the soft tissue. Hemangioendothelioma has good prognosis and is treated surgically and/or by chemotherapy/radiation [2]. Hemangioendothelioma is capable of local recurrence and metastasis albeit at a lower rate as compared to malignant neoplasm. Clinically, it can mimic reactive lesions such as pyogenic granuloma, chronic periodontal disease and peripheral giant cell granuloma.

CASE REPORT

A 39 year old male presented with a freely mobile, hard, gradually progressive swelling measuring 1x1cm over left side of the forehead.

Gross Findings: Received single globular soft tissue mass measuring 1x1cm.

Cut Section

Homogenous, grey-white Tissue was all embedded and after serial tissue processing, sections were ready and microscopically reported as hemangioendothelioma.

Histopathological Examination

Histological examination showed tumor composed of plump, oval to spindle shaped cells with pale eosinophilic nucleoli arranged in nodular pattern. These cells are arranged around central lumen showing few RBCs in some foci. Lymphocytic infiltrate is seen around nodular areas, suggestive of hemangioendothelioma [Fig 1- 4].

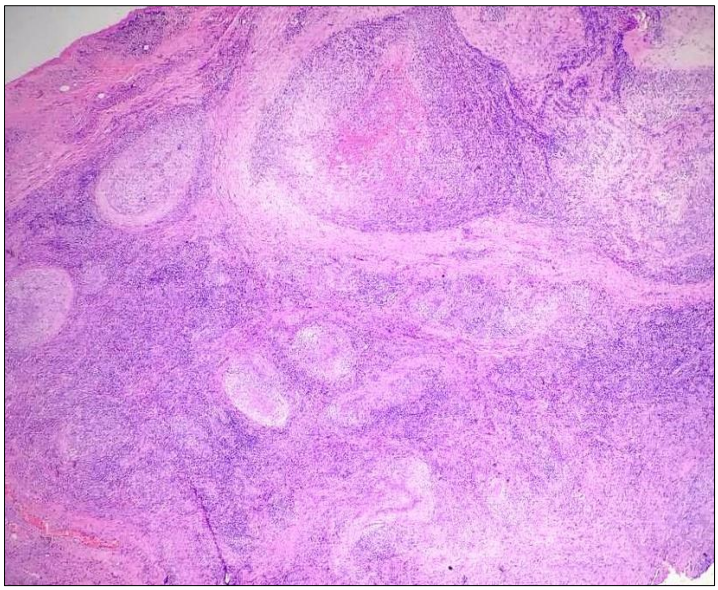

Figure 1: H & E, 4x Biopsy Showing Tumor Arranged In Nodular Pattern

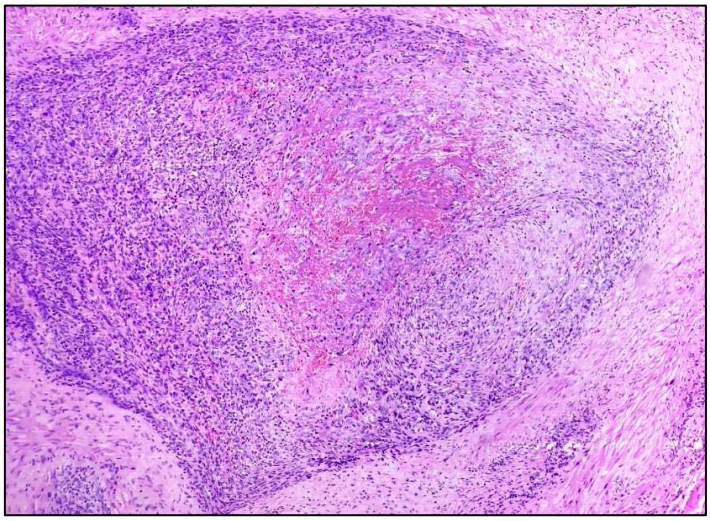

Figure 2: H & E, 10x Vasculocentric Growth Composed Of Proliferating Neoplastic Endothelial Cells Expanding The Vessel Wall

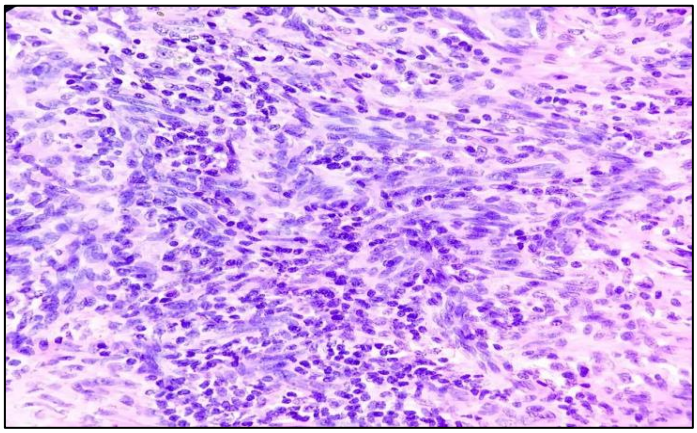

Figure 3: H & E, 40x High Power Showing Plump, Oval to Spindle Shaped Tumor Cells Arranged in Interlacing Fasicles

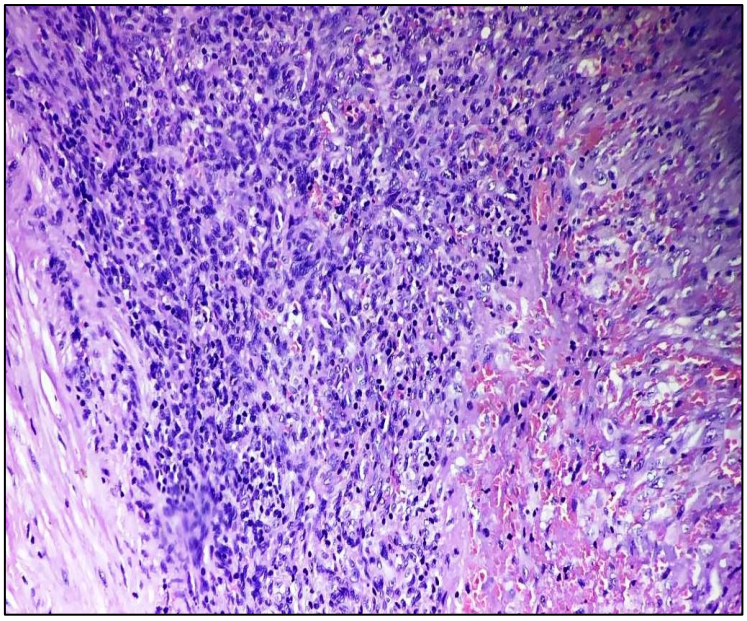

Figure 4: H & E, 40x Section Showing Lymphocytic Infiltrate around the Vessel Wall in Nodular Areas

DISCUSSION

Hemangioendotheliomas are vascular neoplasms seen commonly in soft tissue of the extremities and bone. Head and neck is an uncommon site. Diagnosis is made with biopsy and histopathologic features include angiocentric growth composed of proliferating neoplastic endothelial cells expanding the vessel wall, often with surrounding sclerosis Based on the histological presentation, they can be classified as Retiform, Epithelioid, Hobnail, Kaposiform, Epithelioid-sarcoma like, Composite, etc [3].

The most characteristic finding of papillary intralymphatic hemangioendothelioma consists of papillary tufts, with a central hyaline core lined by hobnail-like endothelial cells protruding into the lumina [4].

Retiform hemangioendothelioma is an infiltrative neoplasm composed of elongated arborizing vessels, arranged in an anastomosing pattern that resembles that of the rete testis, and lined by a single layer of hobnail-like endothelial cells that protrude within the narrow lumina. Kaposiform hemangioendothelioma is composed of several solid poorly circumscribed nodules, and each nodule is composed of a mixture of small capillaries and solid lobules of endothelial cells arranged in a glomeruloid pattern. A frequent finding consists of the presence of areas of lymphangiomatosis adjacent to the solid nodules [5, 6].

Epithelioid hemangioendothelioma is composed of cords, strands, and solid aggregates of round, oval, and polygonal cells, with abundant pale eosinophilic cytoplasm, vesicular nuclei, and inconspicuous nucleoli, embedded in a fibromyxoid or sclerotic stroma. Many neoplastic cells exhibit prominent cytoplasmic vacuolization as an expression of primitive vascular differentiation. Pseudomyogenic hemangioendothelioma is a poorly circumscribed, fascicular lesion with infiltrative borders composed of round or oval neoplastic cells, with vesicular nuclei and inconspicuous nucleoli, and ample homogeneous eosinophilic cytoplasm, giving them a rhabdomyoblastic appearance [7].

Finally, Composite hemangioendothelioma is the term used to name locally aggressive vascular neoplasms of low-grade malignancy showing varying combinations of benign, low-grade malignant, and high-grade malignant vascular components. From the immunohistochemical point of view, proliferating cells of all hemangioendotheliomas express a lymphatic endothelial cell immunophenotype.

Most hemangioendotheliomas are low-grade vascular neoplasms, with a tendency to recur locally and a low metastatic potential, mostly to regional lymph nodes. Treatment options include surgical excision and/or chemotherapy/radiation. Utility of FISH and RT-PCR in detecting the fusion-genes and immunohistochemistry may add to the diagnostic value. Prognosis is usually good but may vary based on site, local aggressiveness, etc. Epithelioid hemangioendothelioma, especially large lesions and those located in deep soft tissues, seems to have a more aggressive biological behavior [8].

CONCLUSION

Hemangioendotheliomas are uncommon borderline vascular tumors with high chances of local recurrence. Rarity of the tumor combined with wide age distribution at presentation, the variety of anatomic sites,

local aggressiveness, metastases and multifocality lead to a wide range of differential diagnoses and affect the prognosis. Complete surgical excision is the primary stay of treatment.

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