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Kaposiform Hemangioendothelioma in an Adult- A Case Report

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Abstracts: ***Kaposiform Hemangioendothelioma is a rare vascular neoplasm that previously has been described in infancy and early childhood. It has rarely been observed in head and neck region. We report a case of nineteen year-old female who developed a lesion in superficial soft tissue of lateral aspect of neck. Tumor size was 1.8x1.8x1.5cm. Clinically, diagnosis of accessory thyroid was given. Histologically, it was involving the lymph node. Immunohistochemistry was positive for CD31 and negative for CD34. A diagnosis of Kaposiform Hemangioendothelioma was given.***

Keywords: Adult, Hemangioendothelioma , Kaposiform

1. Introduction

The term Hemangioendothelioma was introduced by Mallory in 1908 to include all tumor arising from blood vessels endothelium.[1]

Zuckerberg et al described it as an intermediate/borderline vascular neoplasm between a hemangioma and angiosarcoma[2] It is a locally aggressive rarely metastatic neoplasm, does not have tendency for spontaneous regression and has characteristic histopathological features, including tumor cell architectural pattern resembling a Kaposi Sarcoma. [3]

It is usually identified in infancy and first decade of life at sites like extremities and retroperitoneum and uncommonly in head and neck region. It is known for its association with lymphatic component namely lymphangioma/lymphangiomatosis and Kasabach Merritt phenomenon (KMP). At times, Kaposiform Hemangioendothelioma (KHE) can occur without KMP.[2] It has not been documented primarily in the lymph node.

Herein, we present an extremely uncommon case of KHE in a lymph node unassociated with KMP and lymphangioma, in a nineteen year-old female.

2. Case Report

A nineteen year-old female presented with swelling neck ?Lymph node ?Accessory thyroid clinically diagnosed as accessory thyroid on lateral aspect of neck.

Pathological findings - On gross, a grey brown nodular soft tissue piece was measuring 1.8x1.8x1.5cm. External surface was smooth to rough. Cut surface grey brown tan with central cystic space 1 cm in diameter filled with ?blood.

Histologically sections from different areas of the specimens were studied. Sections predominantly revealed partial effacement of lymph node with a few remnant follicles. There was vascular proliferation along with nodules showing proliferation of endothelial cells and malignant cells having vesicular nuclei and showing spindling at many places and the nodules were surrounded by fibrous bands [Fig.1]. Spindled tumor cells exhibited the vasoformative slit like lumen [Fig.2]. Extravasated blood, hemosiderin pigment lying free and in macrophages (Iron positive) was seen [Fig.3]. Numerous plasma cells were present. This picture was suggestive of Kaposi Sarcoma. Patient's HIV status was negative. PAS stain was negative. Immunohistochemical staining of spindle cells revealed CD31+ve and CD34-ve. A diagnosis of Kaposiform Hemangioendothelioma was given.

3. Discussion

Kaposiform Hemangioendothelioma (KHE) is a locally aggressive, immature vascular neoplasm, characterized by predominantly Kaposi sarcoma like fascicular spindle cell growth pattern.

Synonyms are Kaposi- like infantile hemangioendothelioma [4], hemangioma with Kaposi sarcoma like features.

The tumor most commonly occurs in the retroperitoneum [2,4] and the skin [3,5] but it can also occur in the head and neck region, [6] deeper soft tissue of extremities of the trunk and extremities. [2,7] No case of KHE primarily in lymph node has been reported except for a case of KHE in tonsil of child associated with cervical lymphangioma. [9]

KHE typically occurs in infancy and first decade of life, but adult cases are also recognized.[3,6,8] Lymphangioma and consumption coagulopathy (Kasabach-Merritt Syndrome) may complicate the larger tumor. Sometimes it may not be associated with Kasabach Merritt syndrome.[8] Soft tissue

tumors are greyish to reddish multinodular and may coalesce and encase surrounding structures.

Microscopically, the tumor grow in the form infiltrative vague lobules separated by fibrous septa. It consist of criss-crossing spindle cell fascicles interspersed with slit like sieve like blood vessel.[4,6] Nuclear atypia and mitotic activity are inconspicuous.[4,6] Fibrin thrombi, in the capillaries and areas of haemorrhage and hemosiderin deposit were seen.[6] No known association with HIV infection or HHV-8 was seen.

In adults the differential diagnosis of KHE comprises especially Kaposi sarcoma and spindle cell hemangioendothelioma, further differential diagnosis include tufted hemangioma and cellular capillary hemangioma which occurs rarely in adults.[6]

Immunohistochemical staining in spindle cells were positive for CD31 and negative for CD34. In our case, patient's HIV status was negative though Immunohistochemistry for HHV-8 was not done.

To conclude, KHE is an uncommon tumor with a distinct clinicopathologic features, including IHC profile and differs from a Kaposi Sacroma and other histological mimics. Careful attention towards its histopathological features coupled with IHC, is helpful in its identification, especially at rare sites like lymph node.

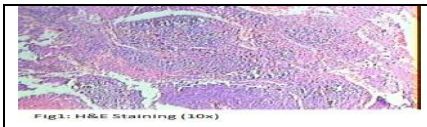

Fig.1
KaposiformHemangioendothelioma(H & E Section) : low power view (10x) showing lobular growth pattern of tumor cells.

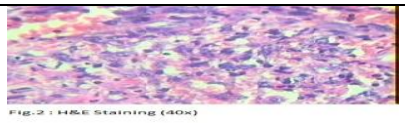

Fig.2
KaposiformHemangioendothelioma(H & E Section): High power view (40x) showingspindled tumor cells with slit like vascular spaces and extravasated red blood cells.

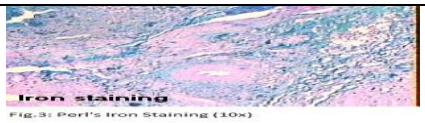

Fig.3
KaposiformHemangioendothelioma**:**Low power view(10x) showing Iron Stain positive for hemosiderin.

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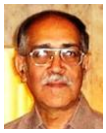

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