



EPITHELIOID HEMANGIOENDOTHELIOMA A CASE REPORT

Pathology

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ABSTRACT

The epithelioid hemangioendothelioma (EHE), is a rare tumor of vascular origin. It is composed of a distinctive type of endothelial cells having an epithelial-like appearance. We present a case of epithelioid hemangioendothelioma in a 16-year male with a well-defined nodular lesion of size approximately 4x5 cm projecting from left anterior nares with proximal verrucous appearance and distal erythematous border. The lesion slowly increased in size in the last 3 months and was associated with pain and bleeding. It was provisionally diagnosed as verrucous hemangioma but diagnosed as EHE on histopathology examination of biopsy tissue and confirmed by immunohistochemistry.

KEYWORDS

EHE (Epithelioid hemangioendothelioma), pure cutaneous involvement, IHC (Immuno histochemistry).

INTRODUCTION

Epithelioid hemangioendothelioma, an angiocentric vascular tumor, can present at almost any age but rarely occurs during childhood. (1,2) It affects both sex equally. The tumor develops as a solitary, slightly painful mass in either superficial or deep soft tissue, although it may occur multifocally. Approximately 50% cases are closely associated with or arise from a vessel, usually a vein. In some cases, occlusion of vessel accounts for more profound symptoms, such as edema and thrombophlebitis. Those tumors that arise from a vessel have a variegated, white-red color and superficially resemble organizing thrombi, except that they are firmly attached to the surrounding soft tissue.

Here in, we describe a case of EHE in a 16-year male with emphasis on its histopathological appearance and immunohistochemistry.

CASE REPORT

A 16-year male with a well-defined nodular lesion of size approximately 4x5 cm projecting from left anterior nares with proximal verrucous appearance and distal erythematous or indurated border presented in OPD of Department of Dermatology and USG report was unremarkable. CT scan thorax was unremarkable and CT scan of face – there is soft tissue density lesion, centered at left naso-labial fold. The lesion was measuring approximately 2.1x3.8x4 cm. The lesion is extending inferiorly up to lower margin of upper lip, medially the lesion minimal narrowing of left nasal cavity. Underlying bony outline is well preserved.

It was provisionally diagnosed as verrucous hemangioma.

Incisional biopsy from indurated part and verrucous part was done and sent to Histopathology Section of Department of Pathology.

Vial-1 (indurated part)- contained single skin-covered soft tissue piece measuring 0.8x0.4x0.3 cm.

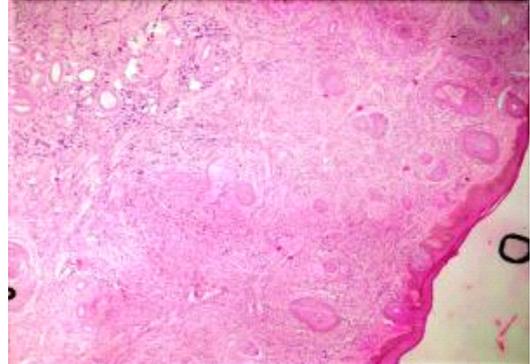
Vial -2 (biopsy from verrucous part)- comprised of multiple gray-brown soft tissue pieces collectively measuring 0.4x0.3x0.2 cm.

H&E section showed diffuse infiltration of ovoid, cuboidal and spindle-shaped cells with eosinophilic cytoplasm, mild atypia and increased mitotic figures. At places tumor cells were forming vascular channels lined by epithelioid type of endothelial cells. Hyalinization of stroma was also seen.

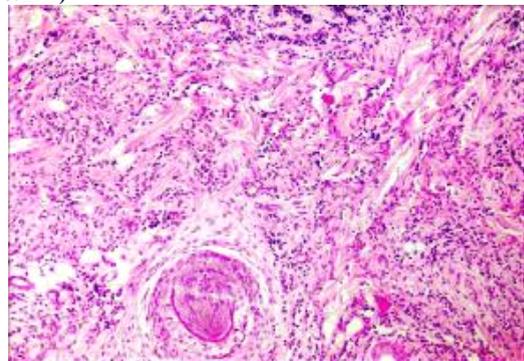
IHC Study: tumor was positive for CD31, CD34 and FLI-1 and negative for S100, CD3, CD20, CK.



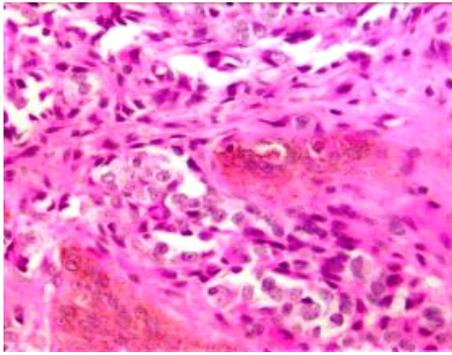
Pic1: Clinical Presentation of Epithelioid Hemangioendothelioma



Pic2: Fairly circumscribed collection of tumor cells in the dermis (H&E 4x)



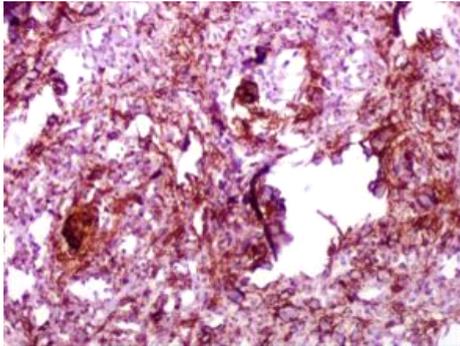
Pic3: Short fascicles of small tumor cells in the dermis (H&E 4x)



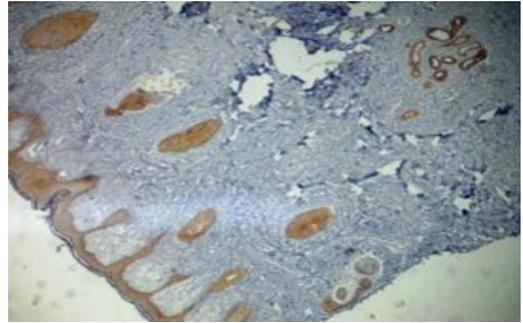
Pic4: Vascular channels lined by Epithelioid type of endothelial cells (H&E40x)



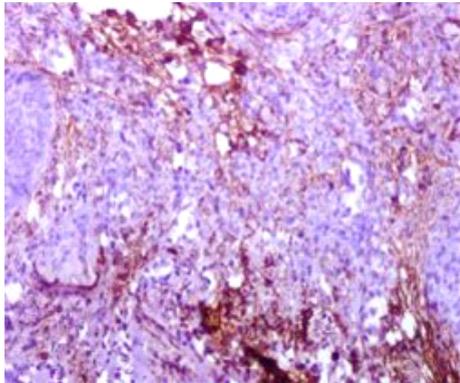
Pic9: Negative staining of tumor cells with CD3 (IHC 4x)



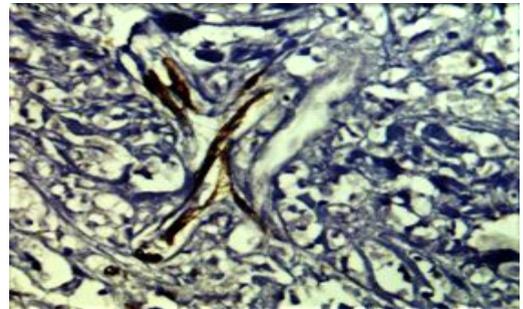
Pic5: Tumor cells stain positive with CD31(IHC 10X)



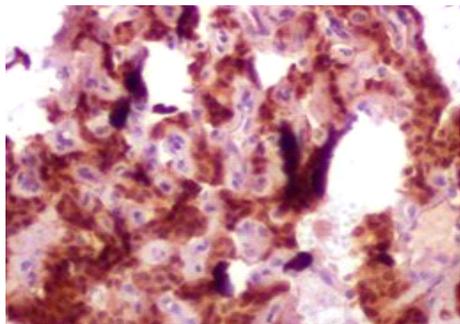
Pic10: Negative staining of tumor cells with CK (IHC 4x)



Pic6: Tumor cells stain positive with CD34 (IHC 10)



Pic11: Tumor cells stain negative with S100(IHC 40x)



Pic7: Tumor cells stain positive with FLI-1(IHC 40x)



Pic8: Negative staining of tumor cells with CD20 (IHC 10x)

DISCUSSION

The epithelioid hemangioendothelioma, an angiogenic vascular tumor, can occur at almost any age but rarely occurs during childhood^{1,2}. It affects both sexes equally and no predisposing factor identified⁵. The tumor develops as a solitary, slightly painful mass in either superficial or deep soft tissue. 50% cases are closely associated or arise from vessel, usually a vein. Those tumors that arise from vessels usually have a variegated, white red color and superficially resemble organized thrombi, except that they are firmly attached to the surrounding soft tissue.

Microscopic features: lesion that arise from vessels have a characteristic appearance when seen at low power. They expand the vessel, usually preserving its architecture as they extend centrifugally from the lumen to the soft tissue. The lumen is filled with a combination of tumors, necrotic debris and dense collagen. Unlike epithelioid hemangioma, in which vascular differentiation proceeds through the formation of multicellular, canalized vascular channels, vascular differentiation in these tumors is more primitive and is expressed primarily at the cellular level. The tumors are composed of short strands or solid nests of rounded to slightly spindled endothelial cells. Rarely are large, distinct vascular channel seen, except in more peripheral portions of the tumors. Instead, the tumor cells form small intracellular lumens, which are seen as clear spaces, or vacuoles, that distort or blister the cell. In most cases the tumors appear quite bland, and there is virtually no mitotic activity. In about ¼ cases the tumors contain areas with significant atypia, mitotic activity (more than 1 mitosis per 10 high power field (HPF)), focal spindling of the cells, or necrosis. When metastasis occurs in this disease, they usually develop from tumors with these atypical features.

Differential diagnosis:

1. Metastatic carcinoma (or melanoma)
2. Angiosarcomas with epithelioid appearance.
3. Epithelioid Hemangioma

In general carcinomas and melanomas metastatic to soft tissue display far more nuclear atypia and mitotic activity than epithelioid hemangioendothelioma and are rarely angiocentric and metastatic carcinoma are FLI-1 negative, melanoma is S100 positive.

Angiosarcomas with epithelioid pattern are composed of solid sheets of highly atypical, mitotically active, epithelioid endothelial cells. Necrosis is common, and vascular differentiation is expressed primarily by the formation of irregular sinusoidal vascular channels.

Epithelioid sarcoma is perhaps the closest mimic of the epithelioid hemangioendothelioma. Composed of rounded eosinophilic cells that surround cores of necrotic debris and collagen. In ambiguous cases, immunohistochemistry and electron microscopy may provide the most reliable clues for differentiation. About ¼ of epithelioid hemangioendotheliomas express cytokeratin³, but usually the staining is less intense and focal compared to epithelioid sarcoma. With optimal material, Von Willebrand factor can be demonstrated in the cytoplasm of most epithelioid hemangioendothelioma. The cells of epithelioid hemangioendothelioma express CD31 and CD34. By electron microscopy, the cells have the characteristics of endothelium, including well-developed basal lamina, pinocytotic vesicles and occasional Weibel-Palade bodies⁴.

Epithelioid hemangioma has lobular appearance and prominent endothelial cells with perivascular fibrosis and neutrophilic and eosinophilic infiltration.

CONCLUSION

Pure cutaneous involvement of EHE is very rare and it can have variable clinical appearance in the form of verrucous lesion or as an indurated lesion. Therefore, it should also be considered in the D/D of such type of lesions.

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