

CASE REPORT

CAVERNOUS HAEMANGIOMA WITH RETIFORM HAEMANGIOENDOTHELIOMA - A CASE REPORT

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ABSTRACT: INTRODUCTION: Retiform hemangioendothelioma is a rare variant of low-grade angiosarcoma with an indolent clinical behaviour, with predilection for young adults. Mostly is seen in the extremities, especially the distal lower limbs. **CASE REPORT:** An eighty year old lady, presented with complaints of a solitary, well defined swelling below the nape of the neck since two years. The swelling was clinically diagnosed as cavernous haemangioma. However the colour of the lesions was looking like a melanocytic naevus. Excision was done under local anaesthesia and was sent for histopathological examination. Histopathological diagnosis was confirmed as "Cavernous haemangioma with Retiform Haemangioendothelioma".

KEY-WORDS: Elderly female, Cavernous haemangioma, Retiform haemangioendothelioma.

INTRODUCTION: Retiform haemangioendothelioma is a rare variant of low-grade angiosarcoma with an indolent clinical behaviour, with predilection for young adults. It presents as a slow growing non-distinct tumour with equal gender incidence. Retiform haemangioendothelioma has a predilection for extremities, especially the distal lower limbs.

Haemangiomas are basically hamartomatous malformations and are observed since birth. As age advances, they spontaneously regress. In literature, no such cases have been demonstrated having both cavernous haemangioma and features of retiform haemangioendothelioma.

The true neoplastic proliferation of blood vessels can be divided into:

- Benign
- Low malignant potential
- High grade malignancies

Among the tumours of low malignant potential, there is a family of vascular tumours characterized by epithelioid endothelial cells. These represent the benign end of the spectrum which in turn are characterised by hobnail endothelial cells (match stick) appearance. This entity includes:

1. Dabska haemangioendothelioma
2. Retiform haemangioendothelioma
3. Hobnail haemangioma (Targetoid Haemosiderotic Haemangioma)

CASE HISTORY: An eighty year old female presented with a solitary swelling in the upper back just below nape of neck since two years. The swelling was associated with pain. There was no other swelling in the body. There was no other relevant history. On clinical examination a single well defined reddish swelling measuring 1x 1 cm was present in the upper back just below the nape of neck. Swelling was soft in consistency, and tender. The swelling completely disappeared on compression, and gradually refilled very slowly. Surrounding skin was normal. Excision was done under local anaesthesia, as a day care procedure. The excised tissue was sent for histopathological examination.

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MACROSCOPY: Container had a single skin covered grey black, grey white tissue piece measuring 1x1 cm in diameter. Cut section showed solid, grey white areas.

MICROSCOPY: The sections were studied with Haematoxylin and Eosin stain. The epidermis was unremarkable. Beneath the epidermis there were cavernous vascular spaces which were lined by endothelial cells [Figure 1, 2 & 4]. There were also papillary configuration and tufts of capillaries with hobnail pattern or match stick pattern [Figure 3, 5, 6 & 7], suggesting that there is an overlap of cavernous haemangioma with retiform haemangioendothelioma. Some proliferating endothelial cells were very compact forming slit like vascular spaces [Figure 8 & 9]. The histopathological diagnosis was confirmed as cavernous haemangioma with retiform haemangioendothelioma.

DISCUSSION: Cavernous hemangiomas are less frequent and are most common during childhood, located in the upper portion of the body. Superficial lesions are blue, puffy masses with irregular surface caused by dilatation of the vessels. Deep lesions impart little or no colour. They show no tendency for spontaneous regression ^{1, 2}. It is rarely associated with an overlying capillary haemangioma. Cavernous haemangioma is composed of large, dilated, blood filled vessels arranged in a roughly lobular or diffuse haphazard pattern, and lined by flattened endothelium.

Retiform hemangioendothelioma is a rare variant of low-grade angiosarcoma with an indolent clinical behaviour^{3, 4, 5} with predilection for young adults. It presents as a slow growing non-distinct tumour with equal gender incidence and a predilection for extremities, especially the distal lower limbs. Multiple recurrences are common, but metastasis is rare. Tumours are ill-defined and involve the reticular dermis with frequent extension into the subcutis. In low power the presence of elongated, arborizing blood vessels resembling the shape of rete testis, are lined by monomorphic bland endothelial cells with prominent apical nuclei and scanty cytoplasm. These cells have been described as having a “matchstick” or hobnail appearance. A lymphocytic inflammatory infiltrate, though not invariably present, is seen in the stroma and in the vascular lumina. Occasional intravascular papillae with hyaline cores can be seen.

Differential diagnosis of retiform haemangioendothelioma:

1. Dabska's tumour	2. Angiosarcoma
<ul style="list-style-type: none">• Hobnail haemangioma (targetoid hemosiderotic haemangioma).• More superficial.• Lacks a retiform architecture.• Has hobnail endothelial cells mainly seen in vessels near the surface.	<ul style="list-style-type: none">• Presents in a different clinical setting.• Cytologic atypia and mitosis are present.• Absence of hobnail endothelial cells.

The present case was clinically suspected as cavernous haemangioma, however the colour was looking like a naevus. Microscopically, the lesion had the features compatible with both cavernous haemangioma and retiform haemangioendothelioma. Both of these lesions appear to co-exist in the same lesion. These tumours cannot be clinically differentiated. So the role of pathologist in making the final diagnosis is important.

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MICROSCOPIC PICTURES: H & E SECTIONS:

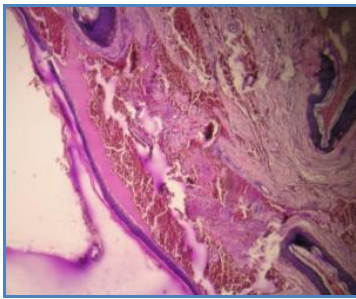


Figure 1. 4X

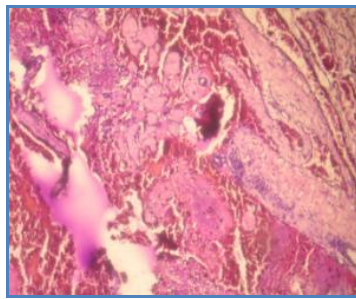


Figure 2. 10X

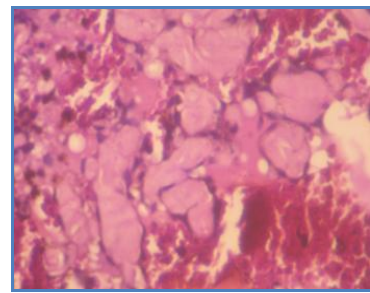


Figure 3. 20X

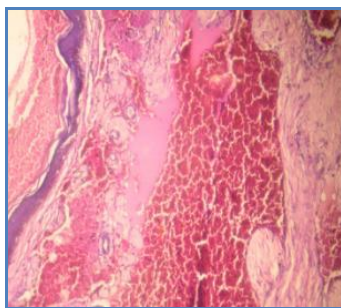


Figure 4. 10X

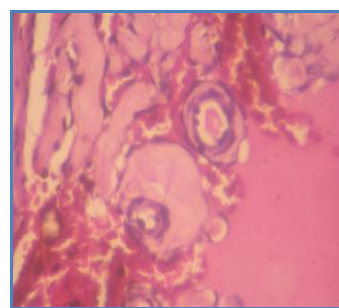


Figure 5. 20X

Figure 1, 2 & 4:- Squamous lining below which cavernous vascular spaces were observed, which were lined by endothelial cells.

Figure 3 & 5:- Also seen were capillaries with hobnail or match stick pattern- Retiform Haemangioendothelioma like pattern.

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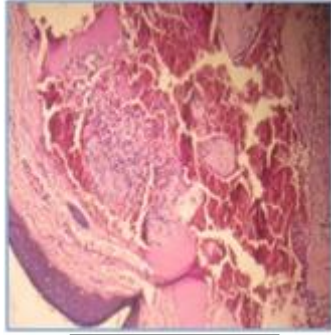


Figure 6. 10X

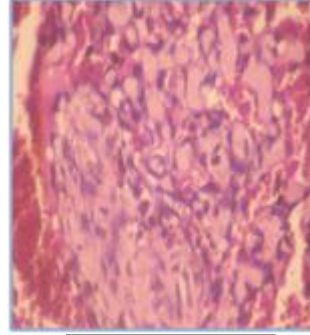


Figure 7. 20X

Figure 6 & 7:- Capillaries with hobnail or match stick pattern- Retiform Haemangioendothelioma like pattern.

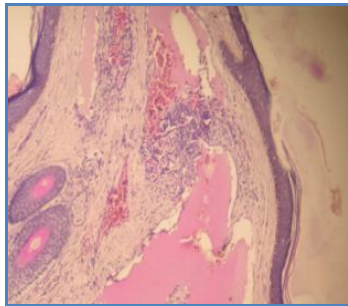


Figure 8. 4X

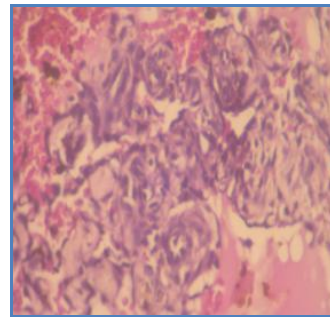


Figure 9. 10X

Figure 8 & 9: Some proliferating endothelial cells were very compact with formation of slit like vascular spaces.

- All the microscopic pictures were taken using Nikon Cool pix Model 8400.
- X - Indicates the power of Objective.
- Stain used - Haematoxylin and Eosin.

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