



## A rare presentation of Retiform Hemangioendothelioma in sacral area

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### ABSTRACT

*Retiform Hemangioendothelioma is an extremely rare tumor. It was first explained by Calonje et al in 1994. It is a vascular neoplasm of borderline malignancy, mostly seen in young age, and extremities. A 65 years old female patient presented with a recurrent growth in the sacral region, along with skin ulceration. FNAC revealed a cystic lesion with inflammation. Previous biopsy showed granulation tissue only. The growth was surgically removed for the third time and sent to Histopathology lab. Gross examination revealed a partial skin covered tissue mass of 15x10x3 cm. Cut section showed solid cystic growth of 2x2x1.5 cm. Microscopically, proliferating neoplastic elements, vascular in nature, in a net like pattern seen. Monomorphic cells with scant cytoplasm with hobnail nuclei lined the vessels. Scant mitotic figures, lymphocytic infiltration, and focal fibrocollagenous material were seen. Dermis was focally involved. On immunohistochemistry, Ki-67 (low proliferation) and CD 34 were positive. Hence the diagnosis of Retiform Hemangioendothelioma was made.*

**Key words:** Hemangioendothelioma, tumor

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### INTRODUCTION

Retiform Hemangioendothelioma is one of the most common variants of Composite Hemangioendothelioma – a vascular neoplasm expressing blending of ingredients like benign, low-grade malignancy, and frank malignancy, hence infinite variants and sub-variants of vascular tumors show sharing of features amongst themselves. Therefore diagnosing such kind of tumor can be challenging, especially when the tumor site is rare for its occurrence, and age of the patient doesn't match with the most common incidental group. The tumor also shows high recurrence, so an accurate diagnosis and surgical resection with follow up is very essential.

### CASE HISTORY:

A 65 years female patient presented to the Surgery OPD of JNMC, AMU, Aligarh, India with the chief complaint of recurrent growth in the sacral region, along with skin ulceration. Previous two biopsies showed granulation tissue only. The growth was surgically removed for the third time and sent for histopathology.

### Case Details:

#### Histopathology

Gross examination revealed: A partial skin covered tissue mass of 15x10x3 cm, showed an ulcerative growth of 8x5cm. Cut surface of the mass showed solid cystic growth of 2x2x1.5 cm. Multiple sections were taken for histopathological assessment. Microscopic examination revealed: H& E stained sections showed an infiltrating lesion with proliferating neoplastic vascular elements arranged in a net like pattern. Vessels were lined by round monomorphic hyperchromatic cells with scant cytoplasm, protruding into lumina imitating "hobnail" appearance [Fig. A, B]. Scant mitoses, focal atypia, and focal fibrocollagenous material were noted. Lymphocytic infiltration was present. Since this tumor has high recurrence rate, the margins of the tumor were assessed and found to be free from the neoplasm.

On immunohistochemistry, the vascular marker CD 34 was positive [Fig. C] along with Ki-67 (low proliferation) [Fig. D]. Hence the diagnosis of Retiform Hemangioendothelioma was made. The patient is on follow up for 10 months and no recurrence is seen.

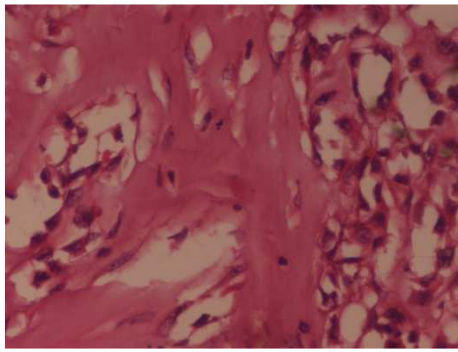


Fig. A - H&amp;E (200X)

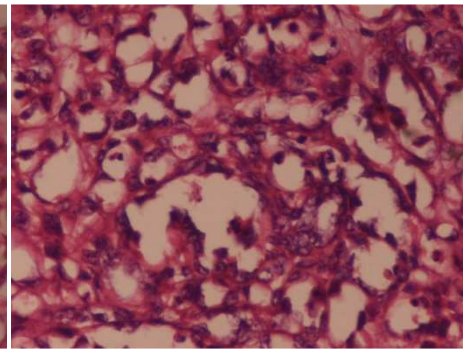


Fig. B - H&amp;E (200X)

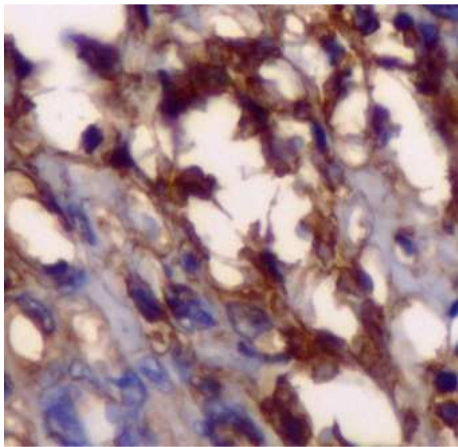


Fig. C - CD34 positive (400X)

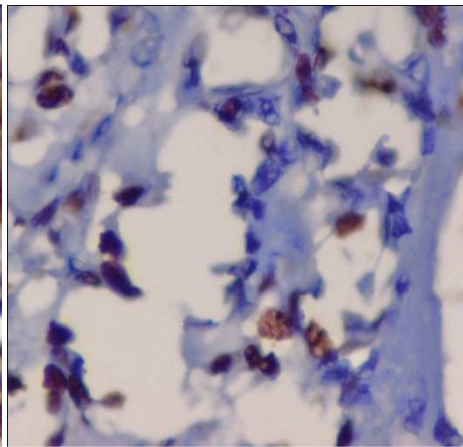


Fig. D - Ki-67 (low proliferation) (400X)

### DISCUSSION

An intermediate lesion between Hemangioma and Angiosarcoma, the age group of Retiform Hemangioendothelioma ranges from 6 to 78 years, most patients presenting between second to fourth decade of life and the mean age being 36 years [1]. Male to female incidence ratio is 1:2 [1]. Local recurrence is seen in 60% of cases [1]. The most common presentation of Retiform Hemangioendothelioma is in the skin, especially in lower limbs. Most patients present with an exophytic, slow growing nodules or plaques in the dermis or subcutaneous tissue that may vary from 2 months to 10 years. Tumor size varies from 1 to 30 cm, mostly being <5.0cm [2]. Prox1 (Prospero Homeobox 1), a transcription factor that regulates and maintains the lymphatic endothelial phenotype, has a role in the development of Retiform Hemangioendothelioma. It also has potential diagnostic use as an immunohistochemical marker [3].

Histologically, Retiform Hemangioendothelioma mainly occurs in the dermis, more specifically the reticular layer, but larger tumors extend to the subcutaneous adipose tissue, occasionally interspersed with collagen and skin appendages surrounding the invasive growth [4]. On light microscopy, Retiform Hemangioendothelioma shows a retiform pattern of multiple interconnecting, arborizing blood vessels with the lining of monomorphic hobnail endothelial cells with mild atypia and no mitotic figures [5]. Additionally, there is lymphocytic infiltration [6]. On Immunohistochemistry, the neoplastic cells of retiform hemangioendothelioma show positivity with CD31, CD34 and factor VIII related antigen [5]. In our case, the neoplastic cells were morphologically and immunohistochemically in favour of Retiform Hemangioendothelioma. MIB-1 index of Ki-67 showed low proliferation. To rule out any evidence of Angiosarcoma, multiple sections from the same resected specimen were examined. The case fulfilled the criteria for Retiform Hemangioendothelioma.

**Differential diagnosis:**

1. Hobnail Hemangioma can be differentiated from retiform hemangioendothelioma by the presence of elongated vascular spaces with the lining of hobnail-like endothelial cells, confined to the superficial skin without extending to the deep dermis or subcutaneous tissue, along with hemosiderin deposits [8].

2. Dabska tumor can be differentiated from retiform hemangioendothelioma by the presence of papillary tufts along with a central hyaline core with lining of hobnail-like endothelial cells with protrusion into the lumina whereas in retiform hemangioendothelioma, rather than papillary tufts there are papillary infoldings along with arborizing rete testis like architecture of blood vessels, which are not present in Dabska tumor [9].

3. Angiosarcoma can be differentiated from Retiform Hemangioendothelioma by the presence of significant nuclear atypia and conspicuous mitotic figures and the lack of the typical retiform pattern of blood vessels. Additionally, Angiosarcoma has higher invasive and metastatic potential than Retiform Hemangioendothelioma [7, 10]. A true Retiform Hemangioendothelioma may be misdiagnosed as Angiosarcoma if it involves skin along with invasive growth in the dermis. On the other hand, a true Angiosarcoma may get misdiagnosed as Retiform Hemangioendothelioma if the endothelial cells due to intravascular protrusions may be mistaken for spikes of angiosarcoma [4]. Recurrence rates are high, therefore the preferred treatment is wide surgical excision with histopathologically certified tumor-free margins [11]. Our patient was diagnosed for Retiform Hemangioendothelioma on sacral region growth, an atypical site.

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**Conflict of interest:** None

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