Retiform hemangioendothelioma with lymph node metastasis: A rare entity

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ABSTRACT
Retiform hemangioendothelioma (RH) is a rare, recently described, unique kind of low grade angiosarcoma. The tumor is characterized by distinctive arborizing blood vessels that mimic the appearance of rete testis. The tumor chiefly originates from the skin and subcutaneous tissue and has a tendency to recur locally. Only one case with lymph node metastasis has been reported previously. Another such rare case of RH is being reported in an adult female who presented with a subcutaneous nodule, underwent multiple recurrences and uniquely metastasized to regional lymph node.

Key words: Lymph node metastasis, Recurrence, Retiform hemangioendothelioma

INTRODUCTION
Retiform hemangioendothelioma (RH) was first described in 1994 as a distinctive form of low grade angiosarcoma.[1] RH is a locally aggressive, very rarely metastasizing vascular lesion, characterized by distinctive arborizing blood vessels lined by endothelial cells with characteristic hobnail morphology.[2] The low power appearance of the tumor resembles that of rete testis and hence the name. The tumor principally involves the skin and subcutaneous tissue. RH is an uncommon tumor and only 24 cases have reported in a recent review.[3] Only one case with lymph node metastasis has been identified in the literature.[4]

One such rare case of RH is being reported in an adult female who presented with multiple recurrences and regional lymph node metastasis.

CASE REPORT
A 35-year-old female presented with a slow growing, shiny subcutaneous nodule over mons pubis. The nodule was about 4 cm in diameter and the overlying skin did not show any ulceration. The patient did not have any significant medical history and there was no history of previous radiotherapy or pre-existing lymphedema. All routine laboratory examinations (including complete blood count and biochemical parameters), chest radiography, and abdominal ultrasonography were unremarkable. The nodular lesion was about 6 cm when it was first seen. The tumor recurred two years after the excision and was excised again. The second recurrence occurred seven months later, and at this time it presented as an ulcerated nodule. Metastasis to the right inguinal lymph node was then found. Biopsies were collected from the initial lesion as well as from the two recurrent lesions. After the second recurrence, the patient was lost to follow up.

Pathological findings
The cut section of the initial lesion biopsy, 5 x 4 x 4 cm, lobulated nodule [Figure 1] covered by skin, was grayish white in color and predominantly solid with occasional spongy areas. Microscopically, the tumor was located deep in the dermis and involved subcutaneous fat. The lesion showed multiple interconnecting blood vessels arranged in retiform pattern. The endothelial cells lining the vessels were single layered and showing monomorphic nuclei with scanty cytoplasm and distinctive hobnail pattern [Figure 2]. In focal areas, papillae projecting into lumen were noted [Figure 3]. There was absence of solid zones and lack of nuclear atypia and mitosis. Stroma in between the tumor cells showed prominent infiltration by lymphocytes.

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Histopathologically, the inguinal lymph node showed vascular spaces lined by tumor cells, still showing hobnail morphology at places amid the lymphoid tissue indicative of metastatic deposit [Figure 4]. The tumor and metastatic deposit were negative for PAS stain.

DISCUSSION

Retiform hemangioendothelioma is a distinctive low grade angiosarcoma of skin. Since its original description of 15 cases in 1994, some sporadic cases have been reported. Most patients present between second to fourth decade of life, the youngest being 9-years old and oldest 78 years (mean age 36 years). There is a female predilection. Exceptional cases occur in the setting of previous radiotherapy or pre-existing lymphedema. Typically, RH is a slow growing nodular or plaque-like lesion. RH usually occurs as a single lesion, but multiple lesions affecting different anatomic sites have been reported by Duke et al.

The term “low grade angiosarcoma” refers to a group of vascular neoplasms that have a histopathologic appearance intermediate between hemangioma and angiosarcoma. Epithelioid hemangioendothelioma, Dabska tumor, and retiform hemangioendothelioma are examples of low grade angiosarcoma.

The clinical picture does not provide help and the exact diagnosis therefore depends entirely on typical histological features. The clinical differential diagnosis in such a tumor prior to biopsy includes lymphoma, dermatofibrosarcoma protuberans, and cutaneous angiosarcoma.
Vascular tumors that mimic the histological picture of RH include angiosarcoma, tumors with hobnail endothelial cells, such as targetoid hemosiderotic hemangioma (hobnail hemangioma) and Dabska’s tumor.

The present case involved skin and subcutaneous tissue over mons pubis. Histologically, the blood vessels showed typical hobnail pattern. The tumor usually shows lymphocyte infiltration in stroma, which was obviously evident in the present case. It recurred twice and also showed lymph node metastases.

There were no solid areas, vacuolated endothelial cells, nuclear pleomorphism, or mitotic activity which excluded the possibility of an angiosarcoma. Hobnail hemangiomas have a characteristic clinical appearance characterized by a small solitary lesion, consisting of a brown to violaceous papule surrounded by a thin pale area and a peripheral ecchymotic ring; microscopically, these tumors are located more superficially, lack a retiform architecture, and have hobnail endothelial cells that are mainly seen in the vessels near the surface.

Dabska tumor was also ruled out as there were only focal papillary projections and no hyaline cores containing basement-membrane-like material and that it usually occurs in children. Another differential diagnosis was apocrine carcinoma. Typical vascular appearance of tumor focally with papillary projection and absence of characteristic morphology of apocrine cells helped to exclude such possibility.

This neoplasm frequently recurs but rarely metastasizes. Lymph node metastasis was seen in single case reported so far where the tumor was present in biphasic pattern and only spindle-shaped component metastasized. PAS stain was negative in the present case, thus, excluding the possibility of metastatic deposits from an adenocarcinoma. RH tumor cells react with vascular endothelial markers CD31, CD34, and factor VIII-related antigen.

Due to the tendency for recurrence, the treatment of choice is surgical excision with histopathologically tumor-free margins. Radiation therapy could be effective as demonstrated by the successful treatment of the aforementioned RH lymph node metastasis.

The present case is extremely rare and has got distinctive presentation with multiple recurrences and even regional lymph node metastases.

REFERENCES