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Case Report

Retiform hemangioendothelioma of breast: A rare case report

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Abstract

Retiform hemangioendothelioma is a very rare vascular endothelial tumor which belongs to rarely metastasizing subgroup of intermediate category. It commonly affects children and young adults and involve skin and subcutaneous tissue of lower extremities. Here we describe a case of 52years female who presented with lump in left breast for 2 years in upper inner quadrant. Her biopsy was in favour of malignancy for which mastectomy was done but finally she was diagnosed with retiform hemangioendothelioma of left breast which is an extremely rare site for this tumor to occur.

Keywords: Breast, Mastectomy, Retiform hemangioendothelioma, Vascular endothelial tumor.

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1. Introduction

Retiform haemangioendothelioma was first described by Calonge et al. in 1994.[1] They are named because of similarity to rete testis histologically lined by endothelial cells. They are exophytic and slow growing vascular tumors found in children and young adults.[2] They have equal predilection for males and females and usually occur on extremities or trunk.[2,3] According to recent WHO classification it is grouped under intermediate category of vascular tumors with rarely metastasizing nature. [2,3] Only one case of lymph node metastasis has been reported.[1] Only 33 cases have been described till date in English literature.[4] To the best of our knowledge no case have been reported in breast.

2. Case Report

A 52years presented to out-patient department with complaint of lump in left breast for 2 years. It was painless and gradually progressing with no history of nipple discharge. There was no history of swelling in opposite breast or any other site, fever, weight loss and trauma to breast. There was no significant family or past history and patient was not on any medications.

On examination 11x10cm firm, non-mobile and non-tender lump was palpable in upper inner quadrant of left breast with peau d'orange appearance and retraction of nipple was also seen. No axillary lymphadenopathy was noted.

Left breast mammography showed ill-defined asymmetric density in upper inner quadrant of left breast with no calcifications. Right breast and both axillae were unremarkable. Left breast core biopsy showed clusters of atypical cells with increased nuclear to cytoplasmic ratio and hyperchromasia and moderate pleomorphism with few cells seen invading the stroma. Findings favoured the diagnosis of invasive breast carcinoma: no special type. Following this her left radical mastectomy was performed.

Grossly whole specimen measured 21x15x8cm and on serial sectioning 10x10x6.5cm tumor was identified in upper inner quadrant. Cut section of tumor was variegated in appearance with areas of haemorrhage (**Figure 1**). Tumor was grossly involving the medial margin and base. 5 lymph nodes were retrieved.

On microscopic examination sheets of retiform pattern of blood vessels was seen which were lined by monomorphic

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hobnail endothelial cells with round naked type nuclei and scant to nil cytoplasm (**Figure 2,3**). All the margins and base were uninvolved. All 5 lymph nodes were free from tumor cells. Tumor cells showed positivity for CD31 and CD34 (**Figure 4**) while Pan CK was negative.

On the basis of history, clinical examination, radiological and histopathological examination she was diagnosed as a case of retiform hemangioendothelioma of intermediate grade ($pT_{1b}N_0$).

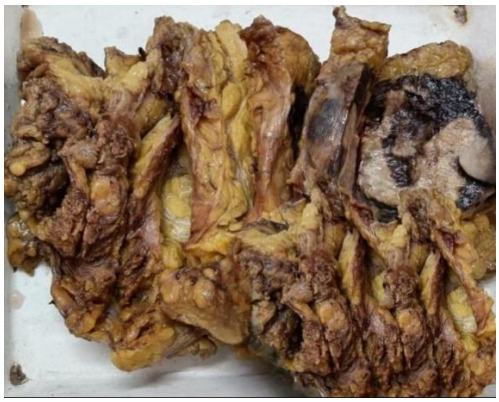

Figure 1: Gross- Left mastectomy specimen measuring 21x15x8 cm. Cut section shows well circumscribed variegated tumor measuring 10x10x6.5cm in upper outer quadrant.

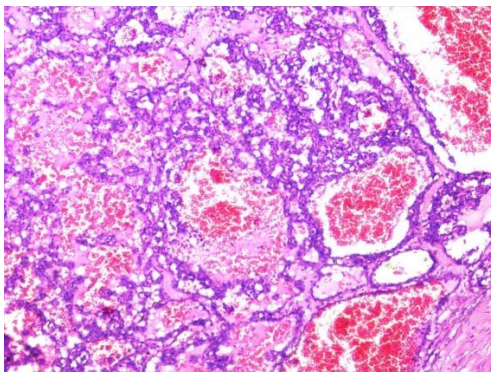

Figure 2: Section shows numerous blood vessels arranged in retiform pattern (H&E x 100)

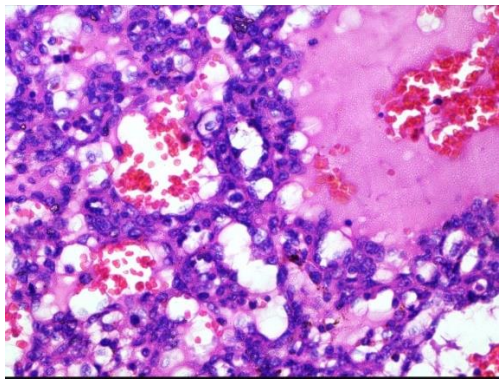

Figure 3: Section shows vessels lined by monomorphic hobnail endothelial cells with scant cytoplasm and round naked type nuclei (H&Ex 400)

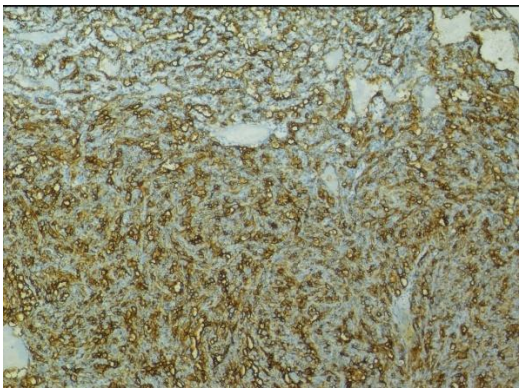

Figure 4: IHC CD34- Diffuse and strong membranous positivity in tumor cells.

3. Discussion

Requena and Kutzner summarized hemangioendotheliomas (HE) and presented seven subtypes including RH, spindle cell HE (hemangioma), papillary intralymphatic angioendothelioma (Dabska tumor), RH, kaposiform HE, epithelioid HE, pseudomyogenic HE (epithelioid sarcoma-like HE), and composite HE.[5] Retiform hemangioendothelioma is a rare vascular tumor that show borderline biological behaviour and belong to intermediate category between hemangioma and angiosarcoma.[6] It is a locally aggressive very rarely metastasizing vascular neoplasm. Histopathologically it is characterized by distinctive arborizing blood vessels lined by hobnail endothelial cells.[7] It has derived its name because of similar morphological appearance to rete testis on low power.[8] One of the differential diagnoses of retiform hemangioendothelioma includes angiosarcoma but angiosarcoma lacks retiform blood vessels and exhibit nuclear atypia and mitotic figures which are absent in our case. In addition, angiosarcoma has higher risk of metastasis and recurrence.[9] Presence of infiltrative vascular spaces allows distinction from benign entities like hobnail hemangioma which is well defined and superficial.[10]

RH usually located usually on limbs and occasionally on trunk.[6] Cases of back, scalp, perineal area, medial canthus, earlobe, pleura, jejunum and labia majora have also been reported. But no case in breast have been reported till date to the best of our knowledge.[11-15] It can present as single or multiple lesions commonly in middle age adults with mean age being 4th decade.[16] RH can also be seen with other intermediate category vascular tumors most commonly papillary intralymphatic angioendothelioma (Dabska Tumor).[17]

Neoplastic endothelial cells in RH consistently express endothelial markers as von Willebrand factor, CD31 and CD34, while the lymphocytic infiltrate shows a mixture of CD3+, T and CD20+ B cells.[18] In our case also tumor cells showed CD31 and CD34 positivity. Surgical resection with free tumor margins is the treatment of choice but recurrence

can occur. Adjuvant radiotherapy has been effectively administered in unresectable cases.[11]

Distinction from other entities, proper management and follow up of patient to look for recurrence are essential.

4. Conclusion

This case highlights the new site of occurrence of retiform hemangioendothelioma. Is a rare vascular tumor and occurrence in breast make even more unique. All breast lumps should be properly examined with immunohistochemistry as prognosis and treatment differ for different entities.

5. Conflict of Interest

None.

6. Source of Funding

None.

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