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Dr. Dharmendra Shah
Head of Unit, Department of Surgery, Medical College Baroda and SSG Hospital, Vadodara, Gujarat, India

Dr. Shivani Chaudhary
Senior Resident, Department of Surgery, Medical College Baroda and SSG Hospital, Vadodara, Gujarat, India

Het Contractor
Final Year Medical Student, Medical College Baroda, Gujarat, India

Milauni Dave
Final Year Medical Student, Medical College Baroda, Gujarat, India

Corresponding Author:
Milauni Dave
Final Year Medical Student, Medical College Baroda, Gujarat, India

Retiform hemangioendothelioma: A rare case of slow growing vascular tumor with review of literature

Dr. Dharmendra Shah, Dr. Shivani Chaudhary, Het Contractor and Milauni Dave

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Abstract
Retiform hemangioendotheliomas are extremely rare tumors of vascular origin. Overall incidence of hemangioendothelioma being <1/1000000. They are low grade angiosarcomas with little risk of metastasis. It usually occurs as a single skin or subcutaneous lesion on the trunk or extremities commonly seen in middle aged adults [1]. Due to its rarity no well-defined guidelines are there for its management, follow up and dealing with local recurrence [2]. Therefore we would like to discuss the case of Retiform Hemangioendothelioma over scapular region in an adult lady which presented with its classical features and how it was managed.

Keywords: Retiform hemangioendothelioma, vascular tumor

Introduction
Hemangioendothelioma is the term used to describe vascular neoplasms that show a borderline biological behavior. They may range from entirely benign hemangiomas to highly malignant angiosarcomas [3]. As per WHO classification Hemangioendotheliomas were included in the mesenchymal tumor category. Amongst its various types Retiform Hemangioendothelioma (RH) also known as Hobnail Hemangioendothelioma is one of the rarest forms. This subtype is commonly seen over the trunk and extremities.

Case report
A 35-year-old lady presented with complaint of hyperpigmented patch on skin over upper back for 5 years. The size of patch has gradually progressed over the period of time. Patient didn't have complaints of pain, itching, trauma over that site, she consulted the dermatologist for her cosmetic concern related to the patch. On examination Dermatologist found there was an ill-defined hyperpigmented plaque with multiple papules over left scapular region. [Fig. 1] The lesion was pulsatile and a mass could be palpated underneath and referred to us. As possibility of vascular malformation, we ordered Magnetic resonant Imaging (MRI), which revealed a focal, very large, well defined, multilobulated, abnormal signal intensity lesion of 4 x 3 x 6 cm with few nodular foci of blooming in the peripheral wall is seen in the intra and inter muscular region of left rhomboid muscle, possibility of slow flow vascular neoplasm. [Fig. 2a, 2b] After discussing the case with plastic surgeon and the vascular surgeon, decision was made to perform wide local excision with intraoperative frozen section. Surgery performed under general anesthesia; wide local excision was done with 2cm margin [Fig. 3a & b]. Intra operative frozen section from its margin showed to be free from tumor so the defect was closed with split thickness skin graft taken from the thigh of the patient. The histopathological examination showed upper dermis with arborizing blood vessels lined by monomorphic bland endothelial cells which showed hob-nailing. Surrounding stroma showed dense sclerosis and inflammation. [Fig. 4a, 4b] On Immunohistochemistry examination, tumor cells stained positive for CD 31, CD 34 suggestive of Retiform Hemangioendothelioma. Post operative course was uneventful with graft uptake of nearly 99%. Follow up at 9 months has shown no recurrence.

Discussion
The risk factors for developing RH formation seem to be linked to a previous exposure to therapeutic radiation, chronic obstruction of the lymph vessels or the presence of other skin cancers, and human herpes virus 8 infections [4, 5].

Diagnosis of vascular tumors has always been a challenging task for physicians as well as oncologists because of the overlapping histopathological features of vascular as well as epithelial components of tumor [6]. Diagnosis depends on the clinical features, histological features, and traditional immunohistochemical markers positivity. When RH is diagnosed as an accidental finding and not causing any trouble to the patient, then non-operative measures are adopted with periodic observations with medications to alleviate symptomatic discomfort if present. Depending on the location, size, and the histological features on the pathology report line of its further management is decided. Any combination of steroids, chemotherapy, radiation therapy, or invasive surgical procedures, are used to treat the tumors. Interferon injections are used to reduce tumor blood supply, which will help by limiting tumor growth. Wide local excision with removal of the entire lesion is the standard treatment mode used. Sometimes limb (hand, finger) amputations may have to be performed, while treating Retiform Hemangioendothelioma, in order to achieve a complete surgical excision [4, 7]. Vascular embolization of the feeding blood vessels is used to provide temporary relief from the symptoms, and reduce blood loss during a surgical procedure for giant RH. When RH is at an inaccessible location, or is unsafe for surgical intervention, then non-invasive procedures, such as radiation therapy and chemotherapy, are adopted. Complications from RH include significant blood loss during invasive or surgical removal, recurrence rate after surgical excision, metastasizing of the tumor to other body regions; though, this is quite infrequent. The long-term outlook for individuals with RH is not predictable and differs greatly among affected individuals. The prognosis depends upon many factors firstly, the type of hemangioendothelioma [8] and whether the condition has metastasized to nearby tissues/ lymph nodes. Secondly on recurrence, with increasing recurrence the prognosis keeps on worsening. For individuals with retiform hemangioendothelioma, local recurrence is as common as up to 50% of cases has shown to reoccur but metastatic disease is rare [7]. Due to its high chances of recurrence, often several years of active follow-up, vigilance is necessary [9]. Current medical research have not established a way of preventing Retiform Hemangioendothelioma occurrence.

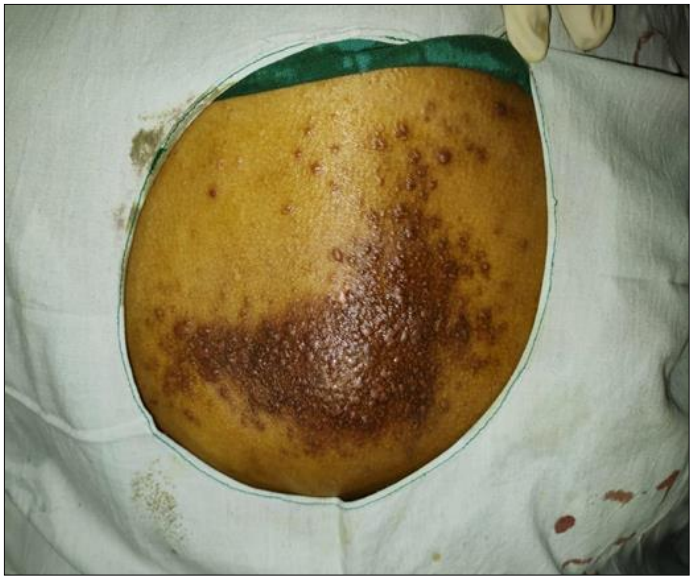

Fig 1: Showing multiple hyperpigmented papules forming a large plaque over the right scapular region with underlying Bulge

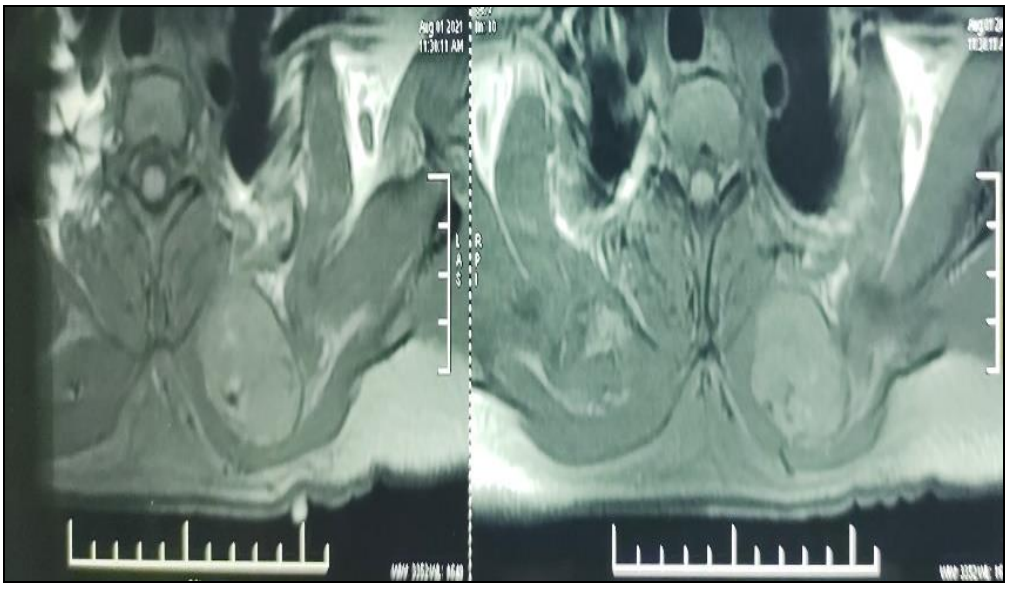

Fig 2a: MRI Showing single, focal, altered intensity lesion over right scapular region extending into the intramuscular planes

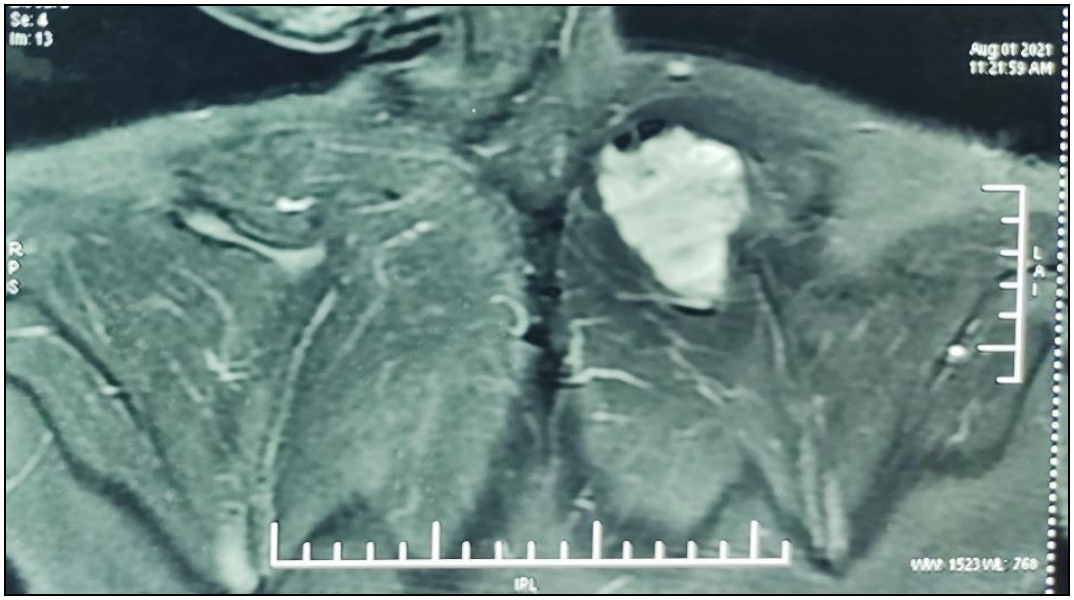

Fig 2b: MRI Contrast film showing single, focal, mainly hyperintense with few foci of hypo intense lesion over right scapular region

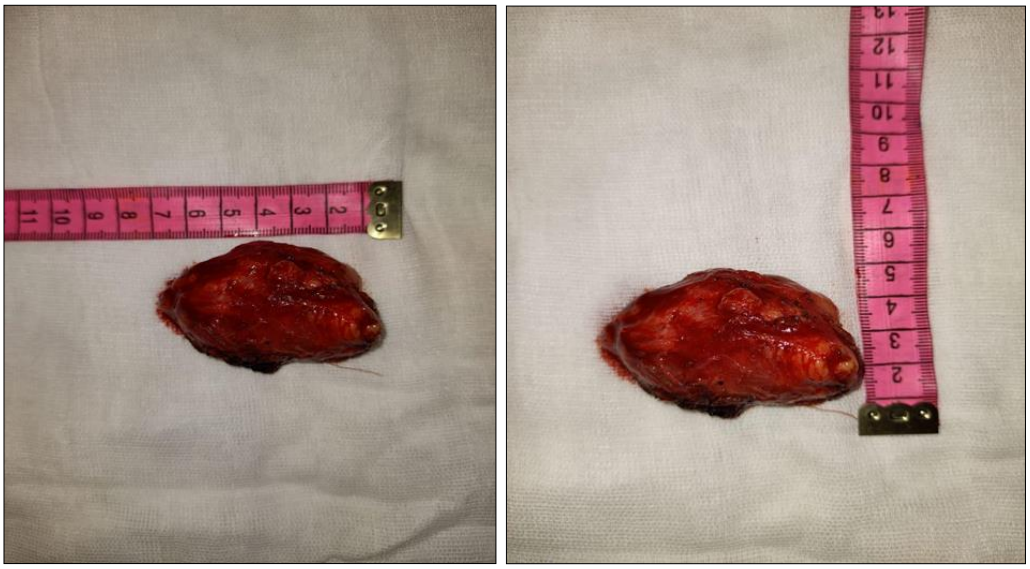

Fig 3a & 3b: Excised specimen of the vascular tumor showing well defined capsule all around with maximum dimension of 7cm and width of 4 cm

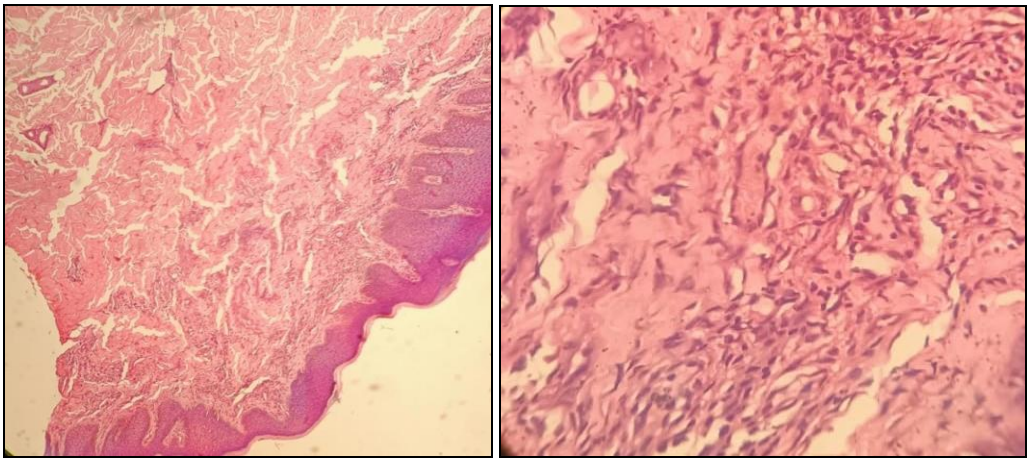

Fig 4a & 4b: Hematoxylin and eosin stain shows vascular proliferation lined with hobnail shaped endothelial cells in the upper dermis 4a- under 10X and 4b – under 40X magnification objective lese

Conclusion

To our best knowledge there are only 41 cases of retiform hemangioendothelioma reported in the literature till date. We would like to report our case of RH with its presentation, management to add in knowledge, in available literature for future reference to define its line of management and follow up.

Acknowledgement

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Conflicts of interest

Authors have no conflicts to declare.

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