Case Report

Retiform Hemangioendothelioma: A Rare Tumor in the Medial Canthus: Case Report and Review of Literature

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Abstract

Retiform hemangioendothelioma (RH) is a rare vascular neoplasm with intermediate malignant potential mostly occurring in extremities. It is important to differentiate this neoplasm from malignant conditions as RH has a good prognosis. As it has a tendency to recur locally, it may be misdiagnosed as a malignant tumor. Herein, we report a rare case of RH occurring in the medial canthus.

Keywords: Retiform hemangioendothelioma, medial canthus, CD31

Introduction

Retiform hemangioendothelioma (RH) is a rare tumor with only about 59 reported cases till date. The first case of RH was described in 1994 by Calonje *et al.*[1] as a vascular neoplasm with intermediate malignant potential and referred it to as low-grade angiosarcoma.

The most common site associated with RH is the distal extremities especially lower limbs.[2] It is rarely seen in unusual sites like penis, scalp, pleural nodule, and mons pubis. RH occurring in the medial canthus has been reported only in two cases earlier.

Herein, we report a case of RH occurring in the medial canthus, which is a rare site.

Case Presentation

A 36-year-old woman presented with a swelling involving the right medial canthus and the bridge of the nose for 1 year. Initially, the lesion was nodular, which progressively ulcerated and was associated with surrounding swelling for which the patient visited the outpatient department. The lesion measured 3×2 cm and appeared ulcerated with violaceous, erythematous inflamed margins and undermined edges with necrotic debris. The base of the lesion was erythematous and necrotic admixed with granulation tissue. The surrounding skin showed cellulitis with an expansion of the ulcer into the right orbit [Figure 1]. On the basis of appearance of the lesion, a clinical diagnosis of malignancy, most likely basal cell carcinoma, was made. The patient did not have any other significant medical history.

An incisional biopsy taken from the ulcer showed an infiltrative lesion in the mid and lower dermis, composed of numerous variably dilated to compressed, arborizing vascular channels [Figures 2 and 3]. The vessels were lined with endothelial cells exhibiting hobnailing, and the lesion resembled rete testis [Figure 4]. Occasional vessels showed pseudopapillary tufts within the lumen. Focally, solid nests of round-to-oval cells with pale eosinophilic cytoplasm were seen. Surrounding stroma showed neutrophils and lymphocytes. There was no significant atypia or mitotic figures. Papillary dermis was uninvolved. On immunohistochemistry, lesional cells were positive for CD31 [Figure 5]. On the basis of histopathological examination, a diagnosis of RH was made. The patient was advised radiology to determine the extent of lesion and was explained the possible role of surgery as the treatment. However, the patient refused any possible surgery on the face for cosmetic reasons and did not come for follow-up.

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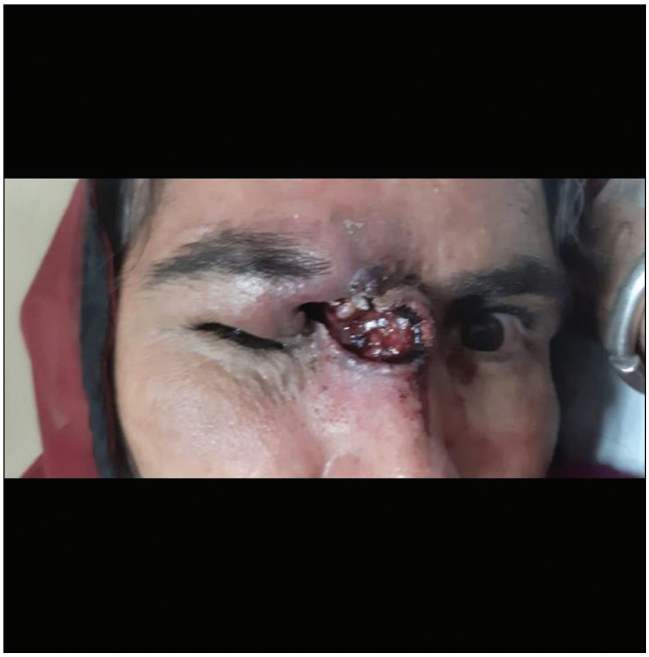

Figure 1: Gross clinical appearance of the lesion in the medial canthus of the right eye showing a nodular, ulcerated growth

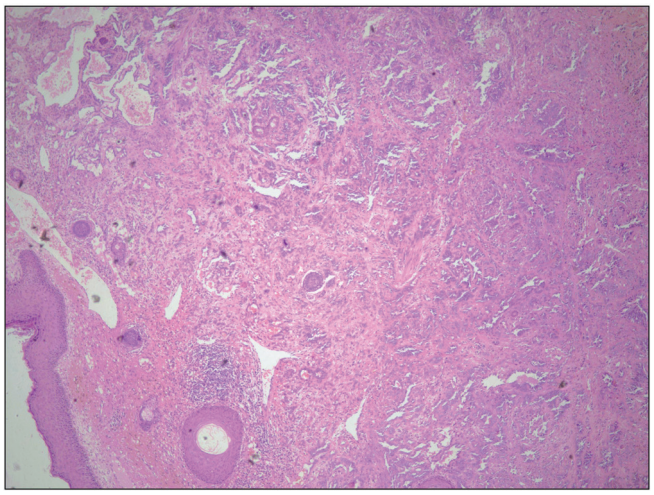

Figure 2: Biopsy showing an infiltrative lesion in the mid and lower dermis, resembling rete testis at scanner view (H&E, ×4)

Discussion

RH is a rare vascular neoplasm, first described by Calonje *et al.* in the year 1994 in a case series of 15 patients. World Health Organization (WHO) classification 2001 included RH as a rare entity with an intermediate malignant potential.[3] Since the first diagnosis of RH in the year 1994, very few cases have been reported in the literature [Table 1]. The etiological factors leading to the formation of RH have not been clearly established, but it has been associated with lymphedema, prior radiation exposure, nonepidermal malignant tumor, human herpesvirus-8, etc.[3]

RH involves a wide age group but predominantly occurs in adults during the second to fourth decade of life[4]

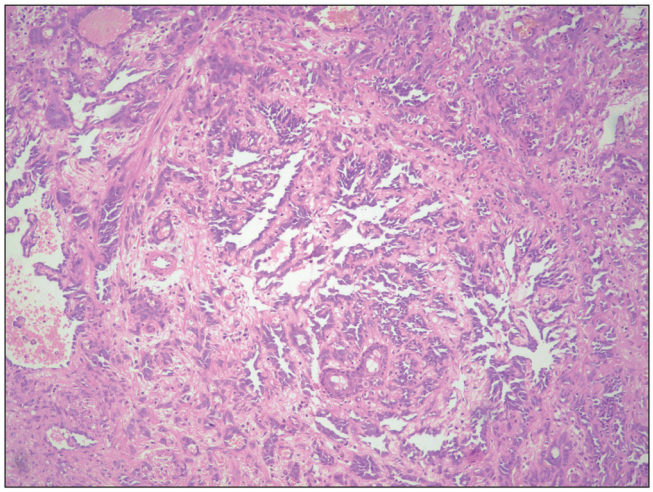

Figure 3: Low power view shows a vascular lesion composed of numerous, variably sized, arborizing vascular channels (H&E, ×10)

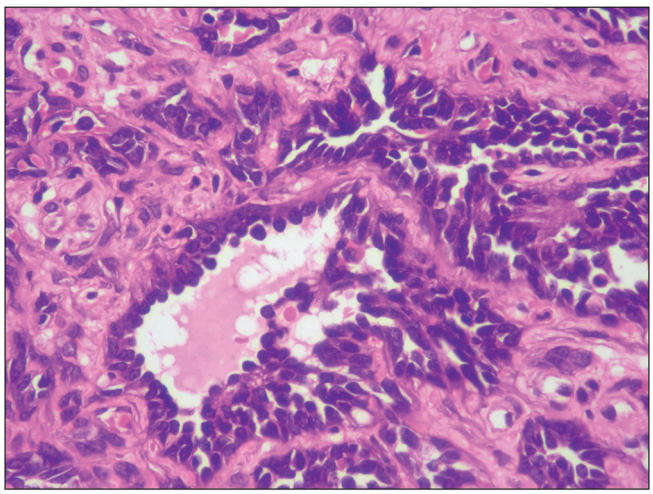

Figure 4: High power view shows the vascular channels lined with endothelial cells exhibiting hobnailing and absent to minimal cytological atypia. No mitoses were noted (H&E, ×40)

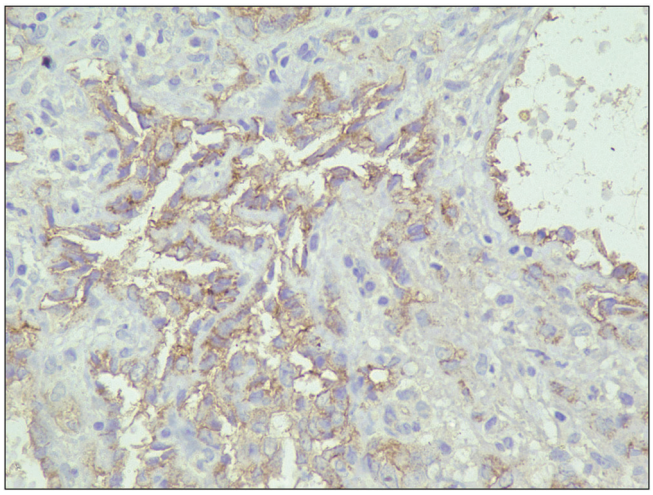

Figure 5: The lesional cells lining the channels show immunoreactivity for CD31 (IHC, ×40)

Table 1: Details of RH reported in the literature including the present case

S. no.	Study	Age	Site	Recurrence	Metastasis
1	Calonje *et al.* (1994)[1]	9–78/9F, 6M	Lower limbs (six), upper limbs (four), trunk (three), penis (one), scalp (one)/2.0–9.0 cm	8/15	1/15
2	Fukunaga *et al.* (1996)	75/F	Lower thigh/3.5 cm	Y	N
3	Duke *et al.* (1996)	30/F	Trunk, breast, and extremities	N	N
4	Dufau *et al.* (1997)	29–40/2F	Lower limb/1.0–1.5 cm	N	N
5	Sanz-Trelles *et al.* (1997)	11/M	Toe/1.5 cm	N	N
6	Mentzel *et al.* (1997)	40/F	Right great toe and right lower leg/ND	Y	N
7	Schommer *et al.* (2000)	73/F	Trunk/30 cm	Y	N
8	El Darouti *et al.* (2000)	32/F	Left upper thigh/3.5 cm	N	N
9	Ulrich and Hrynyschyn (2002)	17/M	Upper limb/0.5 cm	Y	N
10	Escudero *et al.* (2003)	31, 82/F	Sole, dorsum of hand	ND	ND
11	Aditya GS (2003)	36/M	Skull bone/5.0 cm	ND	ND
12	Tan *et al.* (2005)	19/F	Lower limb/4.5 cm	N	N
13	Botros *et al.* (2005)	30/F	Scalp	Y	N
14	Ioannidou *et al.* (2006)	55/F	Left middle finger/1.2 cm	N	N
15	Serel *et al.* (2007)	27/M	Left foot	N	N
16	Parson *et al.* (2008)[8]	17–71/F	Back, upper extremity, medial canthus, foot	ND	ND
17	Bhutoria *et al.* (2009)	35/F	Mons pubis/4.0 cm	Y	Y
18	Emberger *et al.* (2009)[9]	17/M	Back/2.5 cm	N	N
19	Kajo *et al.* (2009)	8/F	Calf	ND	ND
20	Zhang *et al.* (2010)[10]	61/F	Scalp and ear/8.0 and 2.0 cm	Y	N
21	Aydingöz *et al.* (2010)	60/F	Left ankle/6.0 cm	N	N
22	Hirsh *et al.* (2010)	44/M	Scrotum/NA	N	N
23	Keiler SA (2011)	11/F	Left fourth finger/0.7 cm	N	N
24	Albertini *et al.* (2011)	6/F	Neck/NA	N	N
25	O'Duffy *et al.* (2012)	18/M	Left pinna rim/NA	Y	Y
26	Choi *et al.* (2012)	20/M	Left index finger/1.5 cm	Y	N
27	Couceiro J (2013)	42/F	Small finger/1.0 cm	N	N
28	Mota A (2013)	26/F	Right flank/3.0 cm	N	N
29	Al-Faky *et al.* (2014)[7]	9/F	Right medial canthus/0.8 cm	N	N
30	Ezhil Jothi (2014)	58/M	Right ear/4 cm	N	N
31	Zheng LQ (2014)	71/F	Right foot/12.0 cm	Y	N
32	Ranga SM (2014)	9/M	Chest	Y	N
33	Tamhankar *et al.* (2015)[6]	48/F	Forehead	N	N
34	Liu *et al.* (2015)[5]	30/F	Pleural nodule	N	N
35	Kim *et al.* (2016)[2]	13/M	Buccal region	N	N
36	Seema Rani (2016)	23/F	Scalp/1–2 cm	N	N
37	Yang Tan (2017)	42/F	Left leg/1–3 cm	N	N
38	Pan *et al.* (2017)[3]	56/F	Left forearm and right neck/4.0 cm and 2.0 cm	Y	N
39	Shagufta Quadri (2018)	76/F	Right gluteal region/9 cm	N	N
40	Mondal *et al.* (2020)[4]	35/M	Postauricular/3 cm	N	N
41	Present case	36/F	Medial canthus		

F = female, M = male, N = no, NA = not available, ND = not documented, Y = yes

and shows a female predilection.[3] The most common location of this tumor is skin and subcutaneous tissue of the extremities, mostly the lower limb. Other rare sites reported included penis, scalp, pleural nodule, and mons pubis.[5,6] Two cases reported by Al-Faky *et al.*[7] and Parson *et al.*[8] demonstrated RH occurring in the medial canthus, similar to our case.

Clinically, RH usually presents as a painless reddish or bluish slow-growing, indurated nodule or papule. Though many lesions are solitary and painless, multiple, painful lesions are also known to occur.[7]

On histological examination, RH is characterized by elongated, arborizing vascular channels lined with uniform endothelial cells having a "hobnail" appearance. These vascular channels appear similar to normal rete testis. Stromal lymphocytic infiltration is often present.[4,6] Histopathological and immunohistochemical examination is important to make a definitive diagnosis, as the clinical features are nonspecific. The extensive sampling of this lesion is also important as RH can be a part of composite hemangioendothelioma consisting of varied components such as RH, epithelioid hemangioendothelioma, low-grade angiosarcoma, lymphangioma, and hemangioma.[5]

RH expresses immunoreactivity for endothelial markers such as CD31, CD34, factor-VIII, and Ulex europaeus agglutinin-1.[6] Few reported cases in the literature have shown RH to be immunoreactive for D2-40, which is a marker for endothelium of lymphatic vessels.[8,9]

Cutaneous angiosarcoma remains the main differential diagnosis of RH.[6] Angiosarcoma is an aggressive neoplasm having poor prognosis and high recurrence. Clinically, RH can have a variegated appearance, which may mimic angiosarcoma. On histology, angiosarcoma demonstrates marked atypia, mitotic activity, and particularly the absence of hobnail morphology.[10] Although an infiltrative pattern is common to both angiosarcoma and RH, vascular channels of angiosarcoma are more irregular than those of RH. Other differentials for RH include the vascular neoplasms: Dabska tumor and hobnail hemangioma. The Dabska tumor is papillary intralymphatic angioendothelioma and shares some histological features with RH including the presence of lymphocytes in vessel lumina. It is characterized by the presence of "hobnail" cells, irregular papillary endothelial projections and lacks arborizing rete testis-like architecture. The Dabska tumor mainly occurs in children with a tendency to occur in head and neck.[10] Targetoid hemosiderotic hemangioma (hobnail hemangioma) appears brown to violaceous papule with surrounding pale area and an ecchymotic rim, because of the deposition of hemosiderin. These lesions are histologically characterized by the presence of hobnail cells, but the lack of arborizing pattern.[3] Angiomatosis can also be considered differential diagnosis. It usually involves a large part and histologically shows large venous, cavernous, and capillary-sized vessels.[6]

Local recurrence is known to occur in RH, but distant metastasis is infrequent.[4] Wide local excision with free margins of 2–3 cm remains the treatment of choice for RH whenever feasible. Radiation therapy has also been reported to be successful for local as well as regional nodal cases.[1] One case has also been reported to be effectively treated with low-dose Cisplatin and moderate radiotherapy. It is difficult to establish a treatment guideline as of now because of scarce number of reported cases. Other treatment modalities of this tumor are still being investigated.

To conclude, RH is a rare vascular tumor with the medial canthus being even a rarer site. It can present in uncommon locations and can clinically mimic malignancy. Histopathological examination proves to be an important diagnostic tool in this regard.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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