CASE REPORT

Spindle cell hemangioma: Unusual presentation of an uncommon tumor

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ABSTRACT

Spindle cell hemangioma (SCH) is an uncommon tumor that usually presents as subcutaneous or deep dermal nodule affecting the extremities and is typically <2 cm in size. A few cases have been reported in the head and neck region. To the best of the authors' knowledge, there are no previous reports of SCH occurring in the orbit in the English literature. We, therefore, report the case of a large SCH involving the right orbit of a healthy 9-year-old Nigerian girl.

Key words: Orbit, Nigerian, spindle cell hemangioma

INTRODUCTION

Spindle cell hemangioma (SCH) is a distinct benign vascular lesion that typically occurs in the dermis or subcutis of distal extremities. Weiss and Enzinger[1] first described it in 1986 when the name spindle cell hemangioendothelioma was suggested as it was thought to represent an unusual form of low-grade angiosarcoma. However, studies[2-4] have shown SCH to be a benign, possibly reactive lesion thereby prompting its reclassification.

SCH may occur as a solitary or multiple syndromic or nonsyndromic lesion. It has no gender predilection and affects individuals of different ages.[5] Although SCH can occur in a variety of anatomic sites, very few cases have been reported in the head and neck region. Once diagnosed, treatment of choice is conservative excision with long-term follow-up. There is need to recognize this entity to avoid radical or aggressive treatment, especially when located close to vital structures such as the eye. To the best of our knowledge, there are no previous reports of SCH occurring in the orbit. We, therefore, report a case of solitary SCH occurring in the orbit that was managed effectively with conservative excision.

CASE REPORT

A 9-year-old female presented to the ophthalmology and maxillofacial clinics of the University College Hospital with 1-year history of slowly progressive painless swelling below the right eye. The swelling progressively increased in size followed by inability to open the eyelids with obscuration of the right eye.

Examination showed facial asymmetry due to a well-circumscribed swelling in the infero-lateral right orbit, extending to the zygoma and measuring about 8 cm by 6 cm in diameter [Figure 1]. Overlying skin was intact with no color alteration. The right globe was displaced supero-medially with moderate chemosis of the conjunctiva. There was restriction of ocular motility infero-laterally and slight reduction in vision (visual acuity 6/12). The swelling was lobular and firm in consistency.

Computed tomography scan showed a huge multilobulated heterogeneously enhancing mixed density mass in the infero-lateral aspect of the right orbit. The mass remodeled the orbital cavity and protruded beyond the orbital rim inferiorly, overriding the inferior aspect of the globe with minimal superior displacement and compression [Figure 2].

An incisional biopsy was done which showed a lesion composed of thin-walled vessels lined by flattened endothelial cells and between these vascular spaces were spindle, round

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to epithelioid cells which appeared vacuolated, within loose fibrous connective tissue. These features are suggestive of a SCH. CD34 immunohistochemistry was also done to mark vascular distribution [Figures 3-6].

She subsequently had surgical excision under general anesthesia, through an infraorbital incision, the lesion was exposed and dissected out en-bloc [Figure 7]. Postoperative period was uneventful and the patient was subsequently

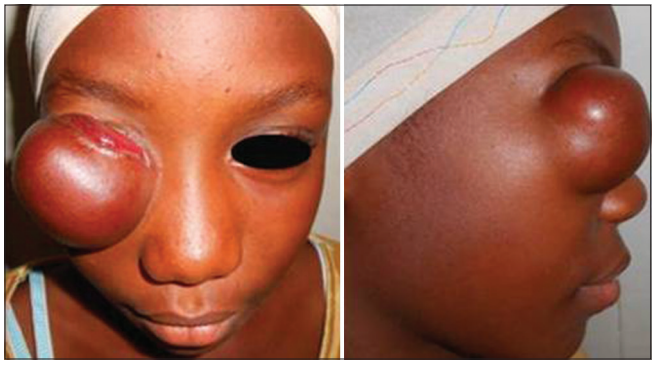

Figure 1: Clinical pictures showing the large well-circumscibed swelling in the inferolateral orbit

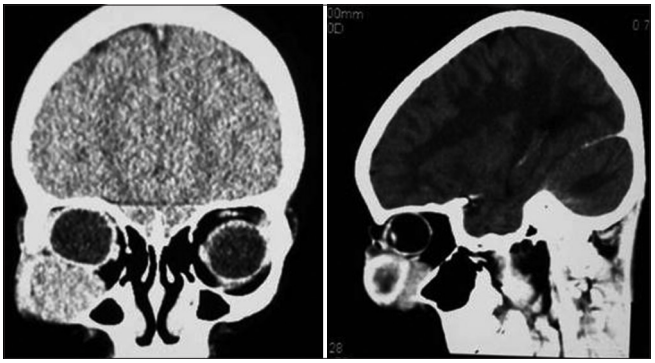

Figure 2: Computed tomography scan showing multilobulated heterogeneously enhancing mixed density mass in the inferior-lateral orbit

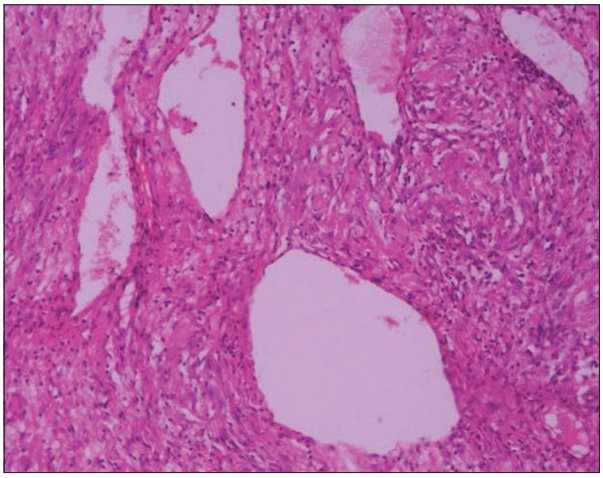

Figure 3: A section showing thin-walled cavernous vessels lined by flattened endothelial cells and containing a mixture of erythrocytes and thrombi (H&E stain, x100)

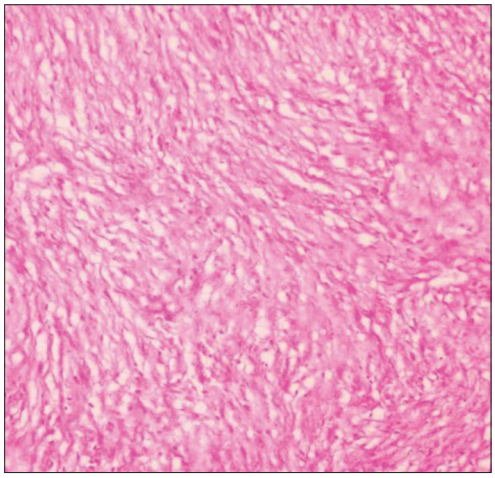

Figure 4: Photomicrograph showing multiple thin-walled vascular channels within spindled stroma that also appears vacuolated (H&E stain, x40)

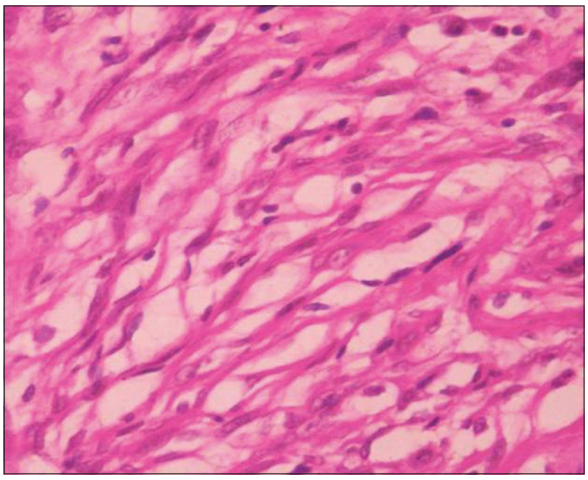

Figure 5: Photomicrograph showing spindled stroma that also appears vacuolated. Some epithelioid cells can be noted in the section (H&E stain, x200)

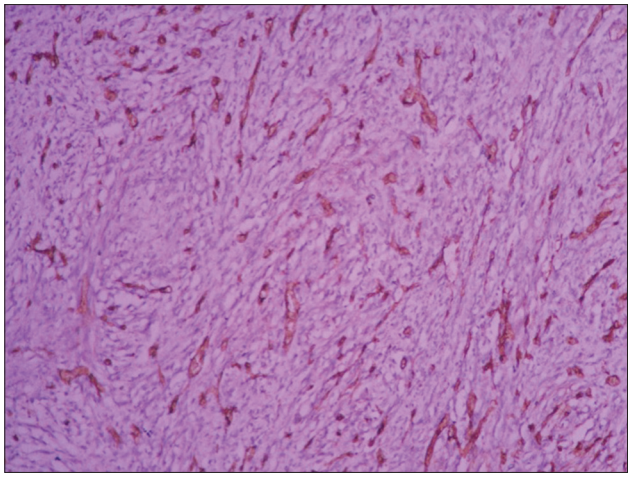

Figure 6: Photomicrograph showing +3 positivity for CD34 vascular immunohistochemical marker (IHC stain, x40)

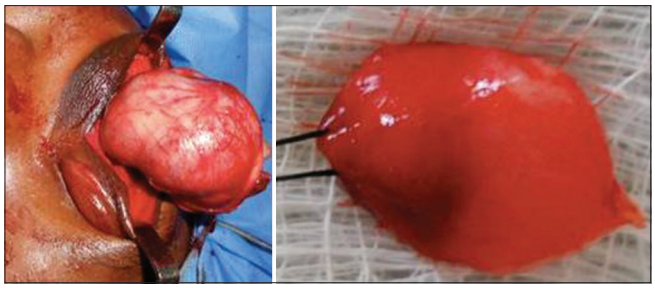

Figure 7: En-bloc dissection of the well-circumscribed mass

discharged under advice for regular follow-up visits. She has had 21 months follow-up with no evidence of recurrence and has corrected visual acuity of $6/6^{-3}$ in the right eye [Figure 8].

DISCUSSION

SCH can present as a solitary lesion as in our patient. Suranagi *et al*. reported that only 11 cases have been reported in the English literature in the head and neck region and none was within the orbit.[6] There is usually no associated discoloration of the overlying skin in SCH, as in our patient, making initial clinical suspicion of a vascular lesion difficult.[7] SCH generally occurs in the dermis or subcutaneous tissue and rarely occurs in the deep soft tissue.[8] In our patient, the deep soft tissue in the anterior orbit was involved with expansion of the orbital floor and displacement of the globe superiorly. SCH is usually asymptomatic in the early stage, but late presentation causing cosmetic disfigurement is a common indication for surgical treatment[7] as was in our patient. Although the pathogenesis and exact biologic behavior of SCH are still not clear, some authors have proposed that it may be a reactive or benign neoplasm,[8] while others have suggested thrombosis and subsequent recanalization in a preexisting vascular lesion.[2] Histologically, the spindle cells seen in both SCH and Kaposi sarcoma could pose a diagnostic challenge. However, cavernous spaces and epithelial vacuolated cells are usually not present in Kaposi sarcoma.[3]

Treatment options that have been used for SCH include surgery, systemic steroids, cryotherapy, laser therapy, radiation therapy, cytotoxic drugs, selective embolization and recombinant interleukin-2 with varying degrees of success.[7,9] SCH is a benign lesion and the most widely acceptable treatment option presently is conservative excision without adjuvant chemotherapy or radiotherapy. Following surgical excision, local recurrence rate of up to 58% has been reported.[8] Recurrences occur more commonly in patients with multiple lesions at presentation, occurring near surgical sites rather than within them.[8] It has been suggested that most of the recurrences represent multifocal tumors arising in the same anatomic region, possibly resulting from an underlying vascular abnormality or intravascular propagation of the lesion.[8] Radiation therapy is not advised for SCH because of the risk of malignant transformation.[7] SCH does not have capacity to metastasize and the only reported case of malignant transformation with metastasis reported in the literature is considered to be a radiation-induced sarcomatous transformation.[8]

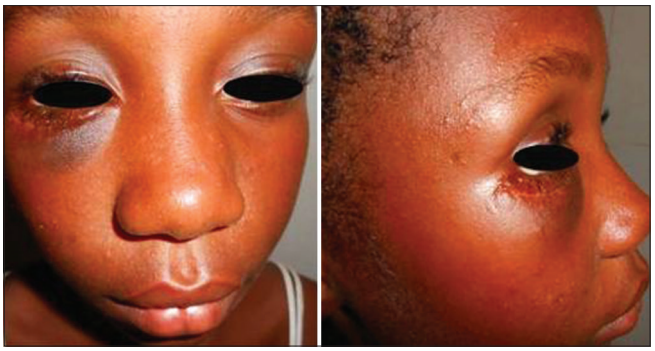

Figure 8: Clinical picture at follow-up showing no evidence of recurrence

CONCLUSION

We present a case of SCH occurring in the orbit that was completely excised surgically with acceptable aesthetics. Although the biologic behavior of this lesion is not yet fully known, it is a benign lesion and surgical excision remains the current acceptable mode of treatment. It is therefore extremely important that clinicians have a high index of suspicion and accurate histological diagnosis to avoid instituting other management modes that may worsen the outcome in the patient.

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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Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Weiss SW, Enzinger FM. Spindle cell hemangioendothelioma. A low-grade angiosarcoma resembling a cavernous hemangioma and Kaposi's sarcoma. Am J Surg Pathol 1986;10:521-30.
2. Imayama S, Murakamai Y, Hashimoto H, Hori Y. Spindle cell hemangioendothelioma exhibits the ultrastructural features of

reactive vascular proliferation rather than of angiosarcoma. Am J Clin Pathol 1992;97:279-87.
3. Tomasini C, Aloi F, Soro E, Elia V. Spindle cell hemangioma. Dermatology 1999;199:274-6.
4. Weiss SW, Goldbum JR. Benign tumours and tumour-like lesions of blood vessels. Enzinger and Weiss's Soft Tissue Tumors. 4th ed. St Louis: CV Mosby; 2001. p. 853-4.
5. Rokunohe D, Takeda H, Kaneko T, Aizu T, Akasaka E, Matsuzaki Y, *et al*. Spindle cell haemangioma and decorin expression. J Cosmet Dermatol Sci Appl 2012;2:8-10.
6. Suranagi V, Harugop SA, Bannur BH, Pilli SG, Mudhol SR. Spindle cell haemangioma of the nasal cavity: A rare tumor with unusual presentation. Clin Rhinol 2013;6:149-51.
7. Dhawan SS, Raza M. Spindle cell hemangioendothelioma. Cutis 2007;79:125-8.
8. Perkins P, Weiss SW. Spindle cell hemangioendothelioma. An analysis of 78 cases with reassessment of its pathogenesis and biologic behavior. Am J Surg Pathol 1996;20:1196-204.
9. Setoyama M, Shimada H, Miyazono N, Baba Y, Kanzaki T. Spindle cell hemangioendothelioma: Successful treatment with recombinant interleukin-2. Br J Dermatol 2000;142:1238-9.