Original Research Paper

Volume - 14 | Issue - 01 | January - 2024 | PRINT ISSN No. 2249 - 555X | DOI : 10.36106/ijar

Pathology

BLEND OF CAVERNOUS HEMANGIOMA AND ANCIENT SCHWANNOMA : A RARE COMPOSITE TUMOR OF ANKLE

Dr. Neelam Gupta	Professor and HOD, Department of Pathology, MMMC&H, Solan, H.P.
Dr. Anantika Thakur*	Assistant Professor, Department of Pathology, MMMC&H, Solan, H.P. *Corresponding Author
Dr. Karishma Sarin	Assistant Professor, Department of Pathology, MMMC&H, Solan, H.P.
Dr. Sanjeev Uppal	Professor, Department of Plastic Surgery, MMMC&H, Solan, H.P.

ABSTRACT Cavernous haemangiomas are vascular tumours that can occur in any organ system. Schwannomas are common benign peripheral nerve sheath tumours arising from differentiated Schwann cells. However, the concurrence of peripheral nerve sheath tumours and cavernous haemangiomas is rare. So far, only 40 cases have been reported in the literature. Most tumours documented to coexist with cavernous haemangiomas are Schwannomas and Neurofibromas. Herein, we discuss a case of a 48-year-old male patient who presented with a slow growing lesion in the right ankle region revealing composite differentiation of both cavernous haemangioma and ancient schwannoma. The aim of this case report is to make clinicians, pathologists and radiologists aware of such uncommon tumours to avoid potential pitfalls during diagnosis and treatment.

KEYWORDS : *Cavernous Haemangioma, Ancient Schwannoma, Ankle, Concurrence, Composite Differentiation*

INTRODUCTION

Cavernous haemangiomas are common vascular malformation that can virtually affect any organ in the body[1]. Schwannomas are also common benign peripheral nerve sheath tumours involving diverse locations, but the overwhelming majority of cases develop in subcutaneous tissue, or less often muscle, with a slight predilection for the distal extremities or head and neck region[1]. The combination of cavernous malformation and neoplastic lesions of nervous system occurring in the same patient is extremely rare. In the literature, a total of 40 cases have been published. Of these, only 24 cases documented the coexistence of schwannoma and haemangioma in the same lesion (conjoint association)[2]. Here, we describe the first case of cavernous haemangioma within an ancient schwannoma arising in soft tissue of ankle region.

CASE REPORT

A 48 year old male presented with the chief complaint of painless swelling over the right ankle region for the past 20 years which was gradually increasing in size. The general physical examination of the patient was unremarkable. Local examination of right ankle revealed a swelling measuring 3.5x2.5x1.5 cm which was soft to firm, mobile and non-tender. The overlying skin showed no visible distended vessels, active discharge or discolouration. On Ultrasonography, iso to hypo-echoic lesion measuring 2.3x0.7 cm in the subcutaneous plane revealing minimal vascularity on CDFI was seen. The lesion was in close proximity to the saphenous vein. A radiological diagnosis of nerve sheath tumour was made. A request for Fine Needle Aspiration Cytology was sent which revealed benign spindle cells lying in a loose fibrillary stroma. Cytological diagnosis of Benign Spindle cell lesion was given . Following this, local excision was done. We received a surgical specimen consisting of single well encapsulated grey brown soft tissue piece measuring 3.5x1.4x1.0cm. On cut section, central part of the lesion is cystic and was filled with grey brown to tan brown hemorrhagic material. A peripheral thin rim of grey white area was seen (Figure 1).

On microscopic examination, the periphery of the lesion revealed hypercellular and hypocellular areas suggestive of Antony A and Antony B. Verocay bodies were also identified (Fig 2). Areas revealing degenerative changes in the form of extensive hyalinisation, pigment laden macrophages, foamy histiocytes and nuclear atypia were seen (Fig. 3&4). The centre of the lesion revealed large vascular channels lined by endothelial cells along with haemorrhage and fibrin (Fig. 5&6). Immunohistochemistry (IHC) was then done. The schwannoma component showed spindle cells(schwann cells) which were positive for S100 (Fig. 7). The endothelial cells lining the cavernous spaces were positive for CD34 (Fig. 8). Based on the above histopathological and IHC findings , a final diagnosis of Cavernous haemangioma within an Ancient Schwannoma was made.

Fig. 1 – Grossly, the lesion is tan brown haemorrhagic in the centre and grey white in the periphery.

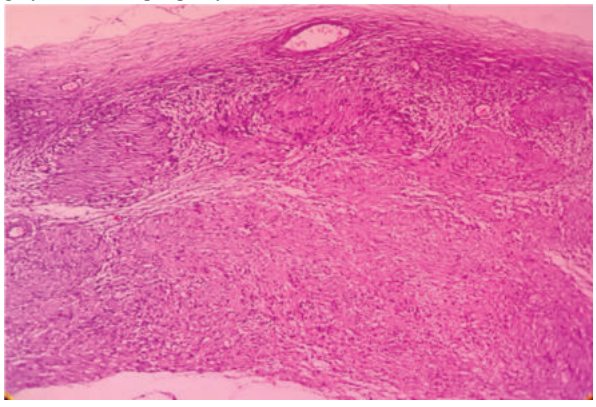

Fig.2 – Cellular areas revealing nuclear palisading around fibrillary process (Verocay bodies). 10x

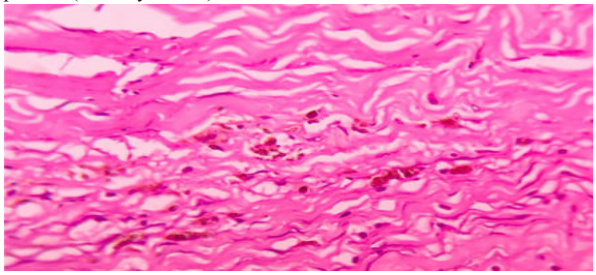

Fig. 3 – Areas with pigment laden macrophages are seen (feature of degeneration). 40x

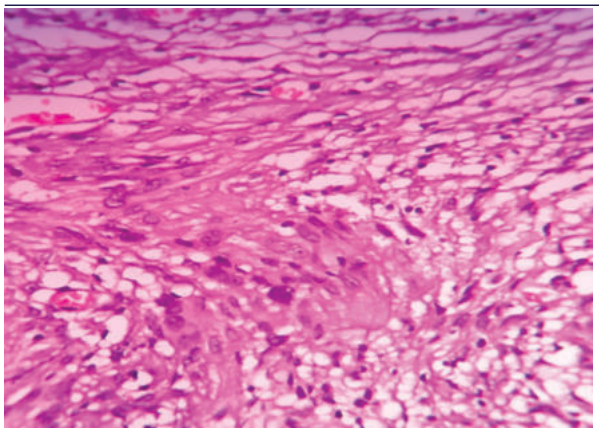

Fig. 4 – Focal areas revealing nuclear atypia are seen (feature of degeneration). 40x

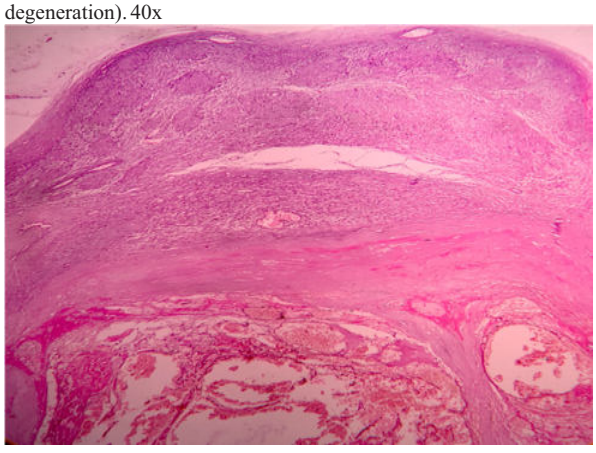

Fig. 5 – Peripheral areas of the tumour reveal spindle cell proliferation while the central area reveal dilated congested variable sized vascular spaces. 4X

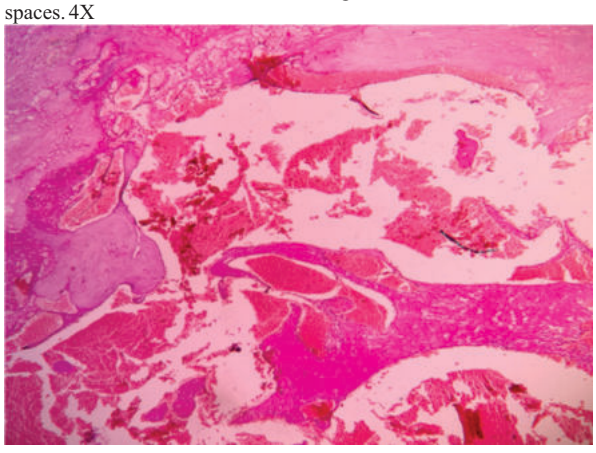

Fig. 6 - Variable sized dilated congested vascular spaces along with haemorrhage and fibrin deposition. 10x

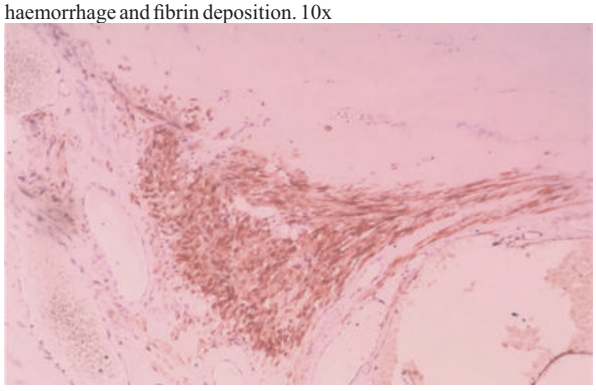

Figure 7- Spindle cells showing diffuse positive staining with S100. 10x

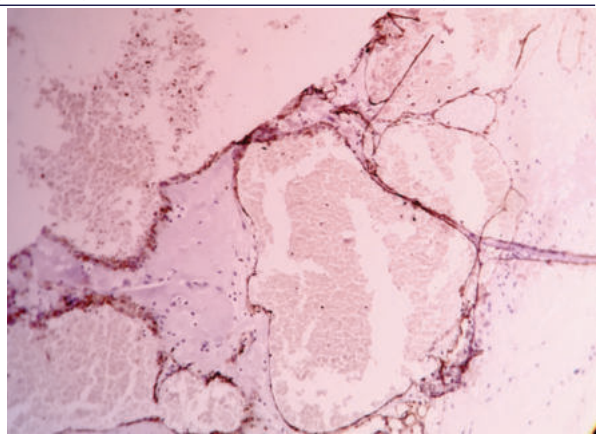

Figure 8- Endothelial cells lining the vascular channels are highlighted by CD34. 10x

DISCUSSION

Schwannoma, also called neurilemoma is a solid, slow growing benign tumour. It was first reported by Verocay in 1910 , and was named 'schwannoma' by Masson in 1932 [3]. These tumours account for about 5% benign tumours as revealed by an analysis of 18,677 benign soft-tissue tumours by Kransdorf and others [4]. It often occurs in the trunk, flexor side of upper and lower extremities, head and neck , mainly in the eighth cranial vestibular nerves , but is uncommon on the foot and ankle [3].

Grossly, schwannomas were round, ovoid, well circumscribed, encapsulated solid masses. The cut surface was often grey to yellow, solid, glistening. Degenerative changes in the form of cystic spaces and haemorrhage can be seen in some cases. Microscopically, the majority of schwannomas exhibited typical biphasic morphology, composed of hypercellular Antoni A and hypocellular Antoni B zones. Antoni A zone consisted of elongated spindle cells with indistinct borders palisaded together, forming a Verocay body, while Antoni B presented reduced cellularity in a myxoid matrix with increased vascularization . Degenerative features (ancient change) include cyst formation, haemorrhage, pigment laden macrophages and nuclear atypia. Immunostain show strong positive cytoplasmic staining for S-100 in spindle cells [1]. Excision is the surgical technique employed for the treatment of benign PNSTs.

Haemangiomas are commonly diagnosed soft tissue tumours in children and are clinically classified as capillary or cavernous. They account for 7% of all benign soft-tissue tumours in the general population. Haemangiomas most commonly occur in the area of the neck and head (60%), trunk (25%), and extremities (15%) [5]. Cavernous haemangiomas are less prevalent than the capillary type and are composed of large dilated vascular channels, with ill-defined edges involving deep structures like viscera, bone and muscles [6,7]. Lesions larger than 5 cm are more likely to be symptomatic and vulnerable to consequences, including bleeding and rupture [5].

Vascular malformations have traditionally been considered to be developmental anomalies.[8] However, a study done by Kim H et al pointed towards an acquired etiologic component, in which these may arise secondary to any number of "inciting event(s)" – ranging from traumatic insult to infection, irradiation and mechanical compression.[8,9]

In the case we discuss here, the coexistence of these pathologies in the same lesion raises question regarding their etiopathogenesis and if their concurrence is a result of similar course. Different theories have been proposed regarding the evolution of such mixed lesions. Kasantikul V et al elucidated the presence of both neurilemmoma and angioma components on the basis of their common origin in ectomesenchyme [10]. Rajkumar S et al did a comprehensive study to understand the molecular pathways which may lead to the coexistence of these pathologies. In their study they concluded that the association of cavernous malformations/haemangiomas with schwannomas might be related to the complex cross-talk between the common tumour-genetic signal pathways that are involved in schwannomas and haemangiomas. Schwannoma-secreted VEGF and angiopoietin-Tie-2 interactions act as the prime modulator in triggering and self-perpetuating complex signalling interactions between Schwann cells

and endothelial cells, which results in concomitant growth of schwannoma and haemangioma in the same tumor.[2]

An exhaustive search of literature yielded 40 cases showing an association between a nervous system tumour and haemangioma. The combination of schwannoma and haemangioma is rare. The cavernous malformation was found within the same tumours (conjoint association) in 29 cases and at different part of body (discrete association) in 11 cases. Schwannomas and neurofibromas were the most common peripheral nerve sheath tumour associated with cavernous malformations, accounting for 29 of 40 cases (72%). Majority (72%) of these tumours were in head and neck region while the remaining others were seen in the extremities and mediastinum.[2]

CONCLUSION

In summary, Schwannomas and hemangiomas are common benign tumours that can affect any organ of the body. Blend of these two lesions in the same tumour is uncommon with only a few reported cases. These tumours pose diagnostic challenges and require adequate sampling to be done by pathologists in order to avoid misdiagnosis. Surgical excision is the current mainstay of treatment however there is an increased risk of bleeding during the procedure.

Funding: No funding sources

Conflict Of Interest: None declared

Ethical Approval: Not required

REFERENCES

1. Fletcher C.D.M. Diagnostic Histopathology of tumors. 3rd ed. Philadelphia: Churchill Livingstone Elsevier;2007.
2. Ramkumar S. Reviewing Schwannoma-Hemangioma Composite Tumors With Their Tumorigenetic Molecular Pathways and Associated Syndromic Manifestations. Cureus 2021; 13:e19839.
3. Hao X, Levine D, Yim J, Qi C, Firestone L, Beiser I, Leone E, Woelffer K, Mirkin G. Schwannoma of Foot and Ankle: Seven Case Reports and Literature Review. Anticancer Res 2019; 39:5185-94.
4. Pilavaki M, Chourmouzi D, Kiziridou A, Skordalaki A, Zarampoukas T and Drevelengas A: Imaging of peripheral nerve sheath tumors with pathologic correlation: Pictorial review. Eur J Radiol 2004; 52:229-39.
5. Ramkumar S. Reappraising Schwannoma-Hemangioma Composite Tumors as Synchronous Tumorigenic Entities With Conjoined Histomorphology: A Case Report. Cureus 2021; 13:e20724.
6. Tripathi AC, Baitule RW, Rathod YB, Pundkar GN, Bhutada GP. A Case Report of an Unusual Case of Cavernous Hemangioma at Retrocalcaneum Region. J Orthop Case Rep 2022; 12:83-6.
7. Eyesan SU, Itie JC, Adesina SA, Kehinde GT, Olasode BJ. Cavernous Hemangioma of the Left Forearm. Rare Tumors 2015; 7:5895.
8. Byrne J V. Cerebrovascular malformations. Eur Radiol 2005; 15:448–52.
9. Kim H, Su H, Weinsheimer S, Pawlikowska L, Young WL. Brain arteriovenous malformation pathogenesis: A response-to-injury paradigm. Acta Neurochir Suppl 2011; 111:83–92.
10. Kasantikul V, Netsky MG. Combined neurilemmoma and angioma. Tumor of ectomesenchyme and a source of bleeding. J Neurosurg 1979; 50:81-7.