Plexiform Schwannoma Clinically Masquerading as Thrombosed Vein

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ABSTRACT

Plexiform schwannoma is a rare benign nerve sheath tumor usually located in trunk, head, neck, upper extremities and occasionally in lower extremities. It is usually less than 2 cm in size and generally affects young adults. The tumour is composed of schwann cells and exhibit a plexiform growth pattern both in gross or histologic examination and is frequently accompanied by multinodularity causing serpentine distortion. Plexiform neurofibroma commonly mimics plexiform schwannoma and it is important to differentiate between them, as recurrence and malignant potential are more common with plexiform neurofibroma. We present the case of a 14-year-old female with history of a nodular soft tissue mass over his right foot measuring 11 cm in length. A Doppler scan of the foot was suggestive of thrombosed superficial vein in right foot. The histological evaluation of the mass revealed Plexiform schwannoma. S-100 immunostaining is diffuse and strong confirming the diagnosis of Plexiform schwannoma.

KEY WORDS

Hematoxylin and eosin, Histopathology, Plexiform, Schwannoma

INTRODUCTION

Schwannomas also known as neurilemmoma are benign peripheral nerve sheath tumors of Schwanncell origin.[1] They comprise around 5% of all benign soft tissue tumors. Plexiform schwannoma, rare variant of schwannoma was first reported by Harkin and Reed in 1978 and accounts for about 5% all schwannomas.[2] This rare benign tumour is composed of Schwann cells and exhibit a plexiform growth pattern either in gross or histologic examination and is frequently accompanied by multinodularity causing serpentine distortion which gives the affected nerve segment appearance of "a bag of worms".[3,4] Plexiform schwannoma are usually solitary, however multiple tumors are seen in individuals with neurofibromatosis type II or schwannomatosis.[4] Plexiform neurofibroma commonly mimics plexiform schwannoma and it is important to differentiate between them, as recurrence and malignant potential are more common with plexiform neurofibroma requiring extensive surgery and rigorous follow-up.

CASE REPORT

A 14-years-old female presented to the surgery department complaining swelling in the right foot for 1 year with crampy pain over the right foot. Skin over the affected site was bluish in color. On local clinical examination there was a irregular nodular lesion over the right foot. A Doppler scan of the foot was suggestive of thrombosed superficial vein in right foot. Based on the clinical and radiological findings, the nodular lesion was excised with the provisional diagnosis of vascular lesion. The specimen was sent for histopathological examination. Gross examination showed a tubular nodular tissue measuring 11 cm in length (Fig. 1). External surface was smooth and cut section showed solid grey white area with no identifiable lumen. On histological examination an encapsulated tumor composed of spindle cells arranged in multiple nodules of varying sizes (plexiform architecture) was seen (Fig. 2a). These spindle cells had wavy serpentine nuclei arranged in a palisading manner at places forming verocay bodies (Fig. 2b). Tumor showed more hypercellular

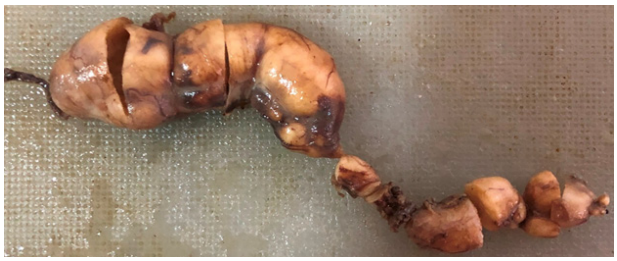

Figure 1. Gross examination shows nodular tissue measuring 11 cm in length (Fig. 1). External surface was smooth and cut section showed solid grey white area.

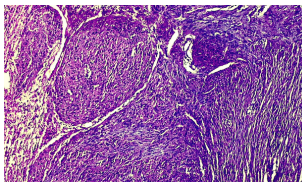

Figure 2a. H&E sectionsshow tumor cells arranged in multiple nodules of varying sizes (20X).

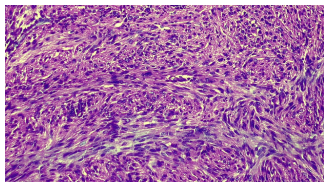

Figure 2b. H&E sections showtumour composed of spindle cells arranged in fascicles inpredominantly hypercellular Antoni A areas with nuclear palisading. Nuclei are elongated wavy and show mild nuclear atypia (40X).

(Antoni A) areas. Nuclear atypia, increased mitosis and necrosis are not seen. On immunohistochemical staining, S100 protein was strongly expressed in tumor cells (Fig. 3). Based on the characteristic microscopic findings and immunohistochemistry report, a diagnosis of plexiform schwannoma was made.

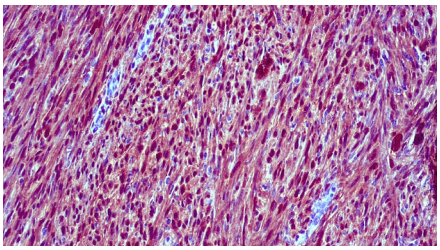

Figure 3. S100 stain demonstrates diffuse strong positive staining of tumor cells.

The patient was reexamined for presence of multiple tumour and evaluated for family history, history of trauma, and other associated syndromes like Gorlinkoutlas syndrome, schwannomatosis. He had no predisposing factors. Patient on follow up of 1 year after surgery is doing well without any recurrence.

DISCUSSION

Schwannomas are benign peripheral nerve sheath tumors of Schwann cell origin.[1] These tumours are common soft tissue tumour with characteristic histopathological features. Most cases of schwannomas occur sporadically and few cases occur in association with multiple meningiomas, Neurofibromatosis type 2 and schwannomatosis and rarely with Neurofibromatosis type 1.[3] Different variants of schwannomas includes Ancient Schwannomas, Cellular Schwannomas, Plexiform Schwannomas, Melanotic Schwannomas, Epithelioid Schwannomas, Hybrid Schwannomas, Reticular Schwannomas, glandular Schwannomas and lymphnodeSchwannomas.[4]

Plexiform schwannoma, rare variant of schwannoma was first reported by Harkin and Reed in 1978 and accounts for 5% all schwannomas.[5,6] Plexiform schwannoma are usually solitary, however multiple tumors are seen in individuals with neurofibromatosis type II, Gorlinkoutlas Syndrome or schwannomatosis.[7] Trauma may play a role in the formation of this tumor. Our patient did not have any presdisposing factors. Plexiform schwannomas are usually superficial and deep seated lesions are quite infrequent.[4] Most common location for this tumour are trunk, head, neck or the upper extremities and occasionally lower extremities.[8,9] The location of the tumor in this case is similar to as reported in literature. These tumors when present as a superficial nodules especially in children can mimic the appearance of vascular lesion. Similar to our case Lo et al. and Mridha et al. had case reported as vascular lesion which was histologically plexiform schwannoma.[10,11]

Most plexiform schwannomas reported measure less than 2 cm and are slow growing.[4] We present a case of swelling measuring 11 cm in the right foot. Usually a congenital plexiform schwannomas arise in lower limbs and are larger in size.[7,10,11] Our patient was 14 years old and had the lesion since approximately for 1 year.

This rare benign tumour is composed of Schwann cells and exhibit a plexiform growth pattern either in gross or histologic examination and is frequently accompanied by multinodularity causing serpentine distortion which gives the affected nerve segment appearance of “a bag of worms”.[6,7] The tumour is composed of tortuous mass histologically composed of multiple discohesive nodules composed mainly of Antoni A-type tissue. Histologically this tumour must be distinguished from plexiform neurofibroma and malignant peripheral nerve sheath tumour (MPNST). Plexiform neurofibroma consists of grossly expanded nerve or nerve fibers replaced by neurofibromatous tissue and marked myxoid change.[9] MPNST histologically shows nuclear atypia, nuclear overcrowding, necrosis with marked mitotic activity.[2,9] MPNST is a malignant tumour requiring wide excision and plexiform neurofibroma has a more tendency for malignant transformation requiring extensive surgery and rigorous follow-up.[2,8] However, plexiform schwannoma’s tendency for malignant transformation has not been described and local recurrence is also very infrequent.[8,9] Local excision is treatment of choice in most of the cases of plexiform schwannoma.[8]

Plexiform Neurofibroma commonly mimics plexiform schwannoma and it is important to differentiate between them, as recurrence and malignant potential are more common with plexiform neurofibroma requiring extensive surgery and rigorous follow-up.

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