

Superficial Plexiform Schwannoma - a report of three cases with review of literature.



Medical Science

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ABSTRACT

Plexiform schwannomas are rare variants of schwannoma, which often occur sporadically. They have also been described to occur in individuals with NF-2 and schwannomatosis. Recurrences as seen in one of our cases are known to occur when these lesions are incompletely excised. This clinical presentation should not be mistaken for malignant transformation, which is often seen in cases of plexiform neurofibromas, which are markers of NF-1.

Introduction :

Plexiform schwannomas are rare benign tumors, often occurring as sporadic lesions though a few cases have been reported in association with Neurofibromatosis -2(NF-2) and Schwannomatosis. [1] They may mimic plexiform neurofibromas (NF-1) by way of recurrence from which they need to be differentiated, as they do not carry the risk of malignant transformation that is seen with NF-1.

Case report:

Case : 1 A 74 yr old male presented with a recurrent painless soft tissue swelling over upper half of the right pinna of 3yrs duration. The tumor was excised and sent for histopathological examination. We received a partly skin covered, nodular, soft tissue mass of 7x5x4 cms. Cut section of the mass revealed multiple well defined solid and grey white nodules in the subcutis.

(Figure -1 here)

Past history of the patient revealed that he had a similar lesion of about 5x 3x 3 cms excised 6yrs back, the details of which were not known.

Case 2: A 24 yr old female came with swelling in the lower part of left side of neck of 2 yrs duration. FNA was reported as a spindle cell lesion. The mass was excised and sent for HPE. We received a 4.5x3.0x1.5 cm grey white, nodular soft tissue mass.

Case 3: A 25 yr old male presented with swelling left side of neck of one and half years duration. Grossly we received a 2x1x-1cm, grey white nodular mass.

Histopathology of all the three cases showed well defined cellular nodules composed of elongated spindle shaped cells with eosinophilic cytoplasm and elongated wavy nuclei showing nuclear palisading at places -suggestive of Antoni A areas. The nodules were surrounded by a capsule and were separated by fibrous stroma.

(Figure-2 here)

A Diagnosis of Plexiform schwannoma was made in all the three cases.

Discussion:

Plexiform schwannomas are rare benign peripheral nerve sheath tumors that mostly affect young adults and occur commonly as solitary, slow-growing, asymptomatic nodules. They are sporadic tumors located in the dermis or subcutaneous tissue, but deep seated and multiple lesions have also been described.[2] Though rare in patients with NF- 1, they have been reported in individuals with NF-2 and Schwannomatosis.[3]

Histopathologically, they show multiple nodules composed of hypercellular Antoni A areas predominantly , though they may show both hypo and hypercellular areas like conventional Schwannomas. [4] They do not exhibit significant mitotic activity, atypia or necrosis.

Immunohistochemically the tumor cells are positive for S100 with the intervening stroma showing positivity for laminin and collagen type IV.[4]

The differential diagnoses include plexiform neurofibromas, Schwannomas with intralesional nodularity ,Neurothekeoma and MPNSTs. [4]

Plexiform neurofibromas are more common and are pathognomonic of Neurofibromatosis of Type1. Morphologically they show mostly hypocellular areas with myxoid stroma in the background. Some of these lesions may show highly cellular areas with schwannian nodules mimicking schwannomas.

Such cases need to be differentiated with the help of immunohistochemical stains for S100 protein, neurofilament, laminin and collagen Type-IV, as plexiform neurofibromas carry risk of malignant transformation unlike Plexiform schwannomas [5]

Schwannomas with intralesional nodularity can be differentiated from Plexiform Schwannomas by the presence of loose Antoni B areas. Immunohistochemical stains also help in distinguishing the lesional nodules from intervening stromal tissue which is seen in Plexiform schwannoma.

Neurothekeomas also show nodular growth pattern with loose myxoid stroma, but the absence of cellular areas helps them in differentiating from schwannomas. Absence of S100 staining and positivity for melanoma specific antigen NKI/C-3 in cellular neurothekeoma are helpful in differentiation.[4]

PS can be mistaken for MPNSTs, when they are associated with marked cellularity, mild atypia and proliferative activity or recurrence. Immunohistochemistry with S100 protein helps differentiate these two.

Recurrences in these tumors are said to be due to incomplete excision, lack of thick encapsulation or irregular growth of tumor. Recurrence in our first case could probably be due to incomplete excision. [5, 6]

At the molecular level, these tumors show loss of NF2 gene which codes for the protein product merlin, whose function is not known exactly but is thought to regulate linkage between cytoskeletal proteins and cell membrane. [3]

Though not pathognomonic, when multiple they may be the external cutaneous manifestations of NF-2 or Schwannomatosis, warranting further work up.

Images



Figure-1: Cut section of the gross specimen from case 1 showing a partly skin covered nodular lesion.

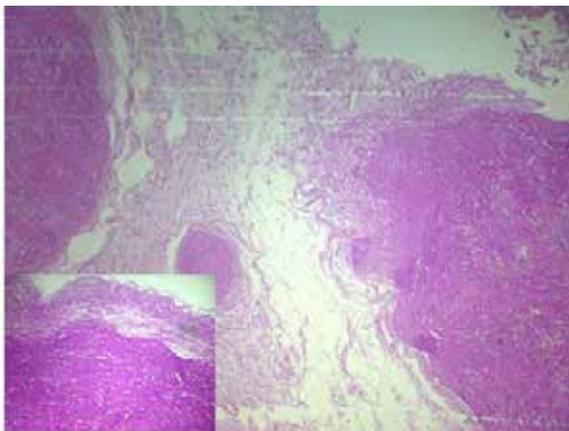


Fig-2: H&E stain, x40x, showing a cellular nodule with foci of nuclear palisading.

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