Plexiform Shwannoma: A Case Report and A Review of Literature

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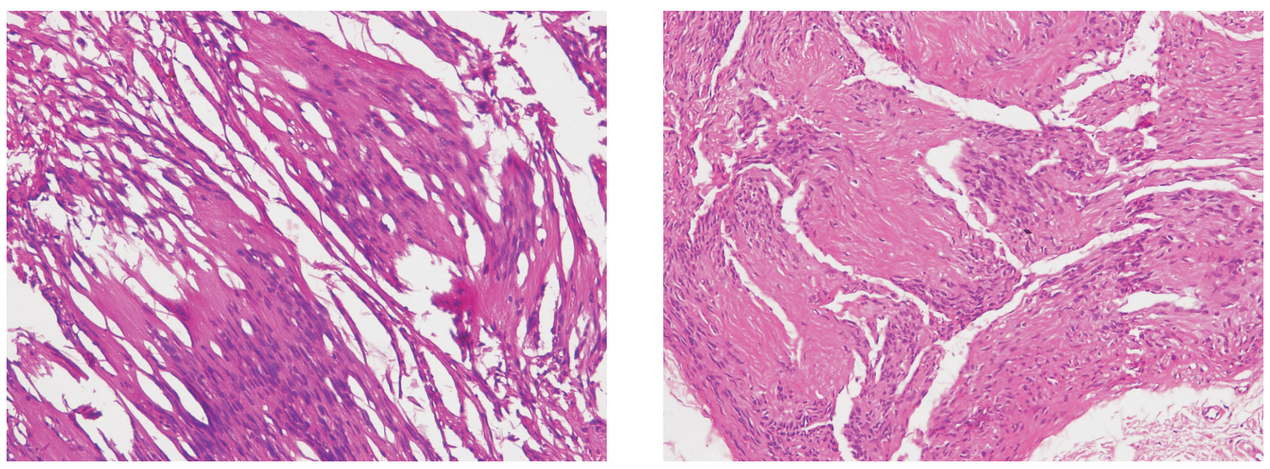
INTRODUCTION:

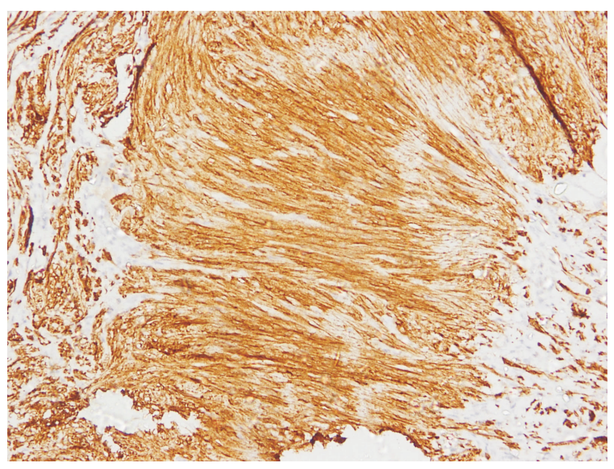
schwannomas are benign peripheral nerve tumors , occuring in third to fifth decad of life and presenting as solitary tumor mostly in head and neck region followed by upper and lower extremities(1) . they account for 8% of the tumors of soft tissue and are composed entirely of schwann cells(2).Various variants of schwanomma have been described as; cellular, plexiform, ancient and psammomatouse. plexiform schwanomma is an unusual varient of schwanomma which represents 3.4% of all schwannomas(3). plexiform schwannoma is a very un usual and rare varient of schwannoma. it mostly occurs in head and neck and extremities of young adults. it presents as solitary nodule in subcutaneouse or superficial areas however rare cases also occur in deeper issue and in muscle. it present as solitary mass or nodule in sporadic cases and as multiple nodules in neurofibromatosis 2. we presen a case of plexiform schwannoma ocuring in young male as recurrent index finger mass

CASE PRESENTATION:

A15 years old male presented with swelling over right index finger. The swelling was painful. He had previous history of same swelling in same place about two years ago which was excised. Previouse biopsy report was not available. No radiological data was provided. Patient had no family history of any disease or co morbidity.
We received multiple whitich tissue fragments measuring 4.5x3.0x1.1 cm in aggregate. On microscopic examination multiple encapsulated nodules were seen separated by thin fibrouse septae. these nodules were composed of spindle shaped cells having wavy nuclei. there were multiple verocay bodies along with congested blood vessles.
Immuno histochemical stain S-100 was applies which showed diffuse positivity. Based on morphological and immunohistochemical features diagnosis of plexiform schwannoma was made.



Histological examination shows multiple nodules of spindle cells



S-100 Immunostain

DISCUSSION :

Plexiform schwannoma is an un usual and rare variant of schwannoma which accounts for 4.3% of all schwannomas. its association is with schwanommatosis ,which is defined as two or more clinically proven schwanommas . least frequently it is also associated with neurofibromatosis-2(NF-2).
It mostly presents in sporadic cases with superficial or subcutaneouse nodule/nodules which is painful or may be assymptomatic. On histological patterns it shows plexiform growth pattern and shows diffuse positivity for S-100 protein. Plexiform schwannomas are located in skin and superficial dermis while deep are located in retroperitoneum,deep in trunk and extremities, parotid, vulva and mediastinal esophagus. Prognostically superficial schwannomas can be excised completely and recurrence is very low while deeply located have high rate of recurrence(about50%). While superficial show normal growth pattern ,deep may have some worrisome features like increased cellularity , pleomprphism and at times mitotic figures. Despite these features it is unlikely for a schwannoma to show malignant behavior or transform into its mailnant counterpart. More importantly differential diagnosis of this entity should be in mind which include plexiform neurofibroma and malignant peripheral nerve sheath tumors. Difference with neurofibroma is important as it has strong association with neurofibromatosis. Like neurofibroma it has plexiform multinodular architecture but neurofibromas show serpentine nuclei and occasional nuclear palisading . neurofibroma shows patchy positivity for s-100 while schwannomas are diffusely positive for this. Second most common differential is malignant peripheral nerve sheath tumor when schwannoma is deeply located and have worrisome nuclear and cellular features as described above. Extensive sampling to search schwannoma and negative immunoreactivity for S-100 rules out the diagnosis.
Schwannomas have excellent prognosis overall. They show trisomy 17 and 18 and have no syndromic association .

CONCLUSION:

Schwannomas are benign tumors of schwannian differentiation. Plexiform schwanomma can be confused with neurofibroma which has syndromic association and with malignant peripheral nerve sheath tumor. Knowledge about different variants of schwannoma , carefull sampling ,history and immuno studies are helpful in making diagnosis .

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