Case Report

Neuroblastoma-like schwannoma in a case of schwannomatosis: Report of a rare case

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Abstract

Schwannomatosis is a term used to describe patients with multiple nonvestibular schwannomas with no other stigmata of neurofibromatosis type-2 (NF2). Neuroblastoma-like schwannoma is a rare subtype of schwannoma, with histological features resembling a neuroblastoma. This case is probabaly the second case of very uncommon neuroblastoma-like schwannoma, in a patient of schwannomatosis.

Key words: Neuroblastoma-like, benign peripheral nerve sheath tumour, rosette.

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Introduction

Schwannomas are benign, solitary peripheral nerve sheath tumours. They can present as typical schwannoma with Antoni A and B areas or rare forms like ancient schwannoma, epithelioid schwannoma, and neuroblastoma-like schwannoma. [1] Cellular and ancient schwannomas are confused with sarcomas, while neuroblastoma-like schwannoma can be misdiagnosed as round cell tumour. [2]

Schwannomatosis is defined as the presence of two or more nonintradermal schwannomas with no evidence of vestibular tumour and no known constitutional NF2 mutation in a patient >30 years of age. [3] It is very rare, having an annual incidence of 0.58 cases/1 million persons withapproximately 20% of cases being familial. [4]

Neuroblastoma-like schwannoma is a rare variant of schwannoma defined by the presence of large, collagenous, rosette-like structures surrounded by small, round, hyperchromatic Schwann cells. Literature search revealed only single report of neuroblastoma-like schwannomatosis. [4]

Case history

A 19 year old woman presented with ten years history of painless, multiple subcutaneous nodules over both upper limbs, lower limbs, abdomen and trunk. (Fig.1)

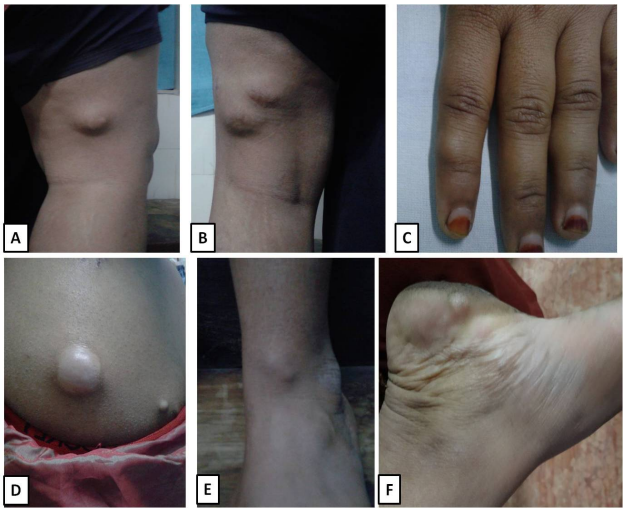

Fig 1: Clinical photograph showing multiple subcutaneous swellings on lower and upper extremities and abdomen.

The largest was over trunk and measured 3X2X2 cm. There were no abnormal neurological signs. Patient showed no evidence of neurofibromatosis.There was no family history of similar lesions. Magnetic resonance imaging brain showed no evidence of vestibular tumour. There was history of excision of two swellings three years back, reported outside as small round cell tumour and benign fibrous histiocytoma. Two swellings, (on the trunk and over anterior aspect of left ankle joint), were excised. Grossly both the masses were nodular, measuring 3X2X2cm and 1.5X0.7X0.4cm respectively. The cut surface was shiny whitish.

Microscopy of the first mass showed conventional schwannoma. Microscopy of the other mass showed a thinly encapsulated tumour composed predominantly of large rosettes arranged radially around central collagenous cores surrounded by small ovoid to spindlecells with rounded hyperchromatic nuclei, and scant cytoplasm. (Fig. 2) In areas, slender, wavy spindle cells arranged in loose background (Antoni B areas) were also present. The diagnosis of Neuroblastoma-like schwannoma was given.

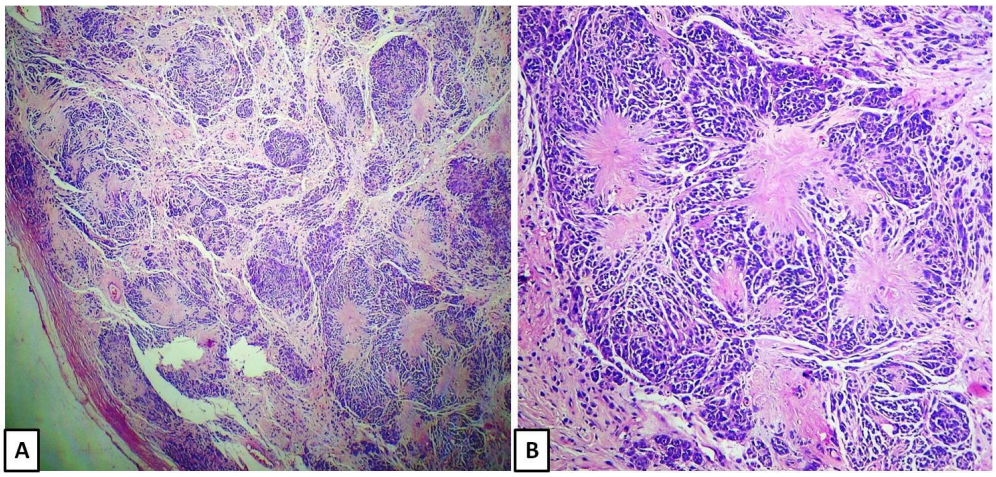

Fig 2: A. Photomicrograph showing thinly encapsulated tumour. (H&E, X100)
B. Photomicrograph showing tumour composed of large rosettes arranged radially around central collagenous cores surrounded by small ovoid to spindle with rounded hyperchromatic nuclei, and scant pale cytoplasm. (H&E, X400)

Immunohistochemistry of both the masses showed strong and diffuse positivity for S 100 protein. Synaptophysin and CD99 were negative. MIB1 labelling index was 5%. (Fig 3)

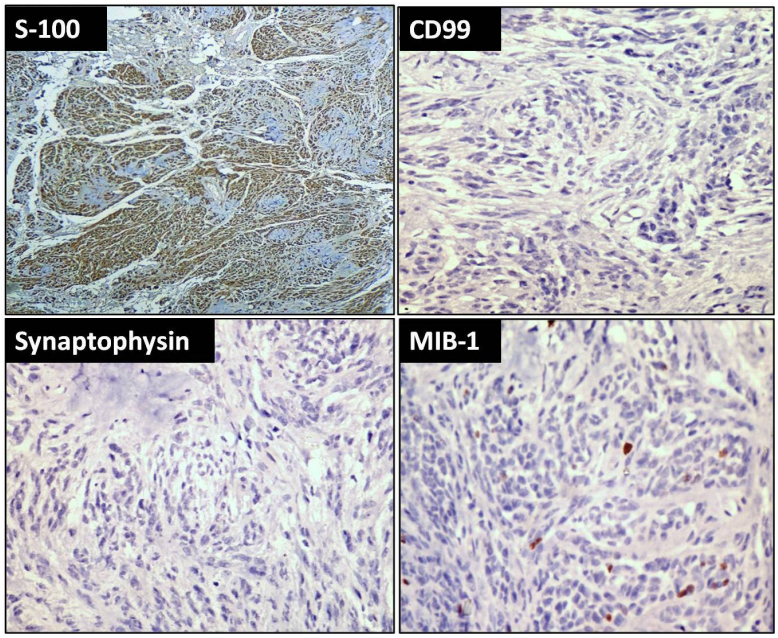

Fig 3: Immunohistochemistry: The tumour cells expressed S100 protein (X40), and were immunonegative for CD99 and synaptophysin. MIB1 was low (5%).(X400)

Discussion

Schwannomas are benign peripheral nerve sheath tumours that have predilection for head, neck and flexor aspects of extremities. These are usually solitary sporadic lesions or can be multiple when syndromic, occurring in association with neurofibromatosis or schwannomatosis.

Schwannomatosisis currently considered a separate entity not associated with germline mutations of NF1 or NF2. [3] It is defined as presence of multiple nonintradermal schwannomas with the absence of vestibular schwannomas, the key feature of NF2. Itmay be associated with germline mutations of the *SMARCB1/INI1* gene. [4] MacCollin et al first proposed the diagnostic criteria for schwannomatosis which were modified and the current working clinical criteria were proposed. [5] Patients can be diagnosed with schwannomatosis if theyhad two or more non-intradermal schwannomas, at least one with histologic confirmation with no evidence of vestibular tumor and no known NF2 mutation, at an age > 30 years or one pathologically confirmed nonvestibular schwannoma and 1st degree relative who meets above criteria. A possible diagnosis may be made if the patient has similar tumors at age < 30 years, or in patient with age > 45 years with no clinical symptoms of eighth nerve dysfunction. Segmental schwannomatosis is the term used when schwannomatosis is limited to one limb or five or fewer contiguous segments of the spine. [3, 6]

Pathologically, schwannoma is an encapsulated tumour composed of alternating Antoni A and Antoni B areas that may blend imperceptibly or change abruptly. Varying degree of nuclear palisading, Verocay bodies and large, gaping thick walled vessels are characteristic. Rarely schwannomas contain a significant population of small lymphocyte-like Schwann cells arranged around collagen nodules forming giant rosettes or around vessels forming perivascular rosettes, referred to as NB-like schwannoma. [3]

NB-like schwannoma, originally described by Goldblum et al, is a rare variant of schwannoma histologically mimicking neuroblastoma or ependymoma. Two out of three cases they described resembled neuroblastoma with Homer-Wright rosette-like structures whilethe third case showed perivascular ependymomatous rosettes. [1]

The small hyperchromatic Schwann cells and rosettes may create confusion with primitive neuroectodermal tumors (PNETs) or neuroblastoma. Homer-Wrightrosettes in neuroblastoma are smaller with Neuron Specific Enolase positive fine fibrillary substance in the centre. PNET shows strong and diffusemembranous positivity for CD99. [2]

In summary, we report a case of an extremely uncommon variant of schwannoma with distinct histology which may be confused with neuroblastomaor PNET but could be diagnosed on the basis of the identification of areas of conventional schwannoma and positivity for S-100 protein.

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