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Original Research Article

Neuroblastoma like Schwannoma: A Diagnostic Dilemma!!- A Rare Case Report

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Abstract:

Neurilemmoma or schwannoma is a common benign nerve sheath tumor composed of spindle cells. They show immunohistochemical and ultra-structural features similar to that of Schwann cells. Although typical cases are easily recognized, certain rare variants of schwannoma do pose a diagnostic challenge for pathologists. We discuss here in a case of a 40 year old male who presented with a scalp mass since 6 months, and was diagnosed as neuroblastoma-like schwannoma based on characteristic histomorphological findings and Immunohistochemistry.

Keywords: Neuroblastoma, Rosette, Schwannoma.

Introduction

Schwannomas are benign, solitary peripheral nerve sheath tumours. They are encapsulated and can present as typical biphasic pattern comprising of a highly ordered cellular component with nuclear palisading forming verrocay bodies (Antoni A) area and a loose myxoid component (Antoni B) area [1] that blends imperceptibly and change abruptly.

Few rare, rather unusual forms of schwannomas include the ancient schwannoma, plexiform schwannomas, epithelioid schwannoma, and neuroblastoma-like schwannoma. [2] Cellular and ancient schwannomas are often confused with sarcomas, while neuroblastoma-like schwannoma can be misdiagnosed as small round cell tumours [3].

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.

This case report deals with this unusual variant of schwannoma, its differential diagnosis along with review of literature.

Case Report: A 40 year male presented with a scalp mass since past 6 months. The swelling was nontender, gradually progressive and it was cosmetic disfigurement along with fear of being malignant that compelled him to seek medical advice. The swelling was subjected to histopathological analysis following excision by the surgeon. Grossly we received a nodular mass measuring 3cm in diameter and cut section was solid, homogenous grayish white.

Two sections were given from two different areas. Microscopy showed a thinly encapsulated tumour composed predominantly of large rosettes arranged radially around central collagenous cores surrounded by small cells that were monomorphic ovoid to elongated spindle cells with rounded hyperchromatic nuclei, and scant cytoplasm resembling lymphocytes. Cells in both components showed strong nuclear and cytoplasmic positivity for S100 protein whereas negative for neuron-specific enolase and synaptophysin.

Corroborating the histomorphological findings and IHC final diagnosis of schwannoma like neuroblastoma was given.

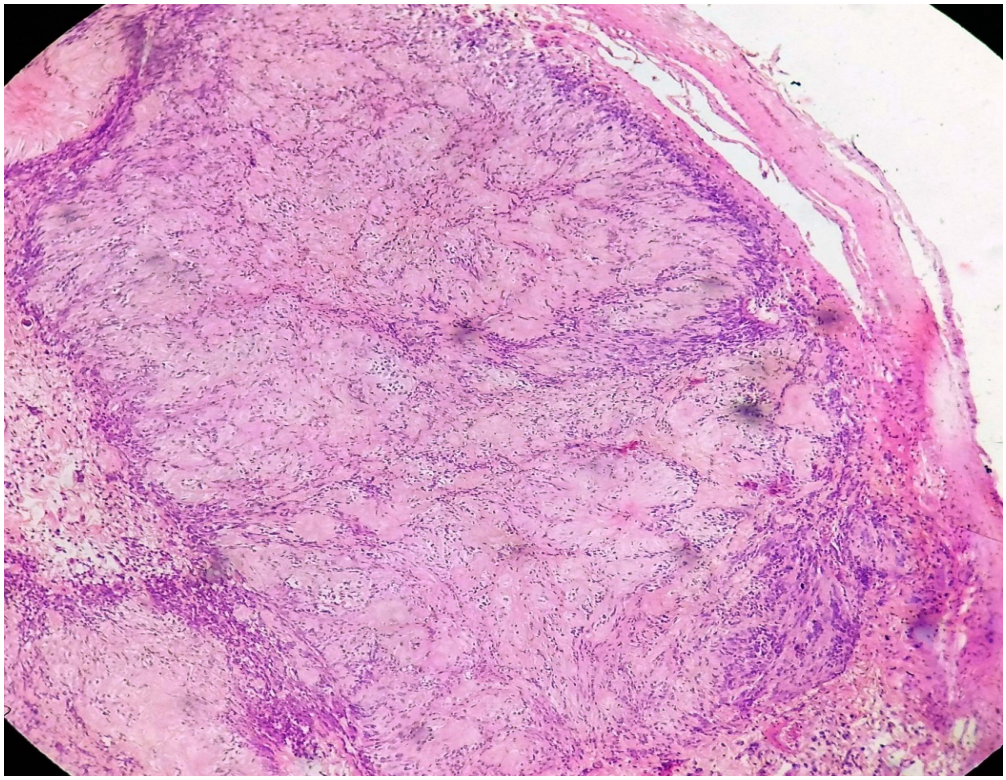

Figure 1: Showing thinly encapsulated tumour (Scanner view 4x)

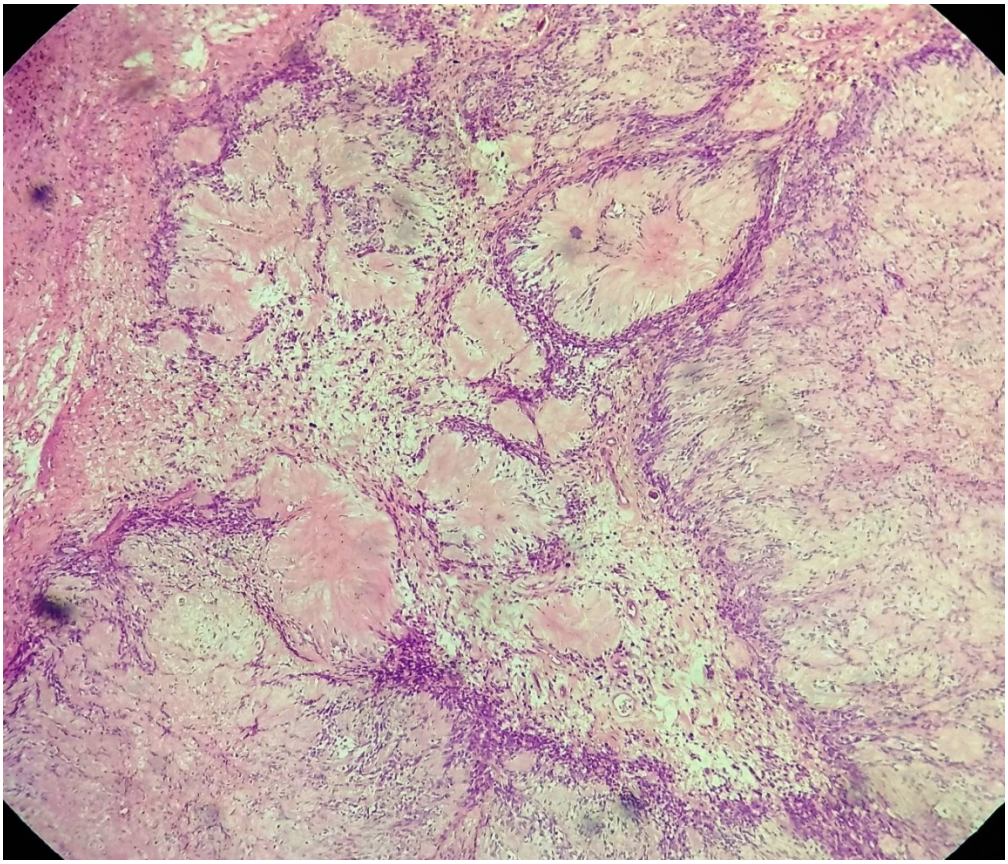

Figure 2: Photograph showing tumour cells composed of large rosettes radially arranged around central collagenous cores surrounded by small to ovoid to spindle with hyperchromatic nuclei and scant pale cytoplasm. [100x]

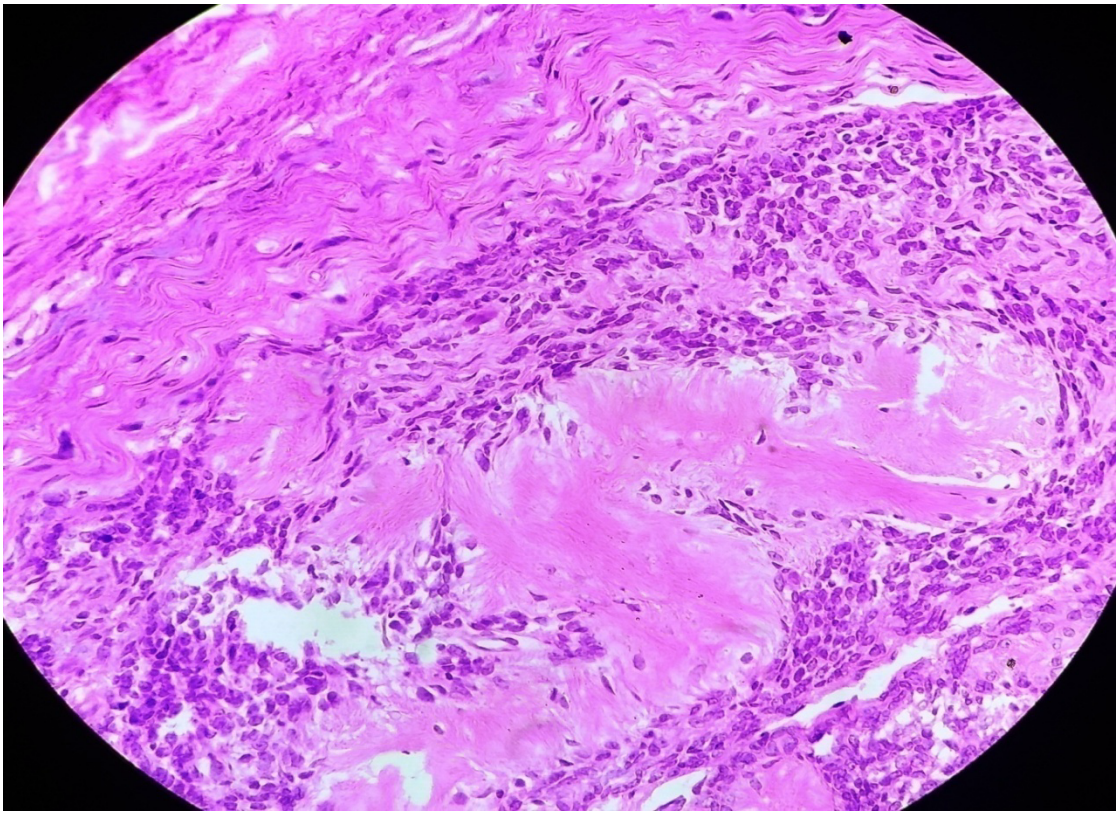

Figure 3: 100x-- Showing small round cells seems to be radially arranged around hyaline central fibrocollagenous cores

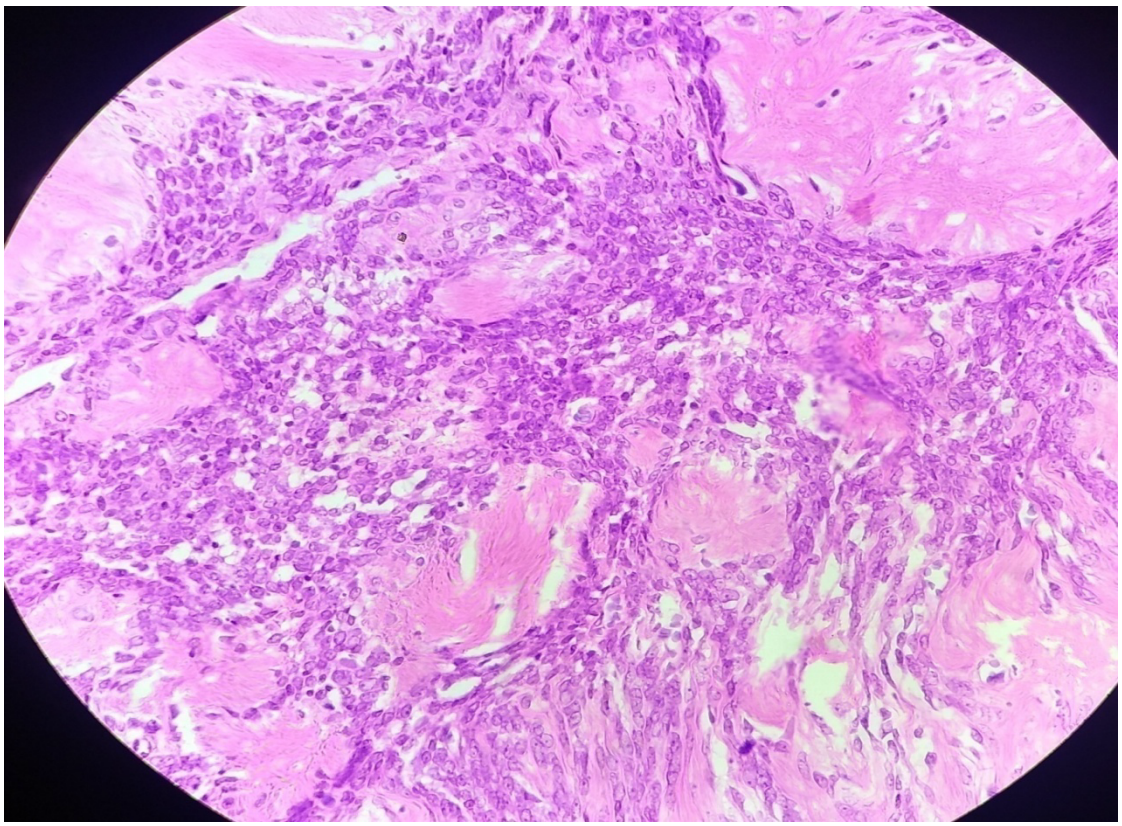

Figure 4: Showing monomorphic looking round to slightly elongated, hyperchrmatic nuclei scant cytoplasm

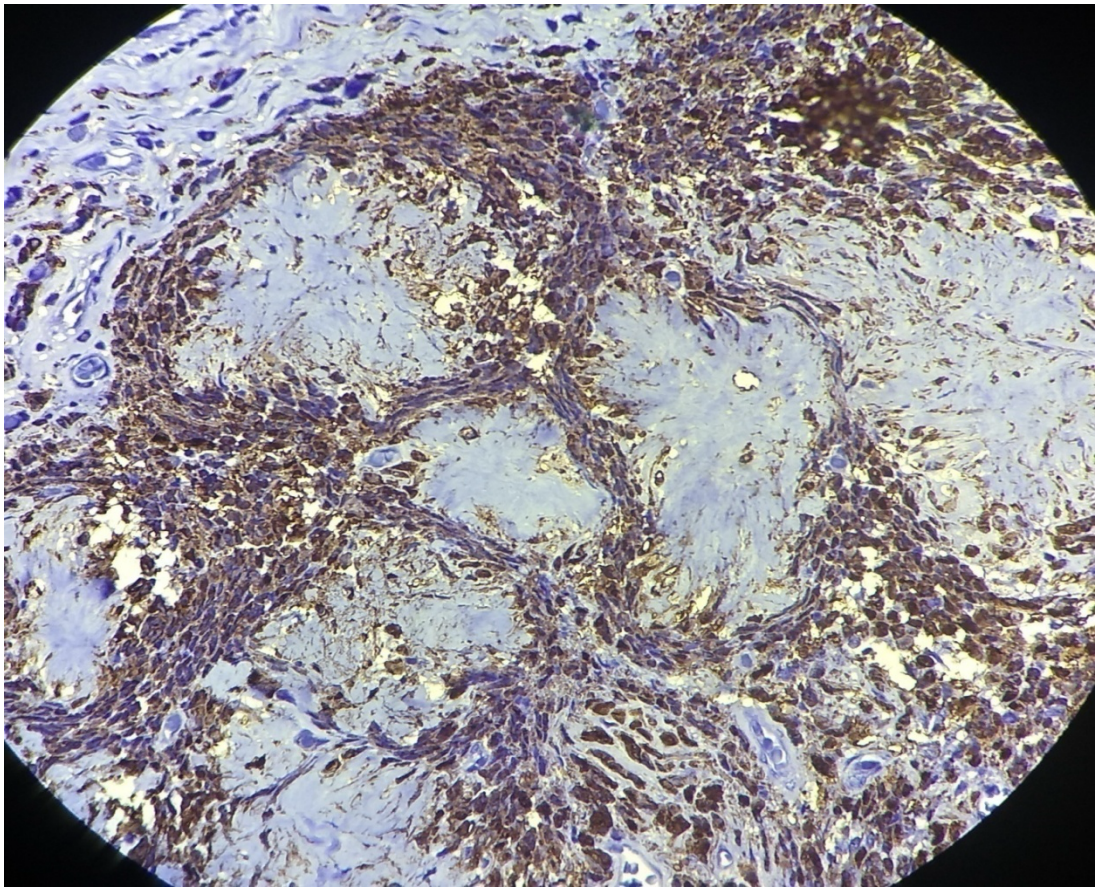

Figure 5: (a)

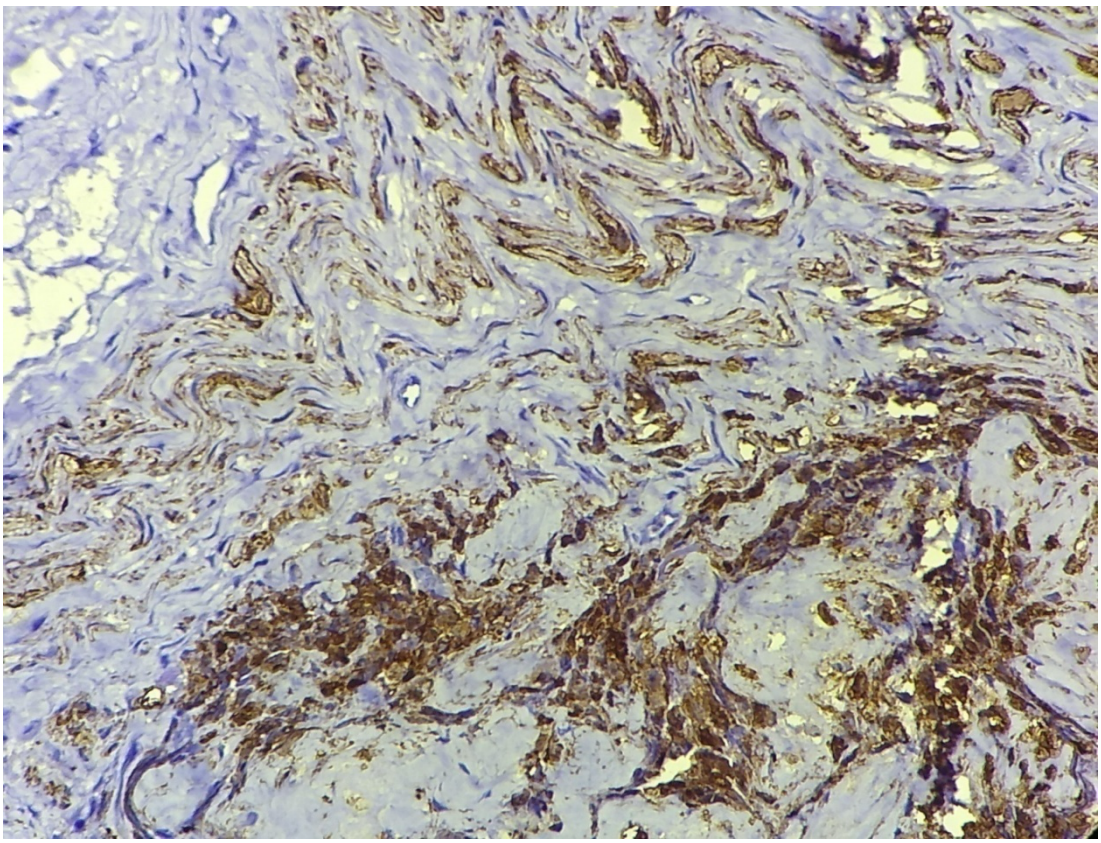

Figure 5: (b)

Figure 5 (a)(b) - Showing IHC nuclear positivity for S100.

Discussion

Cutaneous Schwannomas are benign neoplasms of peripheral nerve sheath composed of varying proportion of Antoni A and Antoni B areas. They have predilection for head, neck and flexural aspects of extremities. They occur as sporadic lesions or can be multiple when syndromic association with neurfibromatosis or schwannomatosis. They are composed of spindle cells with elongated wavy nuclei.

Classical schwannoma seldom pose a diagnostic difficulty for the pathologists, but it is the unusual variants of schwannoma that poses a diagnostic challenge like epithelioid schwannmas, schwannoma with neuroblastma like rosettes and malignant change in schwannoma. Such variants therefore require an extensive clinicoradiolgical corelation and IHC work up for their confirmation.

Review of literatures have shown that Neuroblastoma like schwannoma was originally described by Goldblum et al in the year 1994, is a rare variant of schwannoma histologically mimicking round cell tumours like neuroblastoma or ependymoma. Only two of the three cases reported by him described the morphology to be composed of small round cells that resembled neuroblastoma with Homer-wright rosette like structures whereas the third case showed perivascular ependymomatous rosettes [2]. So far 24 cases have been reported from various sites [3]. Nine reports described subcutaneous nodules at various locations (such as the knee, vulva, thigh, neck, palm, and flank) [2,4,5,6,7] while Kukreja et al [5] described the first case of neuroblastoma like schwannoma of the orbit.

It is the small hyperchromatic Schwann cell component and the rosettes that create a diagnostic overlap with primitive neuroectodermal tumours or neuroblastoma. However in neuroblastoma Homer wright rosette are considerably smaller than neuroblastoma like schwannoma and their centre contains a fine fibrillary substance (neuropil) that is stained by NSE. Primitive neuroectodermal tumour (PNET) on the other hand can be distuinguished from neuroblastoma like schwannoma by the presence of nuclear atypia, mitosis and necrosis and also showing strong diffuse membranous positivity for CD99 [4]. In the literature, only two convincing cases of transformed schwannoma have been described that had histologic features of PNET.[8,9]

Skelton et al in 1994 reported a case of schwannoma with collagenous spherulosis that showed some features similar to neuroblastoma like schwannoma [10]. However bland looking spindle cells and rounded cells were not arranged in rosettes but were compressed by numerous masses composed of radiating collagen fibres. Later, de Saint Aubain Somerhausen et al. described two further cases of neuroblastoma-like schwannoma,[4] and in 1998, Bhatnagar et al. published two new cases of schwannoma with immunohistochemical characterization showing areas that mimicked neuroblastoma/ peripheral primitive neuroectodermal tumor [2].

The recently described dendritic cell neurofibroma with pseudorosettes is a multinodular dermal lesion composed of small dark cells typically arranged concentrically. However, pseudorosettes are smaller than in neuroblastoma like schwannoma, and their central parts are filled with larger cells with pale eosinophilic cytoplasm and vesicular nuclei. Both cell types are immunoreactive for S100 and CD57, a characteristic which highlights the dendritic cytoplasmic expansions of larger cells [11]. Giant collagen-containing rosettes lined by S100 positive cells are also seen in the rare hyalinizing spindle cell tumor with pseudorosettes, which is regarded as a variant of low-grade fibromyxoid sarcoma. Although these lesions are usually large and deep-seated, they may on occasion present in the subcutis [12].

The typical histomorphological clue to the diagnosis is the large rosette like structures, composed of small round cells radially arranged around a central fibrocollagenous core. These small cells were monomorphic, round to slightly elongated, with hyperchromatic nuclei and scant amount of cytoplasm, resembling lymphocytes. The central collagenous network can be highlighted with the use of Mason trichome stain. Immunohistochemiscal results often shows strong positivity for S-100 with both cytoplasmic and nuclear staining and negative for neuron-specific enolase, smooth muscle actin and CD99. In our case IHC S100 was positive and the overall features were consistent with schwannoma having neuroblastoma-like rosettes.

Conclusion

To, summarise we report an extremely uncommon variant of schwannma with distinct histology that cause a diagnostic overlap with PNET or neuroblastoma. Pathologists should be aware of this rare variant of schwannoma to avoid erroneus diagnosis that may lead to inappropiate treatment .Distinction between these entities is important as treatment modalities differ for each entity. Neuroblastomas like schwannoma is a benign entity and mere excision would suffice while their differential diagnosis discussed requires adjuvant chemotherapy and radiotherapy.

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