

Intraparotid Schwannoma in Childhood: A Case Report

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Abstract: Schwannoma is a benign, capsulated perineural tumour of neuroectodermal derivation that originates from the Schwann cells. The tumour is usually solitary, smooth surfaced, slow growing and generally asymptomatic. Intraparotid Schwannomas are uncommon. Preoperative diagnosis of this tumour is difficult due to varied clinical presentation and nerve site involvement. Here we report a case of a 10yrs old female child with complain of right infra-auricular swelling of seven months. Superficial parotidectomy was done and by histopathological and immunohistochemistry diagnosis was confirmed as Schwannoma.

Keywords: Intraparotid, schwannoma

I. Introduction

Intraparotid schwannoma is a rare ectodermal benign encapsulated tumour. Schwannoma is a truly encapsulated solitary neoplasm which contains well – differentiated Schwann cell. They can arise from any part of the nerve along the course. Schwannoma are most commonly seen at fourth to sixth decade. The most common location is the flexor surface of the extremities, neck, mediastinum, retroperitoneum, posterior spinal roots, and cerebellopontine angle. ^[1] The incidence of parotid ranges from 0.2 – 1.5%, ^[2] for which it is rarely diagnosed preoperatively. So our case is unique as its intraparotid schwannoma occurring in childhood.

II. Case Report

10yrs old female child came to surgical outpatient with complain of right infra-auricular swelling of seven months. The swelling was gradually increasing in size and was painless. Ultrasonography reports revealed heterogeneous lobulated space occupying lesion measuring 3 x 2 cm with slightly increased vascularity, suggestive of pleomorphic adenoma. Then superficial parotidectomy was done and the mass was sent for histopathological examination.

Gross examination revealed well encapsulated mass measuring 3 x 2 cm. Cut surface was greyish white. The tissue was processed and stained with haematoxylin and eosin.

Microsection showed a well encapsulated mass having biphasic pattern. Cellular areas having spindle shaped cells arranged in palisading pattern forming *verocay* bodies which were Antoni A areas and a relatively hypocellular area of Antoni B type was seen. Antoni B type areas show haemorrhage, hyalinisation and cavitations. Immunohistochemistry for confirmation was done with S-100 which showed membrane positivity. So it was confirmed as intraparotid Schwannoma.

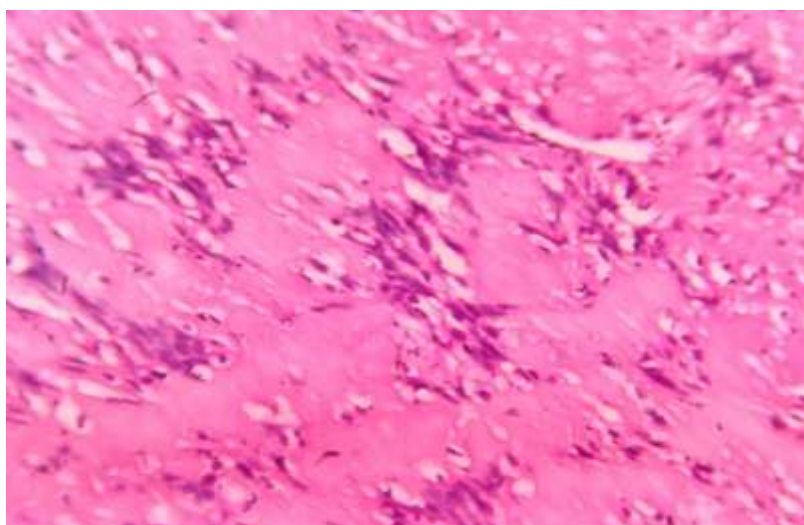


Figure 1: Antoni A and Antoni B areas.

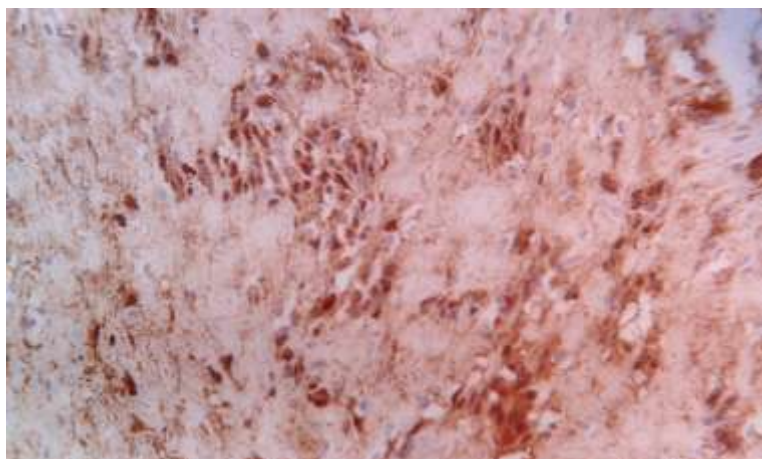


Figure 2: Immunoreactivity for S-100

III. Discussion

Schwannomas were first reported by Virchow in 1908. It's a benign nerve sheath tumour that is typically encapsulated and composed entirely of well-differentiated Schwann cells. Schwannomas represent 8% of intracranial tumours, 85% of cerebellopontine angle tumours and 29% of spinal nerve root tumours. [3] Approximately 90% of cases are solitary and sporadic, while 4% arise in the setting of neurofibromatosis type 2 (NF2), and 5% are multiple but unassociated with NF2. [4] All age group are affected, but most commonly between fourth to sixth decade of life. There is no gender predilection. Most often affected site is skin and subcutaneous tissue. [5] Intracranial schwannomas show a strong predilection for the eighth cranial nerve in the cerebellopontine angle seen in case of NF2. Schwannomas are mostly asymptomatic if located peripherally. Intracranial spinal nerve locations have radicular pain and signs of nerve root and spinal cord compression.

In FNA schwannoma can be diagnosed but sometimes can give misdiagnosis of a pleomorphic adenoma. Cytosmear shows an abundance of large, cohesive tissue fragments with elongated, spindled nuclei having blunted ends with ill-defined cytoplasm and nuclear palisading (*Verocay* body).

The majority of schwannomas are globoid encapsulated masses measuring from a few centimetres to 10 cm in size with pushing border without involvement of surrounding tissue. Outer surface is usually smooth. . The cut surface of the tumour typically reveals light tan glistening tissue interrupted by bright yellow patches with or without cysts and haemorrhage. Infarct-like necrosis related to degenerative vascular changes may be evident in sizable tumour. [6]

Microscopic appearance is distinctive with two patterns in varying proportion. Antoni A pattern where areas of compact, elongated or spindle cells with occasional nuclear palisading or in organoid arrangement (*Verocay* bodies), and less cellular, loosely textured cells with indistinct processes and variable lipidization which is Antoni B. The Schwann cells comprising the tumour have moderate quantities of eosinophilic cytoplasm without definite cell borders. Antoni A tissue features normochromic spindle shaped or round nuclei approximately the size of those of smooth muscle cells, but tapered instead of blunt-ended. In Antoni B tissue, tumour cells have smaller, often round to ovoid nuclei. Nuclear pleomorphism, including bizarre forms with cytoplasmic-nuclear inclusions ("ancient schwannoma") and the occasional mitotic figure may be seen. [6] Blood vessels can be prominent with thrombosis and hyaline thickening. Malignant transformation is rare, if so they have epithelioid morphology.

Palisading of nuclei can also occur in leiomyoma, leiomyosarcoma, GIST, calcifying aponeurotic fibroma, and even in non-neoplastic smooth muscle (most commonly in the appendiceal wall). [7] Different variant of schwannoma are Cellular schwannoma, Plexiform schwannoma, Ancient schwannoma, Melanotic schwannoma, Psammomatous melanotic schwannoma and benign epithelioid schwannoma. Differential diagnosis of intraparotid schwannoma can be malignant peripheral nerve sheath tumor (MPNST) and neurofibroma.

Immunohistochemically, the tumour cells show immunoreactivity for S-100 protein, calretinin, calcineurin, basal lamina components (such as laminin, type IV collagen, and merosin), vimentin and nerve growth factor receptor.

IV. Conclusion

Intraparotid schwannoma is exceptionally rare and it mimics pleomorphic adenoma on FNA and other imaging modalities, but it should be kept in the differential diagnosis of parotid tumour of long duration. Our case is unique due to its location in parotid and in childhood presentation. The diagnosis of schwannoma can be

made by histological features and immunohistochemical profile. Conservative surgery should be planned and regular follow up should be done.

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