Scholars Journal of Medical Case Reports

Sch J Med Case Rep 2015; 3(5)426-429 ©Scholars Academic and Scientific Publishers (SAS Publishers) (An International Publisher for Academic and Scientific Resources) ISSN 2347-6559 (Online) ISSN 2347-9507 (Print)

DOI: 10.36347/sjmcr.2015.v03i05.023

Intraparotid Schwannoma in a Young Adult: A Rare Entity

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Abstract: Intraparotid schwannoma arising from facial nerve are uncommon. These are slow growing, well encapsulated tumors that may arise from the nerve sheath. Preoperative diagnosis of schwannoma is difficult due to varied clinical presentation and usually misdiagnosed as pleomorphic adenoma. Diagnosis is usually confirmed after surgery by histopathology and immunohistochemistry. Till date, few cases of intraparotid schwannoma have been reported in the literature. Here we present a rare case of intraparotid schwannoma in a young adult (18 years) presenting as an asymptomatic parotid tumor with preserved seventh nerve function. Keywords: Parotid, Schwannoma, Facial nerve, Adenoma.

INTRODUCTION

Schwannoma is an unusual encapsulated benign tumor with ectodermal origin. Schwannomas of the facial nerve arise from any part of the nerve along the course, either the extratemporal or intratemporal region [1]. Majority of these tumors are intratemporal. whereas only 9% of schwannoma are located extracranially. They arise from glial Schwann cell transition site at the cerebellopontine angle to the peripheral branches of facial nerve in the parotid gland. They are usually asymptomatic and appear as painless parotid swelling [2]. Intraparotid schwannoma is difficult to diagnose based on clnical and imaging. To the best of our knowledge, few cases of intraparotid schwannoma have been reported in the literature. Incidence of parotid tumor of facial nerve origin ranges from 0.2-1.5% of which majority is schwannoma [1].

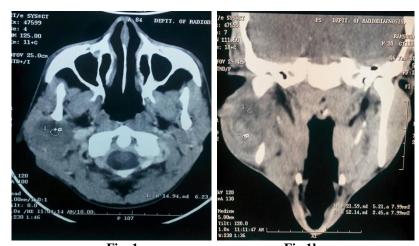
CASE REPORT

A 18 years old male patient presented with a history of a right infra auricular swelling of 10 months duration. It was progressively increasing in size and was not associated with pain. On examination, there was an enlarged right parotid swelling, measuring about 3x3 cm. It was mobile, non tender and firm in consistency. No signs of facial nerve palsy were noted. No evidence of any intraoral swelling or cervical lymphadenopathy was noted.

Computed tomography (CECT scan) revealed heterogenous large lesion measuring 3x 4 cm with slightly increased vascularity suggestive of pleomorphic adenoma (Fig. 1A & 1B). Fine needle aspiration cytology (FNAC) of the swelling showed features suggestive of adenoma. Based on the preoperative investigation, diagnosis of pleomorphic adenoma was made.

Patient planned for superficial was paroitectomy. Intraoperatively, the tumor was found to be superficial, firm, well encapsulated, adhered to lower trunk and marginal branch of facial nerve. Patient underwent superficial parotidectomy with preservation of facial nerve. Postoperative period was uneventful.

Gross examination revealed well encapsulated solid tumour with greyish-white cut surface measuring 4 x 3.7 x 2 cm with thin rim of salivary gland at the periphery (Fig. 2). Histopathological examination of the specimen revealed a benign tumor with biphasic pattern showing Antoni A and Antoni B areas. Antoni A areas were composed of elongated bipolar spindle cells and intercellular fibres and compact whorls forming Verocay bodies were seen. Type B area showed vacuolated spindle cells in loose myxomatous background (Fig. 3A & 3B). Immunohistochemistry showed strong positivity for S-100 protein (Fig. 4) and negativity for p63, vimentin and cytokeratin. Final diagnosis of intraparotid schwannoma was made. The patient was followed up for 8 months and was asymptomatic.



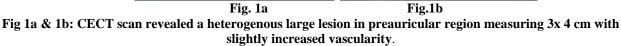




Fig. 2: specimen examination revealed well encapsulated solid tumour with greyish-white cut surface with a thin rim of salivary gland at the periphery

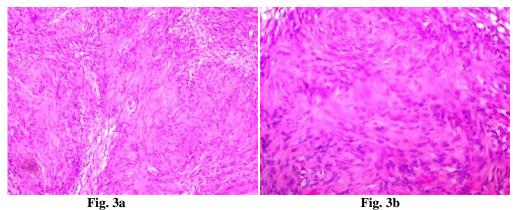


Fig. 3a & 3b: Photomicrograph revealed tumor cells arranged in biphasic pattern showing Antoni A (hypercellular) and Antoni B areas (hypocellular)

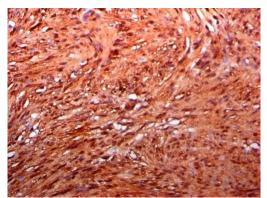


Fig. 4: Tumor cells stained positive for S-100

DISCUSSION

Intraparotid schwannomas are slow growing tumors that arise from nerve sheath of branches of facial nerve. Exact aetipathogenesis is not known. These can occur sporadically or associated with genetic conditions such as neurofibromatosis type 1(NF 1) and type 2(NF 2). [3] Schwannomas usually occur in fifth decade and very rare below 30 years. Liu and Caughey had described mean ages of 42 and 44 years respectively [4, 5]. Purva et al. has reported a case of facial nerve schwannoma in a 12 year old female [6]. To the best of our knowledge, this is one of the rare case of intraparotid schwannoma to occur at younger age. Most common presenting symptom is painless solitary swelling. Features suggestive of facial nerve involvement are usually not seen. Incidence of facial nerve paresis or palsy is around 20% in facial nerve schwannomas [7].

Ultrasonography and CECT Scan may fail to differentiate from pleomorphic adenoma [8, 9]. Preoperative diagnosis of intraparotid schwannoma is difficult and usually misdiagnosed as pleomorphic adenoma based on clinical and radiological presentation. Role of FNAC in diagnosis is not well established with accuracy of 80 % in diagnosis of benign and 62 % for malignant lesion [10]. Definitive diagnosis is based on histopathological examination of the excised swelling. On gross examination, the tumor is usually well encapsulated and in most instances tumor remains adherent to the facial nerve or its branches. Histology reveals tumor consisting of biphasic pattern, Antoni type A (hyper cellular areas with compactly arranged spindle cells) and Antoni type B (hypo cellular areas, sparse spindle cells showing vacoulation and xanthomatous change) [11]. Mitotic figures were infrequent. Schwannomas rarely undergoes malignant change [12]. It is usually confused with spindle cell myoepithelioma. Immunohistochemistry (IHC) study reveals tumor cells positive for S-100 which is a marker of schwannoma, whereas negative for p63 and cytokeratin (CK), a marker of myoepithelioma [13].

CONCLUSION

Intraparotid facial nerve schwannoma is an unusual tumor, often resemble pleomorphic adenoma based on clinical and radiological presentation. Facial nerve paresis or palsy is rarely seen. Diagnosis of parotid schwannomas should be kept in differential diagnosis of parotid tumor of long duration. Final diagnosis of parotid schwannoma was made postoperatively based on histopathological examination of biopsy tissue and immunohistochemical analysis.

ACKNOWLEDGEMENT

Our sincere thanks to Prof Dr. Ravikant, Vice chancellor, King George's Medical College, for guiding and permitting us to publish this article.

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