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Ancient Schwannoma of Paraphryngeal Space

Dr. G.Subba Rao1 M.S, Dr.Sk.Sufiya2 M.S, Dr.B.V.R.S.Sai Virinchi Yadav3

1. *M.S.(General Surgery) , Professor, Guntur Medical college,Guntur, A.P, India.*
2. *M.S(General Surgery),Assistant Professor,Guntur Medical College,Guntur, A.P, India.*
3. *Post graduate, Department of General Surgery, Guntur Medical College, A.P, India.*

Corresponding author: Dr. G.Subba Rao

Abstract
Schwannoma is a benign encapsulated tumor that arises from Schwann cells that form the myelin sheath around myelinated peripheral axons. Tumors are usually solitary and grow to a large size before undergoing notable degenerative changes. The term "ancient schwannoma" is used to describe a schwannoma that has undergone changes such as relative loss of Antoni Type A tissue, perivascular hyalinization, calcification, cystic necrosis, hemorrhage and the presence of degenerative nuclear changes. A case of Ancient Schwannoma of parapharyngeal space in a 28 year- old female is presented here.

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I. Introduction

Ancient schwannoma, a degenerative neurilemmoma or schwannoma are characterized by, distinct degenerative morphological changes in the tumor that includes cystic necrosis, stromal edema, xanthomatous change, fibrosis, perivascular hyalinization, calcification, degenerative nuclei with pleomorphism, lobulation and hyperchromasia. These degenerative features are attributed to the growth and "aging" of the tumor, hence the term "Ancient schwannoma."[1]

Tumors of the PPS(parapharyngeal space) are uncommon, comprising less than 1% of all head and neck neoplasms. Both benign and malignant tumors may arise from any of the structures contained within the PPS. Of PPS tumors, 70–80% are benign, and 20–30% are malignant. Most PPS tumors are of salivary or neurogenic origin.[2] Schwannoma is the most common tumor of neurogenic origin. A case of ancient schwannoma of parapharyngeal space is presented here.

II. Case Report

A 28-year-old female presented with a swelling in the right side of neck since 6 months gradually increasing in size. No complaints of any pressure symptoms noted. Clinical examination revealed a well-defined solitary swelling of size 4 x 4 cm, freely mobile, non-tender and firm in consistency. Cranial nerve and ENT examination were found to be normal.

Fine needle aspiration cytology revealed a rich cell yield consisting of oval to spindle shaped cells with vesicular nuclei against an amorphous background, with possibilities of myoepithelioma and nerve sheath tumor.

Excision biopsy was planned under General Anesthesia. Intraoperatively a well encapsulated firm mass was found in the post styloid compartment of parapharyngeal space. The mass was excised in toto by blunt dissection. Patient was put on post-operative analgesics and antibiotics for five days. Post-operative recovery was uneventful.

On gross examination, single well circumscribed grey brown mass measuring 4 x 4 x 3 cm was received. On cut section it was capsulated, solid, firm gray white with focal dark brown areas. Microscopic examination revealed Antoni A and Antoni B areas. The Antoni A areas showed Verocay bodies. In Antoni B areascells were loosely arranged in a myxoid matrix with areas of microcysts formations, hemorrhage and necrosis. Schwann cell nuclei were large, hyperchromatic and multilobulated. Based on the above, a diagnosis of Ancient Schwannoma was made.

III. Discussion

The parapharyngeal space is often described to be a deep potential neck space shaped as an inverted pyramid. The base of the pyramid is at the skull base, and the apex is at the greater cornu of the hyoid bone. Clinically, the parapharyngeal space should be considered in two compartments: pre-styloid compartment and poststyloid compartment. Fascia from the styloid process to the tensor veli palatine muscle divides the parapharyngeal space into these two compartments. The pre-styloid compartment is anterolateral and contains

the retromandibular portion of the deep lobe of the parotid gland, minor or ectopic salivary gland, a small branch of CN V to the tensor veli palatine muscle, ascending pharyngeal artery, and pharyngeal venous plexus. The majority of the pre-styloid compartment is actually fat. The poststyloid compartment is posteromedial and contains the internal carotid artery, internal jugular vein, CN IX to XII, cervical sympathetic chain, lymph nodes, and glomus bodies

Of the masses found in parapharyngeal space, 40–50% of the tumors are of salivary gland origin. LN metastasis and primary lymphoma represents 15% of lesions. Neurogenic lesions are the most common tumors of the poststyloid PPS and account for 25–30% of PPS lesions.[3]

Schwannoma also termed as neurilemmoma or neurinoma is one of the most common soft-tissue tumors. Tumor consists of two components: Antoni-A areas are more organized and are hypercellular, composed of spindle cells arranged in short bundles or interlacing fascicles. Antoni-B regions are hypocellular, less organized and contain more myxoid, loosely arranged tissue, with high water content. These components are intermixed within schwannomas and occur in varying amounts.[4]

The term "ancient neurilemmoma" was first suggested by Ackerman and Taylor in a review of 48 neurogenic tumors of the thorax. The cited authors reported ten patients with tumors showing features similar to those of typical neurilemmomas, but differing in that significant tumor portions contained only a few cells within hyalinised matrices. They found that these features occurred in schwannomas of long duration, deeply situated, larger masses and hence coined the term "ancient schwannoma" representing 0.8% of all soft-tissue tumors.[5]

Ancient schwannoma of the neck region is a rare benign neoplasm derived from neural crest cells and is usually solitary.The treatment of choice for neck schwannoma is complete surgical resection of the tumor. Recurrence and malignant transformation of schwannomas are very rare.[6]

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