International Journal of Research in Medical Sciences
Rana D et al. Int J Res Med Sci. 2017 Nov;5(11):5066-5068
www.msjonline.org
pISSN 2320-6071 | eISSN 2320-6012

DOI: http://dx.doi.org/10.18203/2320-6012.ijrms20174972

Case Report

Schwannoma in oropharynx: a rare site posing diagnostic challenge

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Received: 09 August 2017
Accepted: 07 September 2017

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ABSTRACT

Schwannomas are benign nerve sheath tumors. These arise from Schwann cells of the neural sheath. Intra oral region is a relatively uncommon site of these tumors. They are solitary, slow growing, smooth surfaced, usually asymptomatic, and encapsulated tumors, about 25% of all schwannomas are located in the head and neck, but only 1% show intraoral origin. A 22-year-old female came with dysphagia since, 3 years. FNAC was not feasible and so excision biopsy of the lesion was performed. Histopathology revealed schwannoma like picture and it was confirmed with diffuse S-100 positivity on immunohistochemistry. Hence, finally confirming schwannoma of the oropharyngeal region. Schwannomas can be found anywhere in the body but a quarter of all occur in the head and neck region. Intraorally its percentage is only 1% with tongue being the commonest. Pharyngeal presentations of schwannoma are rare. Schwannomas are benign tumors having excellent prognosis. Basically, this case report is important as these very rarely occur in the oropharyngeal region and it's a must to consider them in the differential diagnosis of lesions at this site.

Keywords: Benign, Neurogenic, Oropharynx

INTRODUCTION

Neurilemomas or schwannomas, are usually solitary, slow growing benign neoplasm's that are well encapsulated soft tissue lesions deriving from Schwann cells. Schwannomas can arise from any cranial, peripheral, or autonomic nerves that contain Schwann cells, the cells that form the myelin sheath over nerve fibres.[1] Schwannoma was first reported by Verocay in 1910, who called this benign neurogenic tumour as neurinoma.[2] About a quarter of all schwannomas occur in the head and neck region. Only 1% of these occur intraorally, generally in the tongue. Pharyngeal presentations of schwannoma are rare.[3] To the best of our knowledge only 13 cases of pharyngeal schwannomas have been reported and the tumour sites include posterior and lateral pharyngeal wall, pyriform sinus, and lateral glossoepiglottic fold.[4] Although they may arise at any age, the peak incidence of schwannoma is between the third and sixth decade of life, with no gender predilection.

CASE REPORT

This case report is of a 22 years old female who came to the pathology department of our tertiary hospital with an oropharyngeal mass present since, 3 years. She complained of dysphagia and difficulty in swallowing which was progressive in nature. There was significant weight loss. However, there was no history of change in voice or breathlessness. The patient was a non-smoker and non-alcoholic. Her vitals were within normal limits. Her hemogram and other lab investigations were within normal limits. On examination, a huge smooth lobular encapsulated mass arising from posterior and lateral wall of oropharynx extending into nasopharynx was seen. The

mass is free from vallecula and base of tongue. Overlying skin was unremarkable. The surrounding structures like vocal cords, larynx appears normal. There was no cervical lymphadenopathy. Ear and nose examination was unremarkable. Her skin was normal and there was no growth in any part of the body suggestive of neurofibromatosis. Fine Needle Aspiration Cytology could not be performed as the site was not approachable by needle. Excision biopsy was performed and the mass was sent to us in histopathology. We received a well encapsulated globular soft tissue mass measuring 2.5x 2.5x 1 cms. Outer surface is smooth greyish white. On cut, it is greyish yellow and gelatinous. Hematoxylin and eosin stained sections showed a tumour comprising of spindle cells arranged in short fascicles at places with hypercellular and hypocellular areas. Vessels of venous caliber noted with few showing wall thickenings. Periphery of the lesion shows stratified squamous epithelium with subepithelium showing congestion, edema and skeletal muscle bundles (Figure 1).

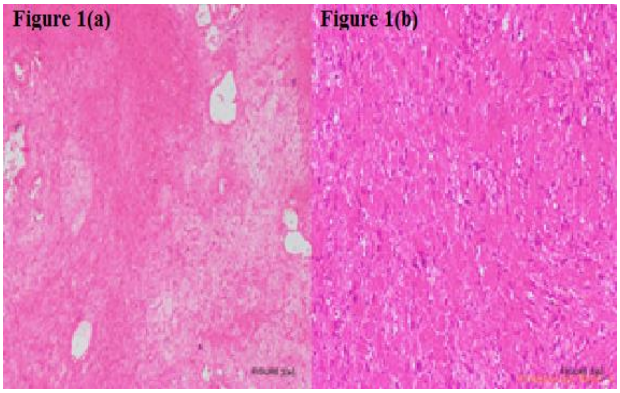

Figure 1: (a) Geimsa stained smears showing spindle tumor cells showing hypercellular and hypocellular areas. Thick walled vessels can also be seen in the section (x100). (b) Giemsa stained smears show verocay bodies seen in the hypercellular area (x400).

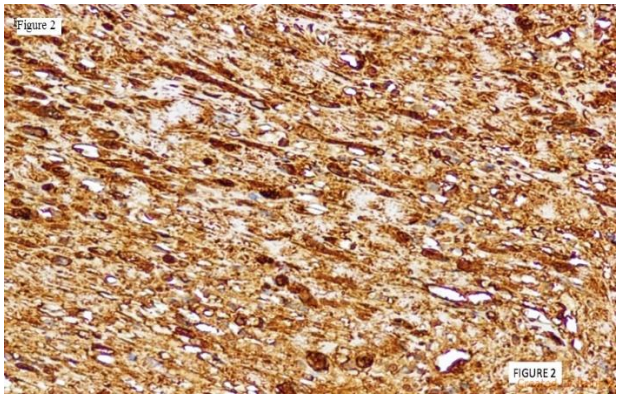

Figure 2: Photomicrograph showing strong diffuse nuclear S-100 positivity. (X100).

Immunohistochemistry showed strong nuclear S100 positivity in the spindle tumour cells along with focal positivity of CD-99 (Figure 2).

DISCUSSION

The schwannoma is also called as neurilemmoma, neurinoma, perineural fibroblastoma and is a solitary, slow growing, usually encapsulated, asymptomatic tumour. They rarely undergo malignant transformation. Although a quarter of all extracranial schwannomas occur in head and neck, pharyngeal presentations are rare. So far, only about 13 cases have been reported in the base of tongue to the best of our knowledge.[4] However, it is more common between the second and third decades of life.[1]

William et al. findings showed that in 83% of the cases studied by them the schwannomas presented in males, while for Lucas there was a greater predilection for females, and for Hatziotis and Asprides; Enzinger and Weiss there was an equal distribution between both sexes.[5-7]

We report a case of a 22 years old female in the oropharynx making it a rare entity. Oropharyngeal schwannomas usually are asymptomatic painless swelling gradually increasing in size. These tumors can also occasionally cause dysphagia, odynophagia and radiating pain. Since, the tumor is well encapsulated, the spread to the surrounding structures does not occur though it can cause compression of surrounding vascular structures. Our case also presents as a slow growing mass initially asymptomatic gradually causing dysphagia with no encroachment to surrounding structures. Clinically, no diagnosis could be made due to the vague history and poor background of the patient. CT could not be advised. The patient underwent surgery and the mass was sent for histopathological evaluation.

Macroscopically, schwannomas are typically circumscribed and encapsulated. Histopathologically, five schwannoma variants have been described: common, plexiform, cellular, epithelioid, and ancient schwannomas.[8] In common schwannoma two distinct histological patterns can be seen: known as Antoni type A and type B. Antoni type A tissues are characterized by compact Schwann cells with nuclear palisading, whereas Antoni type B tissues exhibit a considerable degree of cell pleomorphism in a loosely arranged reticulum network. Vascularity is not a prominent feature, and necrosis and mitotic activity are seldom encountered.[9] Sometimes in type A, the cells are aligned in palisade manner with the nuclei lying side by side in one strip and cytoplasm of cells in the adjacent strip, this pattern known as “Verocay body”.[10] This pattern was also appreciated in our case. As the schwannoma ages, degenerative changes take place, characterized by hyalinized tissue, myxoid areas, and large cystic spaces. The resulting entity is known as ancient schwannoma. Malignant transformation is rare.[11]

In evaluating a patient with a slow growing tongue mass that has been present for a long period of time, benign soft tissue neoplasms and reactive lesions need to be

considered. In addition to schwannomas, the differential diagnosis should include neurofibromas, granular cell tumors, irritation fibromas, leiomyomas, rhabdomyomas, hemangiomas, lymphangiomas, lipomas, pyogenic granulomas, and benign salivary gland tumors, nerve sheath myxoma, palisaded encapsulated neurinoma, mucosal neurinoma associated with multiple endocrine neoplasia III, traumatic neuroma and granular cell tumour.[11] The presence of schwannoma calls for the careful search for nerve tumors in other parts of the body, although in most cases none may be found. The differentiation of schwannoma from neurofibroma is essential, because an apparently solitary neurofibroma may be a manifestation of neurofibromatosis. They are distributed in a centripetal fashion on the body.[12]

Microscopically, both schwannoma and the neurofibroma contain elongated cells with irregular nuclei lying between bundles of collagen fibers. They differ histologically and histogenetically, the schwannoma is derived from the Schwann cells and the neurofibroma from mixture of Schwann cells, perineurial cells, and endoneurial fibroblasts. Neurofibroma is not encapsulated.[13]

The diagnosis of schwannoma presenting in this unusual location in our patient would have been difficult to make based upon routine haematoxylin and eosin stains alone. Immunohistochemistry is a simple, accurate, and cost-effective technique used in the diagnosis of schwannoma. S-100 protein is a diagnostic immunohistochemically marker for schwannoma having 95% sensitivity, in contrast to the cells of neurofibroma, which variably express the antigen.[14]

In our case S-100 showed strong nuclear positivity. Malignant transformation of head and neck schwannoma occurs only in 8-10% of cases.[15] Prognosis is excellent as the tumour is benign, and recurrence is rare unless the resection of the tumour is incomplete. Therefore, it is important to diagnose these tumors as a separate entity as they have different clinical behavior and management protocol.

CONCLUSION

The schwannoma represents a lesion not often encountered in clinical practice. The submucosal forms of this lesion are usually indistinguishable from other benign neoplasms that are also usually seen in the same region. The final diagnosis should be done after histopathological exam and in some cases after immunohistochemically analysis. The therapeutically conduct is the total removal of the lesion. Moreover, histopathology with immunohistochemistry helps in confirmation and further management of the patient. This case highlights the need for studies to elucidate better treatments strategies for these patients.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Rana D, Raychaudhuri S, Sharma N. Schwannoma in oropharynx: a rare site posing diagnostic challenge. Int J Res Med Sci 2017;5:5066-8.