

Case Report

Solitary Neurofibroma of Intramastoid Region in Middle Ear Cavity

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ABSTRACT

Neurofibromas are benign lesions of peripheral nerve sheath tumours comprising of Schwann cell, axons, fibroblast and perineural cells. Auricular nerve involvement is rarely seen in neurofibroma. We present a 34 year female came with complains of right ear pain whose clinical diagnosis of cholesteatoma was given, but on histopathological examination features of neurofibroma was seen. For confirmation immunohistochemistry was done and it showed S-100 positivity.

Key words: Ear, auricular nerve, neurofibroma.

INTRODUCTION

Neurofibroma is one of the benign lesion of a nerve sheath tumour. They are mainly formed by the Schwann cells along with axons, fibroblast, perineurial cells. ⁽¹⁾ Neurofibromas are classified into three subtypes for clinicopathological perspective: Localized cutaneous neurofibroma, diffuse neurofibroma, Plexiform neurofibroma. ⁽²⁾ Neurofibroma stains positively for S-100 protein. ⁽³⁾ Rarely auricular nerve involvement is seen in neurofibroma. Though head neck face is common regions for benign nerve sheath tumours, the middle ear is an uncommon location for neurofibromas. Facial nerve and glossopharyngeal nerve involvement are very rarely seen.

CASE REPORT

A 34-year female came to ENT OPD with complaints of right sided ear pain since 4 months. There was no history of ear discharge, tinnitus or vertigo. No café-au-lait spots were seen anywhere on the body. Audiogram results showed moderate

conductive deafness in the right ear and clinical diagnosis of Cholesteatoma was given. The patient underwent polypectomy and the sample was sent to histopathology for confirmation.

The gross finding was multiple, grey-white to grey-brown, friable, soft tissue all aggregating measuring 0.4cm was received. Microscopic examination of the tissue revealed interlacing bundles of spindle cells with darkly stained elongated wavy nuclei having serpentine configuration and pointed ends. Also, inflammatory cells comprising of mast cells, lymphocytes and few neutrophils dispersed throughout the stroma was noted. Antoni A or Antoni B were not seen in the sections submitted. S-100 was positive for the spindle cells on immunohistochemistry. Based on histopathological examination and immunohistochemistry findings the diagnosis of benign solitary neurofibroma was given.

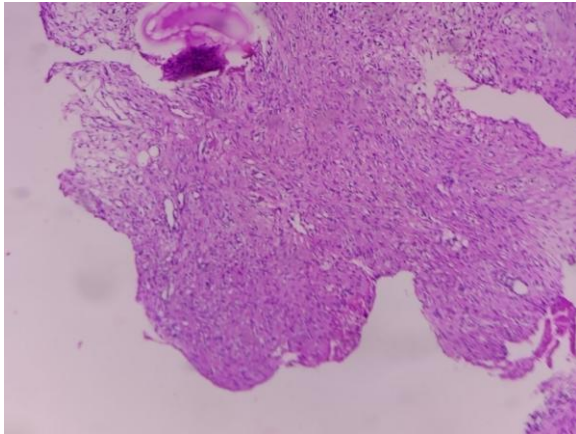


Fig 1. Hematoxylin-eosin x10 : section show proliferation of spindle cells with shredded carrot collagen.

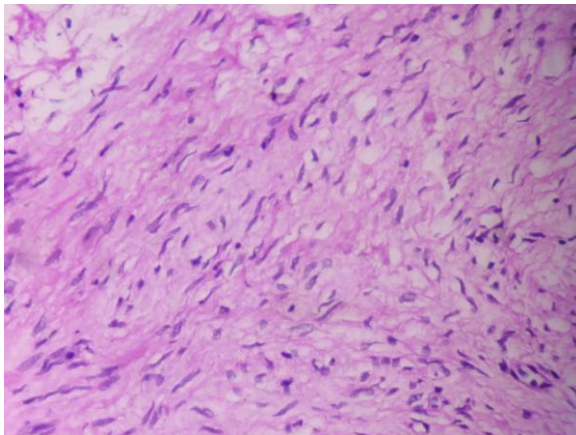


Fig 2. Hematoxylin-eosin x10: Section show interlacing bundles of spindle cells with darkly stained wavy nuclei along with mast cells and lymphocytes.

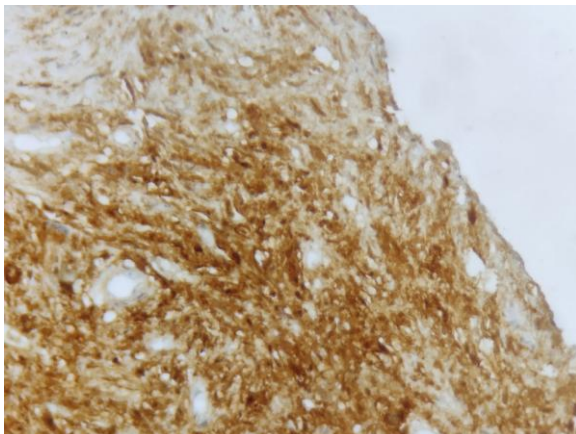


Fig 3. Immunohistochemistry showing S-100 positivity.

DISCUSSION

Solitary tumours are rare along the nerve. [4] The occurrence of Intramastoid location is very uncommon in the Head and Neck region. [5] Glossopharyngeal nerve, Facial nerve and its branches are involved in the middle ear Schwannoma. The cases which are strongly associated

with neurofibroma syndrome. [6] These tumours are rarely diagnosed in the intratemporal region. The MRI and CT scan done preoperatively can aid in the diagnosis of this tumor. [5] But the histopathology with immunochemistry is a must for the confirmation.

Neurofibroma subtypes are multiple cutaneous subtypes, massive subtypes and plexiform subtypes. [2]

The differential diagnosis can be acute otomastoiditis, facial nerve schwannoma, squamous cell carcinoma and adenoid cystic carcinoma of the head and neck region. The rate of malignant transformation of solitary neurofibroma is believed to be low. [5] The plexiform subtype associated with NF -1 has a higher rate of malignant transformation as compared to the localised cutaneous subtype which does not undergo such transformation. [2]

Subtotal resection in the head, neck, face regions are multinodular and multifocal which are factors for recurrence. [2,7] Wood et al in their study proposed multifocality and multinodular patterns as a factor for recurrence. [2] Complete excision of these tumours can be challenging as the morbidity associated resecting of important structure like facial nerve is another complicated part of the surgical resection.

CONCLUSION

Histopathology with immunohistochemistry remains the mainstay diagnostic tool for such solitary neurofibroma As the recurrence rate is more for solitary tumours the confirmatory diagnosis plays a vital role in the follow-up of the patient.

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