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Case Report

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Facial nerve schwannoma: a rare case report

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ABSTRACT

Schwannoma is a benign tumor arising from Schwann cells which is protective covering of nerves, called myelin sheath and can develop anywhere, where Schwann cells are present. Most common schwannomas are found with vestibulocochlear nerve. Facial nerve schwannoma are uncommon tumour involving 7th nerve out of which also most common site of involvement is geniculate ganglion. Facial nerve schwannoma is uncommon benign tumor. There are no typical patterns of presentation and can easily go untreated or misdiagnosed. Facial nerve palsy is most common mode of presentation. Here we present a case of 35 years male who presented with complaint of facial nerve paralysis. High degree of clinical suspicion and early imaging can lead to diagnosis. An early diagnosis is important as morbidity associated with this disease and as well as surgery leads to delay in diagnosis.

Keywords: Facial nerve, Schwannoma, Surgery

INTRODUCTION

Facial nerve schwannomas are the rare tumors of facial nerve which are easily to get misdiagnosed or mistreated. Facial nerve schwannomas (FNSs) are rare lesions that can arise anywhere along the course of the facial nerve, from its origin in the cerebellopontine angle to its extracranial ramifications in the parotid space of the extracranial head and neck.[1,2] They compose only 0.8% of all intrapetrous mass lesions.[3] They do not have any typical pattern of presentation. They arise from the nerve sheath which is made up of schwann cells. We here present a case of facial nerve schwannoma.

CASE REPORT

A 35 year old male presented with hearing loss left side facial palsy for 6 months, gradual in onset, progressive in nature, no history of ear discharge and history of treatment for same complaint for 3 months but with no improvement.

Examination

Left sided facial nerve paralysis with House Brackmann grade IV.

Otoscopic examination

Tympanic membrane found to be intact and retracted. Mass filling the middle ear was visualized.

Investigations

All the hematological and biochemical parameters were with in normal limits. Audiometery showed moderate conductive hearing loss (43 Db) was found in left ear, with normal hearing sensitivity in right ear. X-ray lateral view mastoid depicted partly sclerotic with small areas of lucency. HRCT temporal bone reported soft tissue attenuation in aditus, antrum with few left sided mastoid air cells.

Procedure

He was taken up for exploration of mastoid and middle ear under GA. Tympanomeatal flap was elevated. Reddish mass was seen arising from middle ear. Inside out mastoidectomy was done. Antrum was found to be normal. Facial recess approach was used. Facial canal was drilled out to expose it. Horizontal and vertical part of facial nerve was exposed. Mass arising from vertical part was seen. Facial nerve decompression was done proximal and distal from the diseased segment. Excised mass was sent for histopathology which was reported as benign peripheral nerve sheath tumour–schwannoma. Postoperatively patient was followed up. After 6 months patient has improvement with House Brackmann grade III facial paralysis. Features of neuroma with spindle shaped cells with wavy elongated nuclei with pointed ends Antoni A and Antoni B types of pattern are seen in schwannoma. Verocay bodies are seen in Antoni A pattern which is rows of palisading nuclei aligned in row.

DISCUSSION

Schwannoma is an ectodermal benign encapsulated tumor arising from Schwann cells. Schwannoma of the facial nerve are extremely rare which can arise anywhere along the course of facial nerve.[2] Peak incidence is between 3rd and 6th decade. Incidence is same in males as well as females. These tumors can arise from any segment of the nerve from the cerebellopontine angle to the extra-temporal peripheral portion.[4,5] They can be located intracranially, intratemporally or extratemporally. One study of 600 temporal bone study reveals reported facial nerve schwannoma incidence of 0.8%. The incidence of intratemporal facial neuromas was 0.8% in a cadaveric study, although this figure is higher than the rate of clinical presentation.[6] Most of these tumors are intra-temporal.

Common presenting features

Clinical presentation not only differs on the basis of site but also due to variability of structures getting involved. Facial nerve paralysis, hearing loss, facial nerve pain, hemifacial spasm, decreased lacrimation. Slowly progressive or sudden facial weakness, often preceded by facial twitching, is a common complaint.[7] In about 5% of patients with Bell's palsy, a facial nerve neuroma is found to be the cause.[8] But it has been found, normal facial nerve function has been reported in 27% of patients.[9] Schwannoma can arise anywhere along the course of facial nerve from the cerebellopontine angle to neuromuscular junction but there is predilection for the involvement of the geniculate ganglion. From here it can extend to involve tympanic segment or labriynthine portion of facial nerve. Uncommonly facial nerve can involve middle cranial fossa by direct upward spread through temporal bone. Most common presentation is occurrence of facial nerve paralysis which is progressive and is occurring over months. Hearing loss may be conductive, cochlear, retrocochlear. Origin from mastoid portion can present as ear polyp coming from posterior wall of external auditory canal. In this case ear discharge may be present.

Diagnostic work up include audiological test, auditory brain stem evoked response audiometery, CT, contrast enhanced MRI. CT and MRI are complimentary to each other. As for the diagnosis of facial nerve neuroma, the presence and extension of the tumor can be seen most accurately by radiologic examination. Bone targeted high-resolution CT of the temporal bone is believed to be superior to MRI. Enlargement of the facial nerve suggests involvement of a neoplastic process.[10] Electromyography is helpful for quantifying the nerve's residual motor function and predicting postoperative prognosis of facial recovery after nerve reconstruction.[11] Electro-neurinography can help predict the prognosis of postoperative facial palsy. It is believed that better postoperative results are achieved when the nerve is repaired if compound action potentials decrease no more than 50%.[12]

Treatment includes surgical removal. Approach depends on the site of tumor, size of tumor and hearing loss. Timing for surgery is controversial. Because facial nerve neuromas almost always grow slowly.[12] Waiting to pursue surgery until the facial nerve either shows progressive deterioration or becomes paralyzed in patients with normal facial nerve function has been advocated.[13,14] To delay the need for complete resection further, wide tumor decompression to allow the tumor to expand outside the natural bony confines has also been advocated.[12]

However, some authors believe that the surgical approach becomes more difficult, the likelihood of postoperative complications increases, and the recovery of facial function is poorer if the tumor continues to grow.[5,15] The risk of surrounding structures such as the inner ear being invaded increases and eventually the brainstem may become compressed.[12,13] Surgical resection of facial nerve neuromas is indicated without delay for patients with progressive facial palsy or paralysis, for large cerebellopontine angle tumors compressing the brainstem or producing hydrocephalus, and for tumors invading the inner ear.[12]

The surgical approach should be chosen based on the anatomic location and extension of the tumor.[12] A tumor proximal to the geniculate ganglion with serviceable hearing should be approached through the middle cranial fossa provided the tumor does not extend far into the cerebellopontine angle. If the tumor is proximal to the mid IAC with ≤1 cm of a CPA component, an extended middle cranial fossa approach is best. A retrosigmoid approach gives the best chance of hearing conservation in lesions with a CPA component >1 cm .With non-serviceable hearing, a translabyrinthine approach is the most direct route to the tumor and is the procedure of

choice. This approach also provides the best access for facial nerve grafting. Involvement of the tympanic segment can be reached by a transmastoid approach with facial recess opening. Mastoid segment tumor alone can be extirpated through the mastoid, whereas additional extratemporal involvement may require following the nerve into the parotid gland.[13,14,16] These exposures may be used in combination for tumors involving multiple nerve segments. The tumor has to be removed along the segment of facial nerve with cable grafting of nerve for reconstruction. A further controversy involves re-construction of the nerve.

Some authors have reported success with removal of the tumor and preservation of nerve integrity, but this does not gives good facial function.[17,18] Primary anastomosis without tension is rarely possible without mastoid segment rerouting, which if at all possible may compromise with blood supply of nerve itself. Cable grafting is usually performed with the greater auricular or sural nerves, but any rehabilitative technique that is to be used shall be used before severe atrophy of facial muscles has occurred. The grafting can be done with greater auricular nerve or sural nerve.

CONCLUSION

Facial nerve schwannoma are the rare tumors but can easily mimic other conditions. Delay in diagnosis can lead to increase in morbidity due to increase in area of involvement of the tumor along with increase in site and hence complicating the surgery. A high degree of clinical suspicion along with proper history and examination along with careful interpretation of imaging techniques can help to clinch to the diagnosis. It is possible to surgically remove these tumors while sparing the nerve. Primary facial nerve preservation is often associated with an origin of tumor from nerves other than motor facial nerves, but it may be possible if the tumor is located eccentrically on the facial nerve.

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