CASE REPORT

ISOLATED FACIAL NERVE PALSY DUE TO MASTOID CHONDROSARCOMA: A RARE PRESENTATION

G. Hari Krishna¹, P. Prahaladu², T. Vinay Bhushanam³, K. Siva Ramkrishna⁴, K. Satyavara Prasad⁵

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ABSTRACT: Chondrosarcomas of temporal bone is a rare malignant neoplasm constituting only 0.16% of all intracranial tumors. Cranial nerve involvement in chondrosarcoma of temporal bone is related to specific anatomic location of the tumor. Isolated facial palsy should be evaluated thoroughly and should be differentiated from Bell's palsy. We present a case of isolated facial palsy due to mastoid bone chondrosarcoma in a 25 year old man which was misdiagnosed as Bell's palsy and treated for the same previously. The clinical features, management and outcome are discussed with reviewing the related literature.

KEYWORDS: Bell's palsy, Chondrosarcoma, Facial palsy, Mastoid, Temporal bone tumors.

INTRODUCTION: Chondrosarcomas are rare primary bone tumors. They are composed of cartilage forming cells. Chondrosarcomas of temporal bone are rare neoplasms and account upto 0.16% of all the intracranial tumors.^{1,2,3} It is difficult to differentiate the tumor from other pathologies involving the temporal bone based on clinical signs. Misdiagnosis to have Bell's palsy may mask the primary pathology⁴. We report a case of chondrosarcoma of the mastoid bone presenting with facial palsy which was misdiagnosed and treated for Bell's palsy.

CASE REPORT: A 25 year old male presented 6 months back with sudden onset left sided facial palsy for which he was misdiagnosed to have Bell's palsy and was treated conservatively by a local doctor. He was referred to our centre for evaluation and on examination he had facial nerve palsy with involvement of facial muscles and taste in anterior 2/3rd. There is sparing of lacrimal function. There is conductive hearing loss in left ear. These findings are suggestive of facial nerve involvement in mastoid part. There were no other cranial nerve palsies.

Imaging of temporal bone revealed (Fig. 1: Preoperative images - CT temporal bone and Fig. 2: MRI brain and cervical region) a 4 x 6 cms irregular shaped and ill-defined bordered tumor present in left temporal bone (mastoid) which is extending into jugular foramen anteriorly, retromastoid suboccipital region posteriorly and extending down into the neck upto C2 transverse process. The tumor eroded the hypotympanum and tympanic membrane and extended into external ear. The tumor is osteolytic with spicules of destructed bone in the tumor with sclerosis. The soft part of the tumor is iso to hypodense on CT imaging and predominantly isointense in T1W and hyperintense in T2W images and very minimal patchy enhancement on contrast administration.

The patient subsequently underwent combined lateral skull base approach with presigmoid, retrosigmoid and neck extensions of the surgical corridor. The main bulk of the tumor present in the mastoid bone with extension into neck and retromastoid suboccipital region. Mastoidectomy and tympanotomy was done and the tumor mass present in the inferior half of the mastoid and whole of mastoid tip and extending through the antrum to the middle ear cavity.

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The tumor eroded the hypotympanum and tympanic cavity and extending into the external ear. Whole of the facial nerve in the fallopian canal was decompressed. The lower part of vertical segment of the facial nerve was thinned out by the tumor and corda tympani nerve was destroyed by the tumor. The facial nerve was eventually conserved. Retromastoid craniotomy done and tumor excised. Dura was intact. Subtotal resection of the tumor was done and the surgery was contemplated at the inferior most part due to diffuse oozing from the tumor at the region of C2 transverse process (Fig. 3: Intra-operative pictures). The mastoidectomy cavity filled with abdominal fat. Post-operative imaging showed subtotal resection of tumor with small residue at C2 transverse process (Fig. 4: Post-operative images).

The histopathology of the specimen showed cartilaginous metaplasia. The tumor cells were arranged in sheets and chords of uniform cells overlying a myxoid matrix and enlargement and hyperchromatosis of the nuclei suggesting Grade I chondrosarcoma.

Post-operative radiotherapy was given as there was tumor residue near C2 transverse process in the neck. One year follow up showed no recurrence of the tumor. Facial palsy did not improved.

DISCUSSION: Chondrosarcoma is a malignant neoplasm of cartilage forming cells and accounts for 0.16% of all intracranial tumors.^{1,2,3} Petrous part of the temporal bone is a common site for the development of the tumor.¹ Isolated mastoid bone chondrosarcoma with vertical part facial nerve involvement is very rare. Mastoid chondrosarcomas can present with sudden onset facial palsy.

Many of the patients presented with sudden onset facial palsy were misdiagnosed to have Bell's palsy.⁴ Bell's palsy involves facial nerve in the fallopian tube causing paralysis of all the modalities of facial nerve functions. Most of the cases recover on steroid treatment.^{5,4} Patients not recover on conservative management even for 8 – 10 weeks require further evaluation necessitating imaging of the temporal bone. The typical CT appearance of chondrosarcoma is of a destructive mass with scalloped erosive borders with intralesional calcifications.

Gross total resection followed by radiotherapy⁶ is the standard management protocol. The grade of the tumor reflect the prognosis. The five year survival rates of grade I is 90% whereas the same for Grade III is 43%.^{1,6} Proton beam radiotherapy achieving good control of the residual tumor growth.⁶

CONCLUSION: Chondrosarcoma of the mastoid bone causing isolated facial nerve palsy is extremely rare. However, it should be considered in the differential diagnosis of patients with Bell's palsy. Bell's palsy is a temporary facial palsy in which most of the patients will recover completely. Incomplete recovery or persisting facial weakness even after 8-10 weeks of conservative management prompts further investigations including imaging of the temporal bone. Early diagnosis of chondrosarcoma with gross total resection followed by radiotherapy offers optimal treatment with curative potential.

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Fig. 1: CT bone windows basal skull images showing osteolytic, irregular bordered, scalloping borders and intralesional calcifications and destructed bone extending upto jugular foramen anteriorly, retromastoid region posteriorly and C2 transverse process inferiorly.

Fig. 1

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Fig. 2: MRI Skull base showing T1W isointense, T2W hyperintense, STIR hyper intense tumor extending same as above with minimal contrast enhancement.

Fig. 2

Fig 3: Intraoperative images showing mastoidectomy region and Retromastoid region. Structures defined are Superior semicircular canal, Facial nerve, Mastoid antrum, Tumor tissue, Mastoid tip, Sternocleidomastoid muscle and Middle fossa dura.

Fig. 3

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