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Case Report

A rare case of temporal bone chondrosarcoma

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ABSTRACT

A rare case of lateral skullbase lesion, in a 24-year female, presented with moderately severe conductive type hearing loss, and facial paralysis. Audiological and radiological investigations were done which could not provide a reasonably certain diagnosis. So the patient was taken up for surgery. A subtotal pertrosectomy with blind sac closure was done. The histopathological analysis showed the lesion to be moderately differentiated chondrosracoma, an osseocartilagenous malignancy of the temporal bone. Due to rare nature, difficult location near vital structures, local invasiveness and potential for complications, diagnosis and treatment is difficult.

Keywords: Chondrosarcoma, Petrous apex lesion, Temporal bone chondrosarcoma, Osseo-cartilagenous tumour of skull

INTRODUCTION

Chondrosarcomas are slow growing, locally aggressive malignant tumour of cartilagenous origin mainly found among the long bones of the body. Their presence in the temporal bone or skull base is rare and account for only 0.15% of all intracranial tumours. The location within the temporal bone accounts for 27% among them.[1] It can be either primary or develop as a malignant transformation of benign temporal bone lesions such as an enchondroma or osteochondroma.[2] In the temporal bone, they generally arise from the congenital cell rests in the region of membranous synchondroses or suture lines, such as in the foramen lacerum region where the spheno-petrosal, spheno-occipital and the petro-occipital synchondroses converge.[3,4]

CASE REPORT

A 24-year-old female presented to our tertiary care centre with the complain of decreased hearing and tinnitus in the right ear since 1 year, gradually progressive. She also developed right sided grade III facial paralysis since last 3 months.[5] On otoscopic examination, a soft tissue mass was seen in the external auditory canal. Audiological test in the form of pure tone audiometry was done. It showed a moderately severe conductive type hearing loss with approximately 55 dB air-bone gap. Radiological investigations of high-resolution computed tomography (HRCT) temporal bone and contrast enhanced magnetic resonance imaging (MRI) was done (Figure 1a and b). They showed 3×4×5 cm lesion in temporal bone with aggressive local invasion.

The patient was posted for surgery with the plan of treatment being a subtotal temporal bone resection with blind sac closure by post aural approach. The superfical portion of the tumour mass exposed showed a gelatinous consistency which could be suctioned out easily (Figure 2a). The mastoid bone was drilled posteriorly till the sigmoid sinus, inferiorly till the jugular dome (Figure 2b). The petrous bone was drilled superiorly till exposure of the superior petrosal sinus and petrous apex, skeletonising the carotid artery as much as possible (Figure 2c). The lateral portion of the carotid canal within the petrous temporal bone was dehiscent. The mass was separated from all its

surrounding structures and sent for histopathological typing. The resulting cavity was plugged by transposition of temporalis and occipitalis pedicled muscle flap and abdominal fat plug. Blind closure of the external auditory canal was done. Intraoperatively, inner ear structures were found to be relatively uninvolved, therefore no drilling was done in that area.

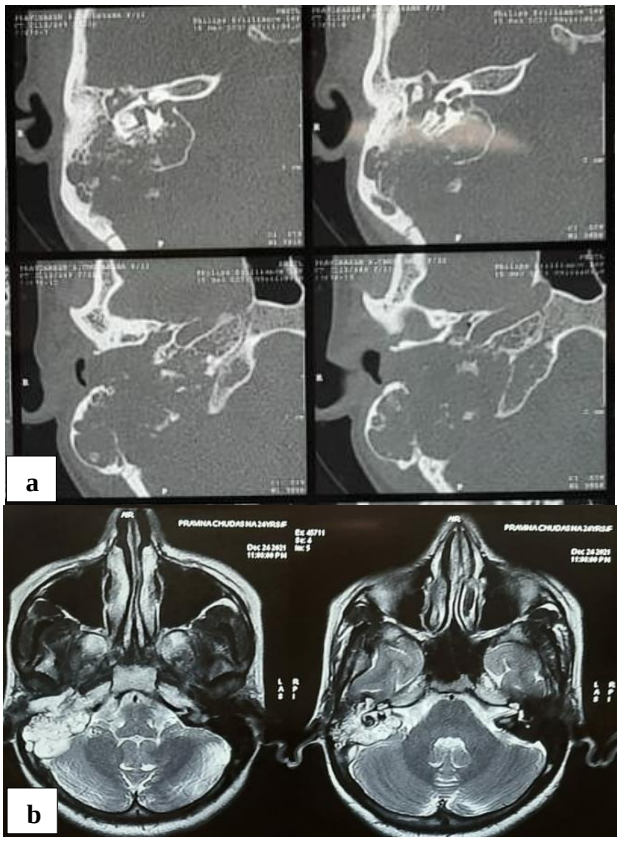

Figure 1: (a) and (b) Radiology showing heterogenously enhancing soft tissue mass with multifocal areas of blooming within the lesion suggestive of intralesional calcifications, involving the right external, middle and inner ear components, invading into posterior cranial fossa displacing the right cerebellum and jugular fossa region, thrombosing the right sigmoid and superior sagital sinus with bony destruction of mastoid and petrous portion of temporal bone with erosions of the carotid canal, extending till the clivus and C1 vertebral arch. The lesion showed heterogenous moderate post contrast enhancement.

There were no significant post-operative or intraoperative complications. There was no change in post-operative hearing and facial paralysis degree as compared to that before surgery. Post-operative stay was uneventful and she was discharged after 5 days.

The histopathological examination was suggestive of grade II (moderately differentiated) chondrosarcoma.[6] The patient was advised for post-operative radiotherapy. The patient refused for it and was therefore kept under regular follow up. On the MRI done at 18-months post op, there has been no findings suggesting any recurrence.

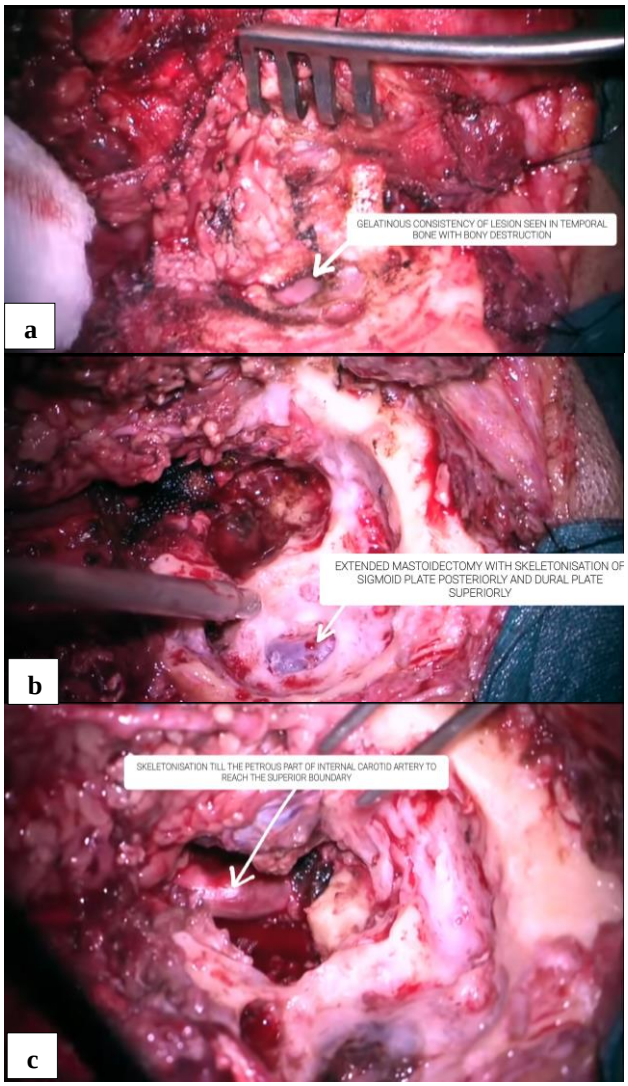

Figure 2: Intra op findings- (a) mastoid bone exposed to show a gelatinous tumour with surrounding mastoid cortex erosions, (b) extended mastoidectomy performed with dural and sigmoid skeletonisation, (c) subtotal petrosectomy done with tumour removal from carotid canal region skeletonising the petrous part of internal carotid artery.

DISCUSSION

Chondrosarcoma of the lateral skullbase is a relatively rare entity that accounts for 6% of skullbase neoplasms and 0.15% of all intracranial tumours.[1] The diagnosis of these cases become difficult because of the unusual presentations in these cases.

In our case, the patient was a young female, with complains of hearing disability and facial nerve paralysis with clinical examination showing a mass in the ear canal. Our first diagnostic suspicions were towards an extensive cholesteatoma with intratemporal complications. But since

she had no previous history of any ear discharge, it was quickly ruled out. Looking at the age and sex of the patient, a malignancy such as squamous cell carcinoma, seemed to be less likely. The most probable diagnosis we were suspecting was that of a benign pathology like paraganglioma (glomus tumours) or schwanomma. In order to aid our diagnosis and to decide a suitable surgical treatment plan, radiological and audiological investigations were done. The radiological findings of extensive bony and soft tissue destruction showed that the lesion was invasive in nature, and non-vascular nature of the lesion (as seen by heterogeneous moderate contrast uptake with internal calcifications) lead us away from our initial suspicions. More differentials were added to our list such as malignancies, and granulomatous diseases. This was the point where relatively rarer and novel diagnoses were thought about. The intraoperative findings of myxoid consistency of tumour gave us a clue that it could be of osseo-cartilaginous origin.

CONCLUSION

Chondrosarcomas of the temporal bone are a rare entity making its diagnosis and treatment difficult. In such cases, diagnostic confirmation can only be by post-operative histopathological analysis. Surgery is proposed as the mainstay of treatment for such osseo-cartilagenous lesions by ENT surgeons with further treatment in terms of need for radiotherapy, be decided after a confirmed histopathological report is obtained with immunohistochemistry as required.

Another major drawback in these cases is deciding the extent of resection for ideal treatment. Due to the difficult location and locally aggressive nature of these tumours, the operating surgeon walks a very thin line between adequate tumour removal and vital structural (cranial nerves, dural venous sinuses, and internal carotid artery) preservation. Adjuvant radiotherapy and long term follow up to look out for recurrence are deemed necessary for optimal treatment. Further research and need for development of fixed treatment protocols for such cases is needed to aid the treating surgeon to overcome these hurdles.

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