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Chondrosarcomas of the temporal bone: presentation and management

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Abstract
Chondrosarcomas (CSA) of the temporal bone are rare primary malignant tumours that are slow growing, but locally aggressive. The management of CSAs involving the temporal bone is challenging and necessitates a multidisciplinary approach in a tertiary referral unit well practised in skull base surgery. Their management with particular reference to modes of presentation and treatment strategies has been reviewed here.

Key words: Chondrosarcoma; Skull Base Neoplasms; Temporal Bone

Introduction

Chondrosarcomas (CSA) of the temporal bone are rare primary malignant tumours that may arise *de novo* or as a malignant transformation of benign chondromas. They are slow growing, but locally aggressive neoplasms with a propensity for recurrence if inadequately treated.[1]

Due to their rarity, their presentation with innocuous symptoms and their locally aggressive nature, the management of CSA involving the temporal bone is challenging.[2,3] This is further complicated by the fact that there are no large series in the literature, and that in the past CSAs of the temporal bone may have been clinically mistaken for multiple sclerosis, glomus jugulare, meningiomas and chondromas. Histological proof is the only definitive diagnostic tool; however, even this is not infallible and CSAs have been histologically misinterpreted as chondromas, osteochondromas and chondroblastomas.

High resolution imaging in the form of computerized tomography (CT) and magnetic resonance imaging (MRI) are the investigations of choice, and surgical resection with post-operative radiotherapy the cornerstone of treatment. Complex surgical clearance is difficult and recurrence rates are high. Post-operative cranial nerve palsies may occur and lead to significant deterioration in the patients' quality of life. Hence management of these tumours necessitates a multidisciplinary approach in a tertiary referral unit well practised in skull base surgery.

Methods

Three patients with CSAs of the temporal bone have been treated at Addenbrookes Hospital, Cambridge, under the care of the senior author (DAM), over the past 10 years. Their management has been reviewed here, with particular reference to modes of presentation, examination findings, investigations, treatment strategies, morbidity and survival (Table I).

TABLE I
Management of chondrosarcomas of the temporal bone

Case	Sex/Age	Location	Symptoms	Management	Morbidity	Follow up
1	F/55	Right petrous apex	3.5 years R hearing loss, tinnitus and headache	R retrosigmoid, retrolabyrinthine approach Post-operative radioptherapy R tarsorhaphy (temporary)	V to XII CN palsies – all recovered VII – improved to grade IV	8 years
2	F/50	Right petrous apex, involving the CPA and extending to the cavernous sinus	10 years R hearing loss, otalgia, vertigo, tinnitus, headache R facial paraesthesia	R translabyrinthine and middle fossa approach Post-operative radiotherapy	VII (Grade VI) CN palsy VI and partial V CN palsies disequilibrium	Disease free at 5 years
3	F/45	Left petrous apex, involving the jugular foramen and clivus	3 years L hearing loss and headache	Pre-operative embolization trans and infratemporal fossa approach Post-operative radiotherapy	Cranial nerves VI to XII palsies swallowing normal	Disease free at 6 years

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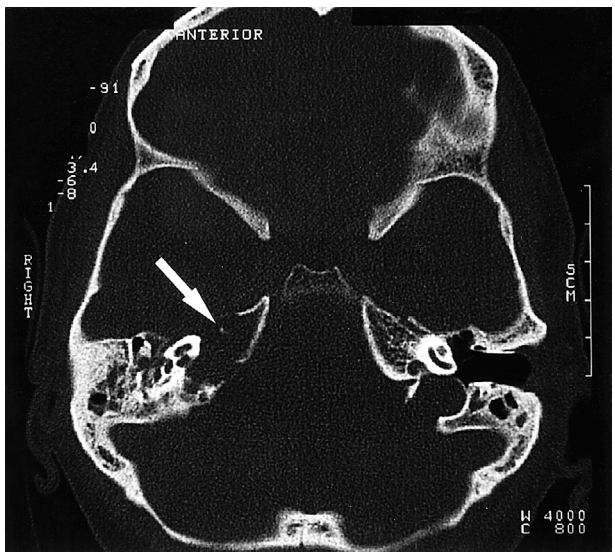


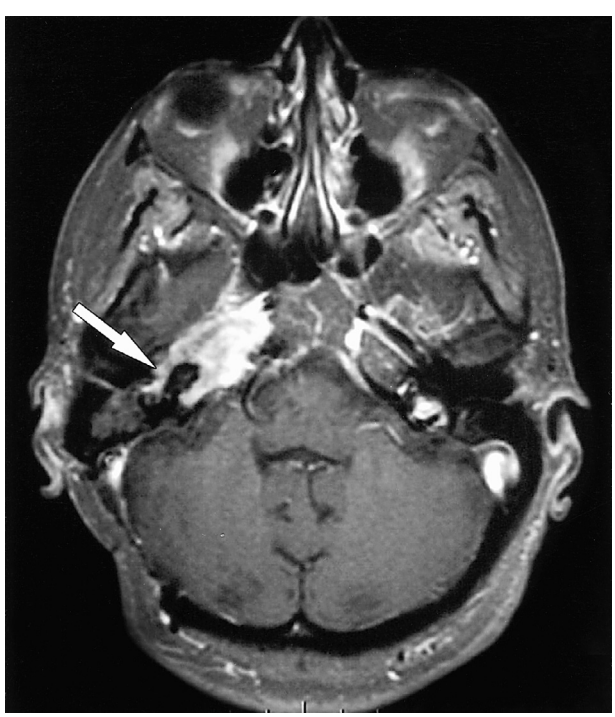
FIG. 1 (*Case 1*)

1(a) Axial CT showing erosion of the right petrous apex by a large chondrosarcoma.

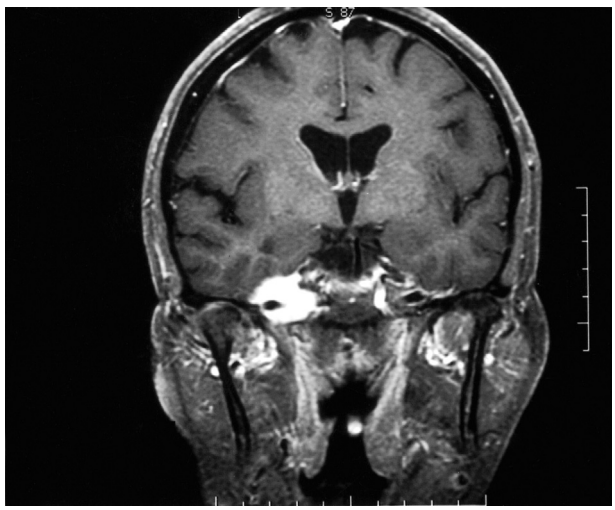
Case report

Case 1

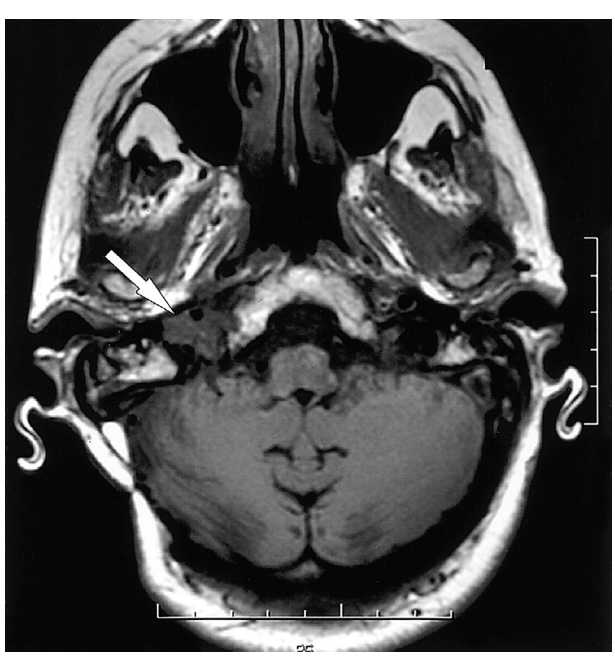
A 55-year-old female presented with a three-year history of right-sided distorted hearing and tinnitus and a six-week history of episodic rotatory vertigo and nausea. Her audiogram demonstrated an average of 25 dB air conduction thresholds and absent caloric responses on the right side. A CT scan of the temporal bone revealed a lytic lesion in the right petrous apex (Figure 1(a)), which enhanced with gadolinium-DTPA on MRI (Figure 1(b) and (c)). The patient declined surgery and three years later presented again but this time with diplopia, secondary to the right abducent nerve palsy. A further CT scan revealed that the mass in the right petrous apex had almost doubled in size. She agreed to undergo excision of the petrous apex lesion by a combined retro-sigmoid retrolabyrinthine and middle fossa approach. Most of the tumour was successfully excised, but a cuff of tissue had to be left around the cavernous sinus in order to limit further morbidity. The defect was repaired using fat and fascia from the right thigh. Post-operatively she developed partial palsies of the Vth to XIIth cranial nerves on the right-hand side, which required intensive rehabilitation. Within six months her cranial nerve functions had improved substantially except for the facial nerve weakness, (House-Brackman grade III). She underwent a single dose of post-operative radiotherapy, following which her facial weakness worsened to a Grade IV (HB). A year later, she developed numbness in the region of the 2nd and 3rd divisions of the trigeminal nerve. The cuff of tissue left around the cavernous sinus has remained stable since her post-operative radiotherapy and the patient remains well at eight years follow up with no further neurological symptoms (Figure 1(d)).



1(b) Axial MRI T1-weighted scan with gadolinium-DPTA showing a contrast enhanced lesion involving the petrous apex on the right surrounding the intrapetrous carotid artery.



1(c) Coronal MRI T1-weighted scan with gadolinium-DPTA showing a contrast enhanced lesion involving the petrous apex on the right surrounding the intrapetrous carotid artery.



1(d) Axial T1-weighted MRI scan at eight years post-operatively. Arrow pointing to the residual tissue around the cavernous sinus.

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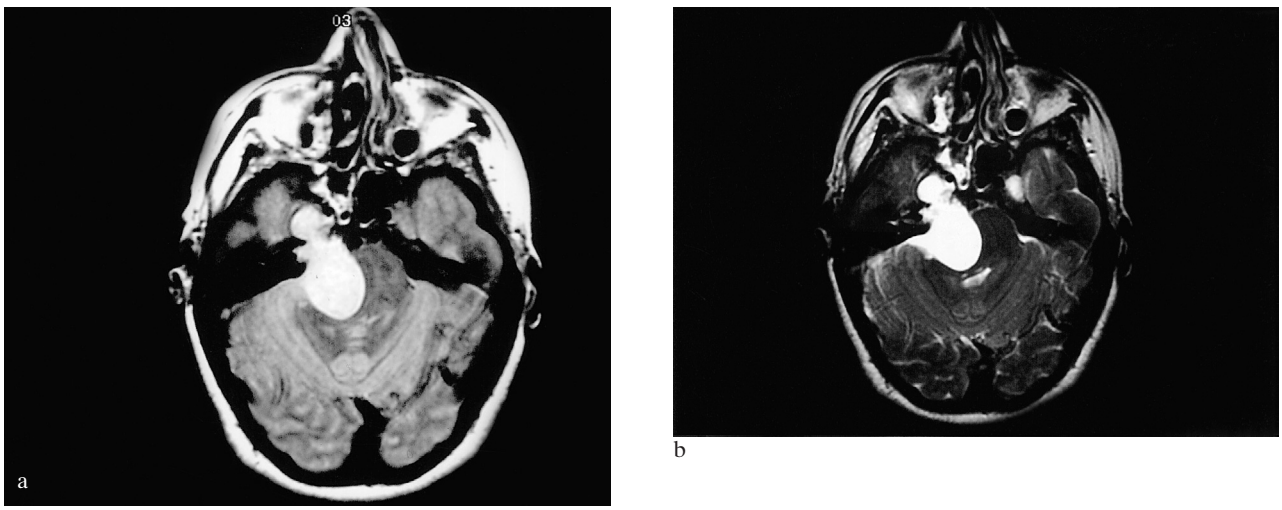
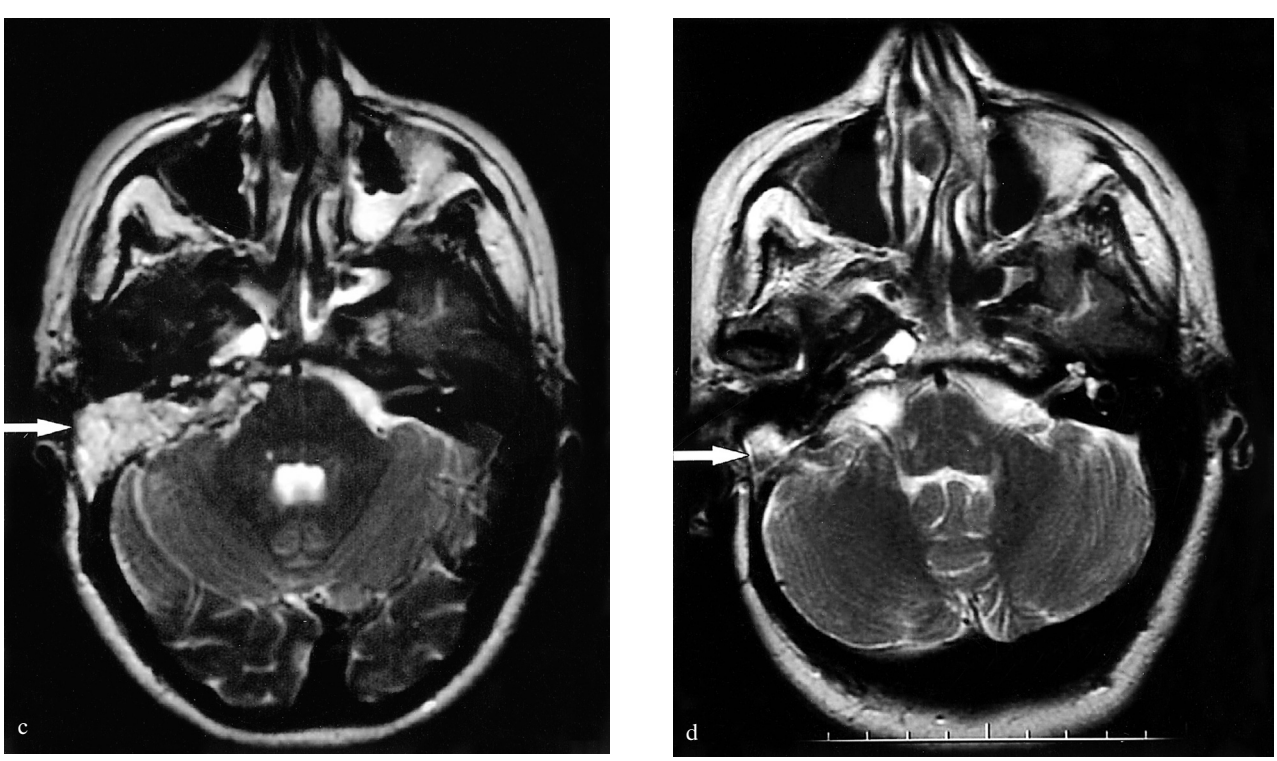


FIG. 2 (*Case 2*)

2(a) T1-weighted and 2(b)- T- weighted axial MRI scans with contrast demonstrating a huge right middle fossa, posterior fossa and petrous apex chondrosarcoma causing significant compression of the brain stem.

Case 2

A 50-year-old female presented with a 12-month history of right-sided deafness, vertigo and high- pitched tinnitus. On examination she had diminished blink and corneal reflexes and a facial nerve palsy (House-Brackman grade 3) on the right side, and a moderate degree of disequilibrium. The pure tone audiogram demonstrated an average of 85 dB sensorineural hearing loss in the right ear. An MRI scan demonstrated a large mass in the middle and posterior cranial fossa, and petrous apex that was causing significant distortion of the brain stem (Figures 2(a) and 2(b)). A translabyrinthine approach was used to resect most of the tumour; however, there was some tumour involvement around the intrapetrous carotid artery and cavernous sinus that was left *in situ*. The defect was repaired using fat and fascia again harvested from the thigh. Histopathological examination of the tissue demonstrated a moderately differentiated CSA. The facial nerve was left in continuity and preserved, however she went on to develop a complete facial palsy two weeks post-operatively. She also had a right VIth and a partial Vth cranial nerve palsy post-operatively. She underwent single dose post-operative radiotherapy and follow-up scans show no sign of recurrence at five years. The MRI images in Figures 2(c) and 2(d) demonstrate the fat and fascia repair of the right temporal bone post-operatively at one year and five years respectively. The axial T2-MRI scans show some degree of resorption of fat and remodelling of the scar tissue over time. There is significant contraction of the repair after five years.



2(c) One year post-operative and 2(d) Five years post-operative axial T2-weighted MRI scans demonstrating the fat and fascia repair and how this is remodelled with the passage of time.

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Case 3

A 45-year-old female presented with a three-year history of headache associated with left-sided deafness and five months of diplopia on left lateral gaze. On examination she had left abducent nerve palsy and a reddish mass behind the left eardrum. An audiogram demonstrated an average 80 dB air conduction threshold in the left ear. A CT scan demonstrated erosion of the jugular foramen, the left side of the clivus and the tip of the left petrous temporal bone. Initially this was thought to be a vascular tumour and hence the patient underwent pre-operative embolization followed by surgery. The tumour was excised via a left transtemporal infratemporal fossa approach. The intrapetrous carotid artery and the cavernous sinus were separated from the tumour, thus enabling a complete resection of the lesion to be undertaken. Histology surprisingly reported a well-differentiated CSA. Post-operatively the patient developed palsies of the cranial nerves VI to XII but her swallowing recovered sufficiently for her to have a normal diet on discharge. Two months after surgery she had a single dose of radiotherapy and the patient remains well at six years follow up with no sign of recurrence.

- **Chondrosarcoma of the temporal bone is rare**
- **This paper highlights management strategies that were used for dealing with this tumour in three patients**

Discussion

Chondrosarcomas account for about 10 to 20 per cent of all malignant primary bone tumours.[1] They are rare within the head and neck, usually occurring in the jaw or nasal cavity,[1,4] and even rarer inside the skull, accounting for six per cent of all skull base tumours. The majority of intracranial CSAs occur in the skull base and develop by endochondral ossification (a pre-requisite for CSAs), unlike the skull vault CSAs that develop by intramembranous ossification.[2,5] Temporal bone CSAs usually arise in the region of the foramen lacerum, where the spheno-petrosal, petro-occipital and spheno-occipital synchondroses converge. It has been suggested that CSAs arise from congenital cell rests within these regions.

Presentation and investigations

The mean age of presentation is in the fourth and fifth decades of life. The clinical manifestations of CSAs of the temporal bone include deafness, pulsatile tinnitus, vertigo, disequilibrium, aural fullness and headache. Cranial nerve symptoms are common and include diplopia, facial pain, hemi-facial spasm, facial palsy, dysphagia, hoarseness, shoulder weakness, and weakness or atrophy of the tongue.[5,6] A variety of inflammatory and neoplastic lesions can occur at the petrous apex and it is difficult to distinguish between them based on clinical history and examination alone. Imaging is, therefore, very helpful in the differential diagnosis of these lesions.

Radiology

Computed tomography and MRI are the investigations of choice for CSAs of the temporal bone. CT scan gives excellent demonstration of bony anatomy and erosion. The typical appearance of CSA is of a destructive lesion with patchy infiltration and a surrounding rim of calcification, although calcification within the tumour can be absent. MRI is superior to CT in soft tissue detail and helps to evaluate tumour involvement of neural and vascular structures. There is a marked degree of heterogeneous enhancement of CSAs with gadolinium-DPTA.[2,5]

Pathology

Many subtypes of CSA have been reported. The conventional subtypes consist of hyaline and myxoid areas or a combination of these. Mesenchymal and poorly differentiated subtypes are rare, aggressive and tend to present with advanced disease. CSAs generally consist of three different grades of tumour based on cellularity and nuclear atypia. Grade I tumours are well differentiated, grade II moderately differentiated, and grade III are poorly differentiated.[5,7] This form of grading is important because it reflects tumour biology irrespective of location or stage of presentation, and will give some indication in terms of prognosis.

Management

Treatment strategies include combinations of surgical debulking, complete surgical excision, radiotherapy and chemotherapy. Due to the rarity of CSAs of the temporal bone there are no large series in the literature and there are therefore no standard treatment regimes and no National Service Protocols.

Recent literature reviews recommend that total *en bloc* resection is better than de-bulking in terms of both local disease control and survival, but total excision is unfortunately associated with a higher cranial nerve morbidity rate.[3,5] More radical surgery has been facilitated by improvements in neuro-anaesthetic and microsurgical techniques and experience in dealing with such complex cases by undertaking them in a tertiary referral multidisciplinary unit where databases can be maintained.

Chondrosarcomas of the temporal bone are rare and very complex and hence their management needs to be customized for each individual patient, in terms of both quality and quantity of life. All three of the present cases underwent total or near total surgical excision of their tumours, where the approach was dictated by the access required and the site and extent of the tumour. Access and control are crucial, particularly in relation to dissection from the intra-petrous carotid artery and the cavernous sinus. In cases where the tumour was close to the carotid artery or the cavernous sinus a cuff of tissue was left *in situ*, and these were subsequently treated with radiotherapy. (e.g. *Cases 2* and *3*). It is evident from the literature that all patients with CSAs should be treated with post-operative radiotherapy.[1,5,6]

Conclusion

Chondrosarcomas, like many temporal bone lesions, present incidentally with non-specific symptoms. They are difficult to diagnose and early imaging permits identification of the disease process and its extent at an earlier stage. Management of these tumours is very challenging and requires a multidisciplinary skull base team. The key is to prolong quality and quantity of life – a balance between total resection and limited neurological complications, followed by post-operative radiotherapy.

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References

1 Burkey BB, Hoffman HT, Baker SR, Thornton AF, McClatchey KD. Chondrosarcoma of the head and neck. *Laryngoscope* 1990;**100**:1301–5
2 Waters GWR, Brookes GB. Chondrosarcoma of the temporal bone. *Clin Otolaryngol* 1995;**20**:53–8
3 Rapidis AD, Archondakis G, Anteriotis D, Skouteris CA. Chondrosarcoma of the skull base: review of literature and report of two cases. *J Craniomaxillofac Surg* 1997;**25**:322–7
4 Finn DG, Goepfert H, Batsakis JG. Chondrosarcoma of the head and neck. *Laryngoscope* 1984;**94**:1539–44
5 Neff B, Sataloff RT, Storey L, Hawkshaw M, Spiege JR. Chondrosarcoma of the skull base. *Laryngoscope* 2002;**113**: 134–9
6 Coltrera MD, Googe PB, Harris TJ, Hyams VJ, Schiller AL, Goodman ML. Chondrosarcoma of the temporal bone – Diagnosis and treatment of 13 cases and review of literature. *Cancer* 1986;**58**:2689–95
7 Lau DPC, Wharton SB, Antoun NM, Bottrill ID, Moffat DA. Chondrosarcoma of the petrous apex. Dilemmas in diagnosis and treatment. *J Laryngol Otol* 1997;**111**:368–71

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