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# Chondrosarcoma of the petrous apex. Dilemmas in diagnosis and treatment

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**Abstract**
A case of chrondrosarcoma of the petrous temporal bone is presented. Chondrosarcomas rarely occur intracranially and typically present as a petrous apex mass. The dilemmas faced in the diagnosis and treatment of petrous apex chondrosarcomas are discussed. This case also gives interesting insight into the natural history of this tumour.

**Key words: Chondrosarcoma; Petrous bone**

## Case report

A 55-year-old lady presented with a four-year history of right-sided hearing loss, tinnitus and increasing episodes of transient dysequilibrium. Neurootological examination was normal apart from a mixed right-sided hearing loss, with a conductive component of 20 db and loss of cochlear reserve at 2 kHz on pure tone audiometry. Impedance audiometry showed reduced compliance of the right tympanic membrane and negative middle-ear pressure on the left. Auditory brainstem responses showed marked prolongation of the I–V interpeak interval on the right and caloric testing showed a right canal paresis. A high resolution unenhanced computed tomography (CT) scan of the temporal bones was performed. This showed an expansile lytic lesion of soft tissue density at the petrous apex extending to the jugular foramen (Figure 1). There was no abnormal calcification or cyst formation within the lesion. A magnetic resonance imaging (MRI) scan demonstrated the mass to be isointense on T1 and hyperintense on T2 weighted images (Figure 2). There was homogeneous enhancement with gadolinium DTPA contrast (Figure 3). Selective carotid arteriography showed no tumour circulation and excluded the possibility of a glomus tumour. A technetium 99m bone scan was carried out to investigate the possibility of a metastatic tumour and was normal. The patient declined exploratory surgery and

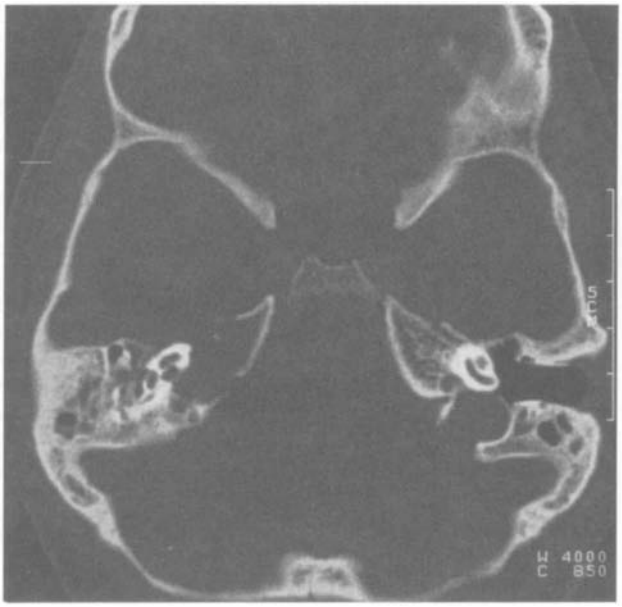


Fig. 1
CT scan showing lytic lesion of the right petrous apex.

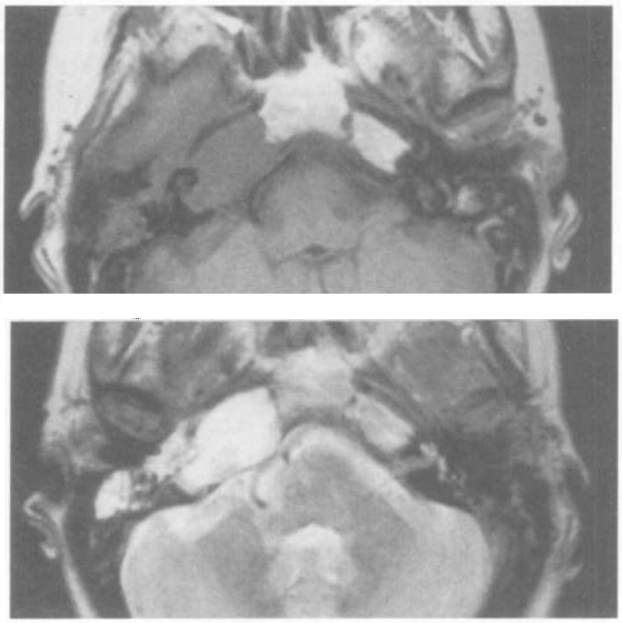

Fig. 2
Axial MRI demonstrating isointensity on T1 (top) and hyperintensity on T2 (bottom).

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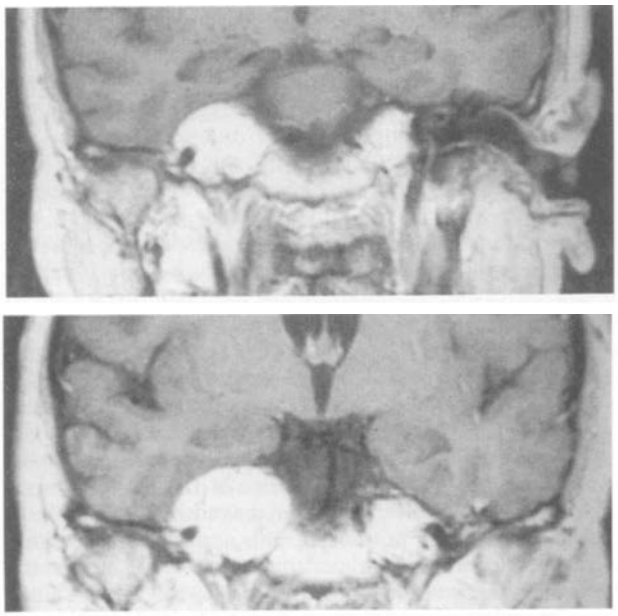

FIG. 3

Coronal MRI demonstrating homogeneous enhancement with gadolinium DTPA at presentation (top) and before surgery (bottom). Note enlargement of the lesion.

was therefore followed-up in the clinic. She remained well and a repeat MRI scan two years later showed no change in size of the petrous apex mass.

Three years after initial presentation the patient represented with intermittent diplopia on right lateral gaze. She was found to have a right abducent nerve palsy. A repeat MRI scan showed an increase in the size of the lesion (Figure 3) and she agreed to undergo surgery. The pre-operative working diagnosis was of a primary tumour of indeterminate nature.

At surgery, an initial Rosen's flap revealed an encapsulated tumour in the hypotympanum and eroding the bone of the promontory. The tumour was removed via combined middle fossa, retrolabyrinthine and retrosigmoid approaches. The tumour was cystic in nature and full of gelatinous material. It was peeled off cranial nerves VII to XII but total excision was impossible due to cavernous sinus involvement. Due to difficulty swallowing and vocal fold paralysis as a result of palsies of the lower four cranial nerves post-operatively the patient required a percutaneous gastrostomy. She received a course of post-operative radiotherapy.

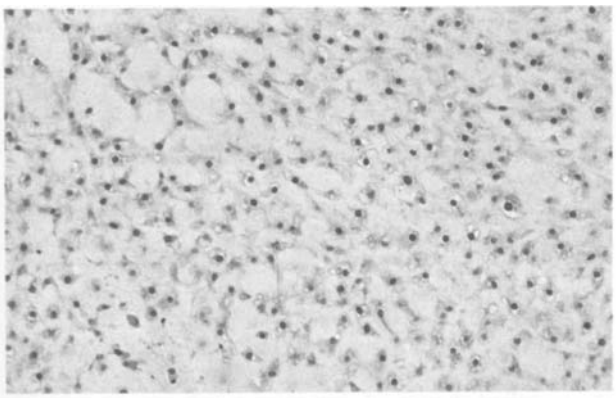

FIG. 4

Low power H & E section showing moderately cellular tumour with chondromyxoid stroma.

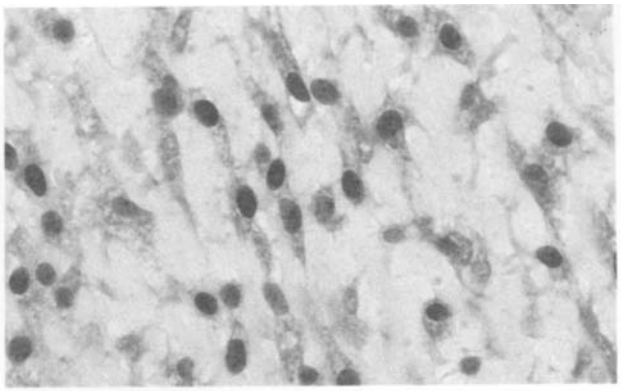

FIG. 5

High power view showing neoplastic cells with round nuclei. Some have cord-like or rather vacuolated cytoplasm.

## Pathology

The pieces of tumour measured in total approximately $3 \times 2 \times 1$ cm and macroscopically had a pale gelatinous appearance. Microscopic examination of haematoxylin and eosin stained paraffin sections showed the tumour to have abundant chondroid stroma (Figure 4). The neoplastic cells were arranged irregularly, either individually or in groups of two or three in a lacuna. In much of the tumour the stroma was more myxoid and the cells possessed more cytoplasm with a rather more 'chordomatous' pattern; some cells showed cytoplasmic vacuolation (Figure 5) but this was not a striking feature. The cells possessed small round nuclei with only mild nuclear pleomorphism and very rare mitotic figures (Figure 6). The tumour showed no evidence of necrosis. Immunohistochemistry revealed that the neoplastic cells expressed S100 and vimentin but not the epithelial markers cytokeratin and epithelial membrane antigen (Figure 7). A diagnosis of well-differentiated chondrosarcoma (grade II) of conventional type was made.

## Discussion

A chondrosarcoma is a primary malignant tumour of bone. It may occur in any bone that develops in cartilage. Although chrondrosarcomas form up to one third of all primary malignant bone tumours, intracranial chondrosarcomas are considerably rarer, constituting less than 0.16 per cent of all intracranial tumours (Berkmen and Blatt, 1968).

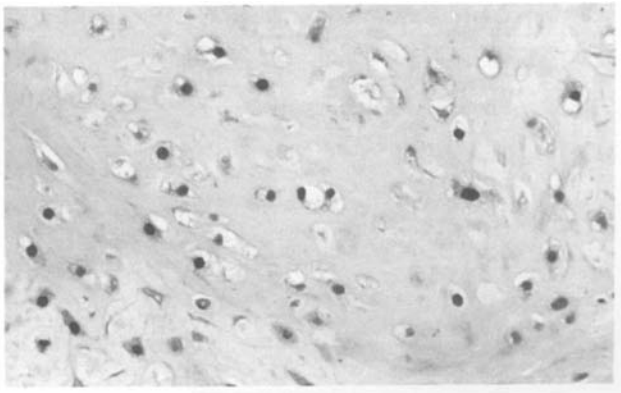

FIG. 6

High power view of a more cartilaginous area of the tumour. Neoplastic chondrocytes sit in lacunae in a chondroid stroma. Irregular cell distribution and mild nuclear pleomorphism is apparent.

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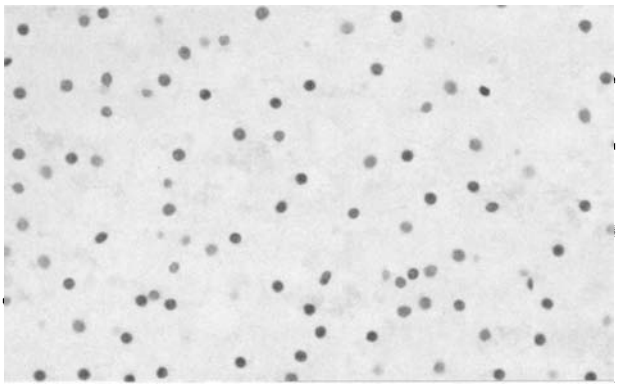

FIG. 7

Immunohistochemistry demonstrating negative staining of tumour cells for the cytoplasmic protein cytokeratin. Nuclei are demonstrated with a haematoxylin counterstain.

Embryologically the bones of the skull base ossify predominantly endochondrally whereas the skull vault ossifies intramembranously. This is thought to be the reason why intracranial chondrosarcomas tend to occur in the skull base. The tumour usually occurs in the region of the foramen lacerum. This is where the spheno-petrosal, petro-occipital and spheno-occipital synchondroses converge. It has been suggested that the tumour arises from congenital cell rests in this region (Jaffe, 1958). Similar tumours have been reported arising from dura (Scheithauer and Rubinstein, 1978), choroid plexus (Scott *et al.*, 1976) and membranous bone (Villani *et al.*, 1984). Alternative aetiological explanations for tumours in these sites have included metaplasia of fibroblasts and origin from primitive multipotent mesenchymal cells.

The mean age of presentation is in the fifth decade (Coltrera *et al.*, 1986). There is no sex bias. A variety of inflammatory and neoplastic lesions can occur at the petrous apex (Table I). It is usually difficult to distinguish between them on the basis of clinical history and examination. Unexplained abducent nerve palsy should prompt an investigation of the petrous apex. Compression or irritation of the nerve occurs as it traverses the supero-medial aspect of the petrous apex in the petro-clinoid ligament or Dorello's canal. Trigeminal nerve involvement may also occur resulting in deep aural or retro-orbital pain. Affectation of these cranial nerves may however be a late presenting feature. It is significant that our case presented initially with non-specific otological symptoms. These preceded the onset of other focal neurology by seven years. This corresponds with observations by Kveton *et al.* (1986) who noted that VIIIth nerve dysfunction was the initial presenting symptom in four out of a series of five patients with chondrosarcoma of the skull base. These findings emphasize the importance of thorough investigation of unexplained and in particular unilateral VIIIth nerve dysfunction. Involvement of the lower cranial nerves suggests a posterior fossa location (Oguro *et al.*, 1989).

TABLE I

DIFFERENTIAL DIAGNOSIS OF PETROUS APEX LESIONS

| | | |
|---|---|---|
| 1. | Normal variants | |
| | a. | Large air cell |
| | b. | Bone marrow asymmetry |
| 2. | Inflammatory | |
| | a. | Effusion |
| | b. | Mucocoele |
| 3. | c. | Petrous apicitis |
| | d. | Cholesterol granuloma |
| | e. | Cholesteatoma |
| 3. | Neoplasms | |
| | b. | Meningioma |
| | c. | Schwannoma |
| | d. | Paraganglioma (glomus tumour) |
| | e. | Chondroma |
| | f. | Chondrosarcoma |
| | g. | Osteosarcoma |
| | h. | Chordoma |
| | i. | Metastatic tumour |

CT, MRI and angiography are the main investigations for suspected petrous apex lesions. They are often helpful in suggesting a diagnosis. CT is usually the initial investigation. It has the advantage of providing detailed images of bone, the primary constituent of the petrous apex. Virtually all petrous apex lesions except for an effusion within a pneumatized apex are detectable as bone erosive lesions on CT. The nature of the eroded margin may give some indication of the diagnosis. Irregular bony erosion occurs in petrous apicitis, meningioma, glomus tumours, chordoma, chondrosarcoma and metastases. Smooth margined erosion typically occurs in schwannomas, chondromas and some metastatic lesions (Jackler and Parker, 1992). Intravenous contrast enhancement may be useful in distinguishing neoplastic from inflammatory lesions. In addition foci of calcification within the tumour creating a 'popcorn' appearance should increase suspicion for a chordoma or chondrosarcoma.

MRI imaging is assuming a greater role in the investigation of petrous apex lesions and is proving helpful in the differential diagnosis. Tumours are typically isointense on T1, hyperintense on T2 and demonstrate enhancement with gadolinium DTPA (Bourgouin *et al.*, 1992; Jackler and Parker, 1992). Cholesterol granulomas are hyperintense on both T1 and T2 images. Cholesteatomas, which are also hyperintense on T2, are usually of low signal on T1 although may occasionally appear isointense and thus similar to tumours. However neither cholesterol granulomas nor cholesteatomas enhance with gadolinium. When used in conjunction with CT scanning, MRI improves the accuracy of pre-operative diagnosis of petrous apex pathology. A primary bone tumour was the most likely diagnosis in our case in view of the location of the lesion as well as its signal and enhancement characteristics. In addition to diagnosis, scanning is essential for surgical planning and detecting residual or recurrent tumour at follow-up.

Histologically the conventional type of chondrosarcoma is characterized by the production of neoplastic hyaline cartilage which may show foci of myxoid change and calcification. Atypia in neoplastic chondrocytes can be subtle in well-differentiated lesions but there tends to be relative crowding of nuclei, double nucleation and hyperchromasia. Several studies have demonstrated the prognostic utility of grading (I to III) based on mitotic rate, cellularity and nuclear size (Evans *et al.*, 1977). At the skull base the well-differentiated chondrosarcoma must be differentiated histologically from a chordoma which is a slowly growing, locally destructive neoplasm derived from the notochord. In the skull base these typically arise in the midline from the clivus. Classically they consist of intersecting cords of cells, some of which are distinctively vacuolated (physaliphorous cells), in a mucoid stroma. The differential diagnosis is a particular problem in the case of chordomas showing pronounced cartilaginous features. Controversy exists surrounding the existence and nature of the so-called chondroid chordoma which may be morphologically indistinguishable from a well-differentiated chondrosarcoma. Immunohistochemistry is considered to be of value in making the distinction. Both tumours express S100

and the mesenchymal marker vimentin, but whereas chordomas and chondroid chordomas also express epithelial markers such as cytokeratin and epithelial membrane antigen (due to their notocord origin), chondrosarcomas do not (Wojno *et al.*, 1992). The immunohistochemical profile in this case, and the origin from the petrous apex rather than the clivus, supports a diagnosis of well-differentiated chondrosarcoma rather than chondroid chordoma.

Surgical excision is the treatment of choice for chondrosarcoma of the petrous apex. Due to the inaccessibility of the lesion intra-operative frozen section is essential to establish a working diagnosis. An exploratory 'burr hole' drilled inferior to the cochlea and superior to the jugular bulb has been described (Brackmann and Giddings, 1994) and may be a suitable route for achieving this aim. Middle fossa, translabyrinthine, transcochlear and infra-temporal fossa approaches to the petrous apex have been described (Kveton *et al.*, 1986; Charachon *et al.*, 1990). Total removal of the tumour is often difficult because of invasion of the cranial nerves, arteries and vital centres of the brainstem (Oguro *et al.*, 1989). Post-operative radiotherapy has been used although its efficacy in controlling tumour recurrence is in doubt (Harwood *et al.*, 1980; Suit *et al.*, 1982).

Prognosis can be predicted by histological grade. In a study of 71 chondrosarcomas, Evans *et al.* (1977) reported five year survival rates of 90 per cent, 80 per cent and 40 per cent for grade I, II and III tumours respectively. The incidence of metastases is also affected by histological grade. In Evans's series no grade I tumours metastasized whereas up to 70 per cent of grade III tumours did. In a review which included 21 intracranial chondrosarcomas, Hassounah *et al.* (1985) reported an overall metastatic rate of 15 per cent. Extremity lesions may carry a more favourable prognosis than head and neck lesions. It is likely that this is related to the difficulty of surgical access to the petrous apex. This may also help to explain the high rate of local recurrence of the tumour.

Chondrosarcoma of the petrous apex is rare. As each major centre treats only a limited number of cases it is difficult to perform randomized controlled trials. Perhaps a central cancer registry should co-ordinate trials of rare tumours on an international basis.

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