Chondroid chordoma of petrous temporal bone with extensive recurrence and pulmonary metastases

ABSTRACT
Chordoma is considered a slow-growing low-grade malignant tumor arising from remnants of the primitive notochord. Chondroid chordoma is a variant of chordoma, with a cartilaginous component. Primary chondroid chordoma of the petrous temporal area is extremely rare; only three case reports are there in the English literature. Occurrence of distant metastases in chordoma is rare and ranges from 5-40%, mostly occurring in patients with advanced disease. We report a histologically proven case of petrous temporal chondroid chordoma with extensive recurrence and pulmonary metastases.

CASE REPORT
This 32-year-old right-handed gentleman was first operated in 2002 for chondroid chordoma of the left petrous temporal bone. Following surgery, he developed facial nerve palsy which persisted. He had a recurrence and was reoperated on two occasions, the last surgery being in February 2004. Near-total excision could be achieved only in the first surgery. There were small residual lesions after both re-explorations. In the course of time he developed a communicating hydrocephalus, and a theco-peritoneal shunt was placed 2 months back for the same.

He presented with a localized swelling in the left retroauricular region, recurrence of headache, mild cough during swallowing, and hoarseness of voice for 1 month. His general and systemic examinations were normal. His higher mental functions were normal. Cranial nerve examination revealed involvement of the left Vth, VIth, VIIth, VIIIth, IXth, XIth, and XIIth cranial nerves. There were no motor or sensory deficits. He had gait ataxia and left-sided cerebellar signs. Blood investigations were normal. A recent MRI showed a large recurrent heterogeneous lesion, which was hypointense to isointense on T1-weighted images and showed high signal on T2-weighted images. The lesion was irregularly enhancing on contrast MRI and was found to have caused destruction of the left petrous temporal bone, compressing the brainstem and extending into infratemporal fossa [Figure 1a and b]. Chest X-ray showed multiple cannonball lesions in both lungs [Figure 2]. In view of the large local recurrence and the presence of pulmonary metastases, the patient was advised radiotherapy. While on treatment, he developed aspiration pneumonia and succumbed to it.

DISCUSSION
Chordomas exclusively involve the axial skeleton and the most common site is the sacrococcygeal region (50%), followed by the skull base (35%) and the vertebral bodies (15%). Chordoma accounts for approximately 1% of all intracranial tumours and the retroclival region is the most common site. Petrous temporal chordoid chordoma is extremely rare. Chordomas usually occur between the fourth and fifth decades of life, whereas chordoid chordomas present at an earlier age and has a female predilection. Cranial nerve palsies due to tumour extension into the neural foramina is the most frequent presentation and there may be associated headache, signs and symptoms of raised intracranial pressure, and pyramidal signs. Because of their proximity to vital structures, including the carotid and basilar arteries and the brainstem, the management of skull...
base chordomas is challenging. Although an extensive and aggressive surgical approach, using advanced microsurgical and skull base techniques, has been advocated to enhance the resectability, complete resection of skull base chordomas is often unsuccessful. Only less than 50% of them can be radically or totally resected, and there is a high recurrence rate even after complete resection.[1] Chordomas are not reported to be sensitive to chemotherapy. The role of radiotherapy as primary or adjuvant treatment has been debated. In patients who have local relapse and have a poor prognosis, both radiation and surgery are used as salvage therapy.[2] Because of invasion of the surrounding structures, in spite of their relatively benign histological behavior, chordomas of the skull base have a poor prognosis and high recurrence rate even after total surgical resection.

REFERENCES


Source of Support: Nil, Conflict of Interest: None declared.