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Review

Skull base chondrosarcoma

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ABSTRACT

Intracranial chondrosarcoma are rare tumours, accounting for approximately one in 1000 intracranial neoplasms. Although more common in the axial or appendicular skeleton, intracranial tumours present a challenging surgical and oncological problem. Chondrosarcoma have a predilection for the skull base and although commonly slow growing, Grade II and III lesions do occur. We present two illustrative patients from the Royal Melbourne Hospital, Australia, demonstrating dramatically differing presentation and clinical outcome and the diagnostic difficulties that may arise. A review of the literature regarding skull base chondrosarcoma is presented. We summarise the clinical, radiological and histological features. The evidence for surgical resection, radiotherapy and chemotherapy is presented and critically evaluated. Based on the available evidence, we advocate maximal safe resection, followed by radiotherapy for Grade II and III tumours. There is no current role for chemotherapy. Radical excision should not be attempted at the expense of neurological function.

1. Introduction

Chondrosarcomas are rare malignant tumours composed of cells derived from transformed chondrocytes and account for approximately 20% of skeletal system cancers [1]. They are a heterogeneous group of tumours that have diverse morphological features and clinical behavior. They commonly occur within the pelvis and shoulder or along the superior metaphysial and diaphysial areas of the appendicular skeleton. Only 2% of all chondrosarcomas occur within the skull base [2], accounting for 0.1–0.2% of all intracranial tumours [3]. They are generally very slow growing and erode the bones of the skull base; however higher grade tumours occur, characterised by rapid growth and early metastasis.

2. Case illustrations

2.1. Patient 1

J.E. is a 68-year-old woman who presented with headaches over a period of 12 months. The headaches were not consistent with any particular pattern and were not suggestive of raised intracranial pressure. Neurological examination was normal. Neuroimaging revealed a contrast-enhancing lesion eroding through the petrous bone into the cerebellopontine angle (Fig. 1–3). A retrosigmoid craniotomy was performed with the intracranial component being resected and a significant amount of the tumour within the skull base debulked. The patient made an uneventful recovery.

2.1.1. Histology and treatment

The histology showed a chondroid tumour forming sheets with a lobulated architecture (Fig. 4). The tumour demonstrated moderate cellularity with the cells being S-100 and M2A positive and CAM5.2 negative (Fig. 5). The Ki-67 index was less than 1% and the features were consistent with a Grade II chondrosarcoma. Targeted radiotherapy was advised given the grade of this tumour and incomplete excision.

2.2. Patient 2

M.H. is a 62-year-old woman who first presented in 1982 with an intracerebral haemorrhage predominantly in the left frontotemporal region requiring evacuation. She was found to have an extensive skull base lesion as the principle cause of her presenting haemorrhage. Her imaging showed a lesion that was centered off the midline and predominantly in the left middle cranial fossa. She underwent a frontotemporal craniotomy and debulking, of which the histology returned as a chordoma. She subsequently had a recurrence in 1987, 1996 and again in 2002, all of which warranted further open surgical debulking. In 2007, she had a further recurrence with extension into the nasopharynx and blocking of

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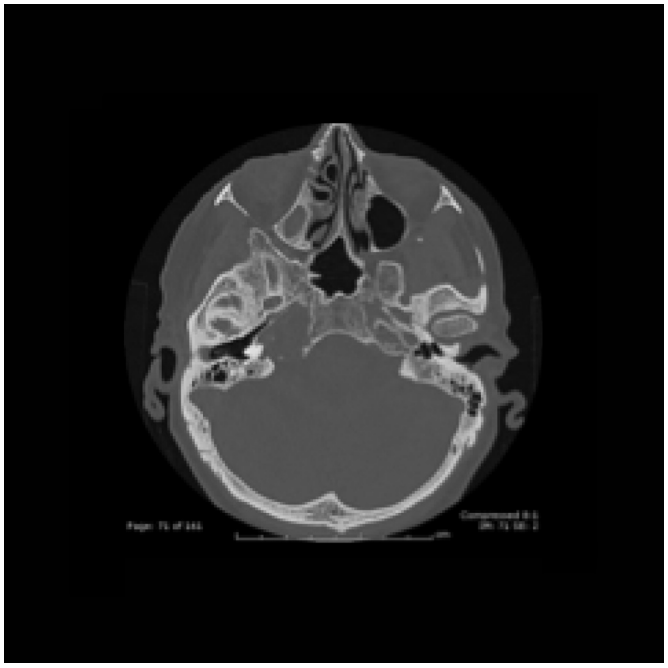

Fig. 1. Axial CT scan of the head (bone window) of Patient 1 showing destruction of the petrous ridge and to some extent the clivus on the right side.

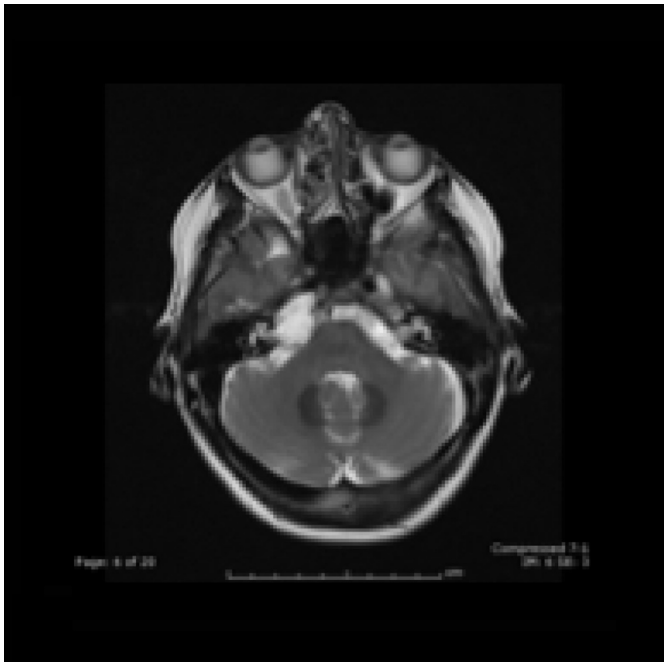

Fig. 2. T2-weighted axial MRI of Patient 1 showing a hyperintense lesion extending from the right petrous region into the cerebellopontine angle abutting the brainstem.

her Eustachian tube, for which she underwent an endoscopic transnasal/transsphenoidal debulking. This was complicated by a left cranial nerve VI palsy. She had a further endoscopic partial resection in 2013, at which time the histology was revised as chondrosarcoma (Fig. 6). The original histology was re-reviewed and this was considered to be more consistent with a chondrosarcoma and not a chordoma. She has subsequently re-presented with a further haemorrhage and multiple left sided cranial nerve palsies requiring a prolonged intensive care stay, a tracheostomy and feeding via a percutaneous gastrostomy tube.

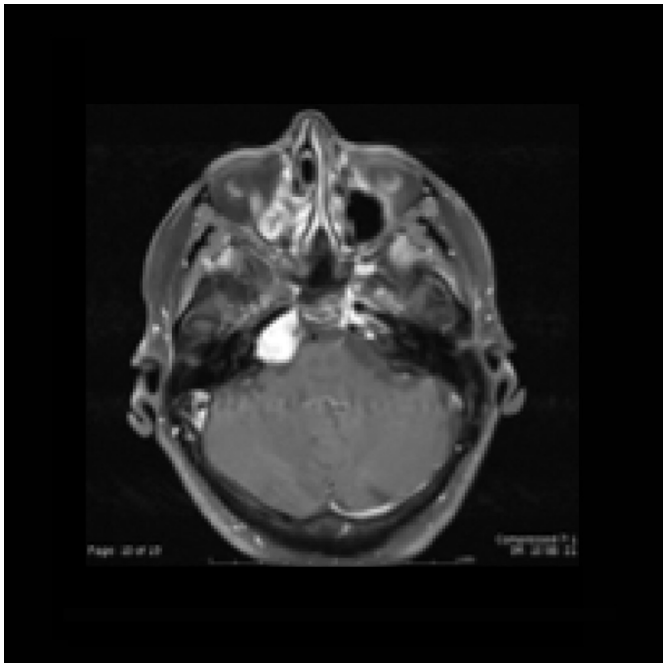

Fig. 3. Axial T1-weighted MRI with gadolinium enhancement of Patient 1 showing the same lesion avidly enhancing.

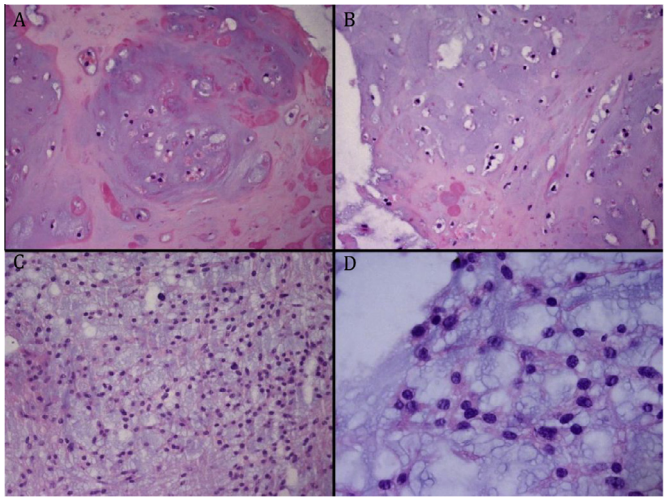

Fig. 4. Histopathology slides of the tumour from Patient 1 stained with haematoxylin and eosin showing chondroid tumour sheets with lobulated structure (A–C original magnification ×200; D original magnification ×400).

This case highlights the difficulties with diagnosis and treatment of these skull base tumours, certainly 30 years ago, but also to this day. It also exemplifies the degree of morbidity associated with the disease and its treatment.

3. Skull base chondrosarcoma

3.1. Epidemiology/presentation

Skull base chondrosarcoma occurs most commonly between the ages of 40 and 70 years. There may be a slight female predominance (1.1:1 female:male [4]) although many studies demonstrate no sex difference. Presenting symptoms vary according to the precise location of the lesion and the compression and/or