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Case Report

Skull Base Parachordoma/Myoepithelioma

Catherine de Cates, Daniele Borsetto, Daniel Scoffings, Dominic O'Donovan, Neil Donnelly

Department of ENT, Addenbrooke's Hospital, Cambridge University Hospitals NHS Foundation Trust, Cambridge, United Kingdom (CdC, DB, ND)
Department of Radiology, Addenbrooke's Hospital, Cambridge University Hospitals NHS Foundation Trust, Cambridge, United Kingdom (DS)
Department of Neuropathology, Addenbrooke's Hospital, Cambridge University Hospitals NHS Foundation Trust, Cambridge, United Kingdom (DO)

ORCID iDs of the authors: C.d.C. 0000-0002-9182-3664; D.B. 0000-0003-3464-2688; D.S. 0000-0002-0220-853X; D.O. 0000-0002-1766-1038; N.D. 0000-0003-3624-689X.

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Parachordoma is a rare soft tissue mixed tumor, associated with soft tissue myoepithelioma. It is typically growing slowly and considered less aggressive than other similar soft tissue tumors. However, it does recur sporadically, and on rare occasions, it has demonstrated the ability to metastasize. Although imaging is important, definitive diagnosis is achieved by histology, and it is typically treated by a wide local excision. We present the first reported case of a skull base parachordoma in a 15-year-old boy, managed with a wide local excision and with no signs of recurrence or metastases after 24 months of follow-up.

KEYWORDS: Parachordoma, skull base, neoplasm, myoepithelioma, temporal bone

INTRODUCTION

Parachordoma is a rare slow-growing tumor arising from soft tissue. It was first described by Laskowski in the 1950s as chordoma peripheric um, and then later by Dabska in 1977, who re-named it as parachordoma [1]. It was previously considered to be morphologically and histologically similar to chordoma (also referred to as central chordoma) but found in the non-axial location. Due to this similarity, the term parachordoma was applied [2].

More recently, new immunohistochemical and molecular tests have helped distinguish parachordoma from other soft tissue tumors such as chordoma and extraskeletal myxoid chondrosarcoma [1]. Since Laskowski first identified this lesion, 68 cases of parachordoma have been described in the literature. Although it is considered a locally invasive lesion, managed by surgical excision alone, there are reports of recurrence and metastases [1-14].

We describe a case of a young man who presented with a right temporal bone parachordoma. To the best of our knowledge, this is the first report of a skull base parachordoma.

CASE PRESENTION

A 15-year-old male presented with a short history of unsteadiness, headache, vomiting, diplopia, and right facial numbness. An examination revealed saccadic eye movements with secondary horizontal nystagmus toward the left and reduced hearing on the right side.

Computed tomography (CT) showed a large posterior fossa extra-axial tumor, with some erosion of the temporal bone. Magnetic resonance imaging (MRI) was performed. It showed a 7 cm extra-axial cerebellopontine tumor in the posterior fossa with a broad base on the posterior surface of the petrous temporal bone. There was obstructive hydrocephalus and compression of the right cerebellum, brainstem, and fourth ventricle (Figure 1). The surface of the temporal bone at the interface with the tumor showed numerous foci of erosion. The tumor was uniformly hyperintense on T2-weighted images and intermediate-to-low signal on T1-weighted images, with some small foci of low signal intensity corresponding to calcification on CT. It showed a heterogeneous

Corresponding Address: Catherine de Cates E-mail: catherine.de.cates@gmail.com
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pattern of contrast enhancement, with areas of non-enhancing cystic or necrotic change. There was no evidence of restricted diffusion within the tumor on diffusion-weighted imaging and no edema in the cerebellum adjacent to the tumor. No abnormal intratumoural or peritumoural vascular flow voids were present.

He was started on dexamethasone and transferred to our unit for surgical excision 5 days later.

Interpretation of the preoperative imaging suggested a large vestibular or lower cranial nerve schwannoma. The surgical approach used was that for a standard translabyrinthine resection of vestibular schwannoma. During the extended cortical mastoidectomy, soft jelly-like tumor was encountered. Intraoperative frozen section was suggestive of chordoma or chondrosarcoma. It became apparent that the tumor was extradural and arising from the temporal bone. The approach was changed to retro-labyrinthine with the tumor and any abnormal bone being removed: there was

Table 1. Summary of Immunohistochemical Findings

Antibody	Result
Brachyury	Negative
S100 Protein	Positive
Vimentin	Positive
EMA	Positive
Desmin	Negative
Actin HHF35	Negative
AE1/AE3 Pancytokeratin	Negative
MNF116 Pancytokeratin	Negative
CD34	Negative
CD117	Negative
SOX10	Negative
p63	Negative
INI1 (BAF47)	Retained nuclear labeling
Synaptophysin	Negative
CD99	Negative

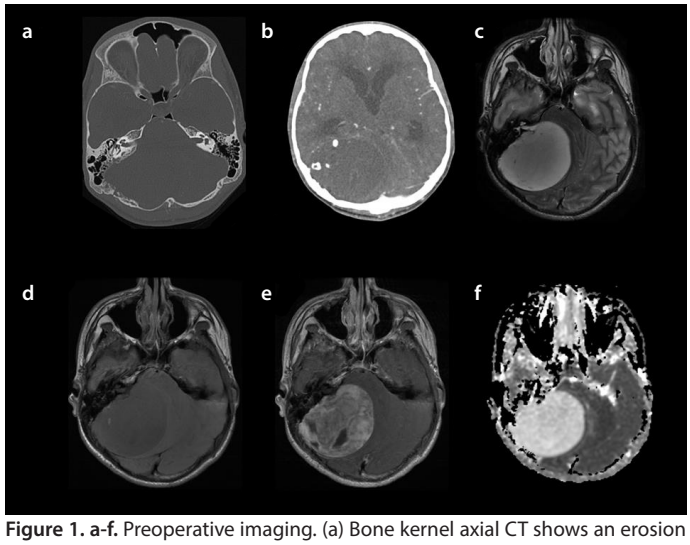

Figure 1. a-f. Preoperative imaging. (a) Bone kernel axial CT shows an erosion of the posterior surface of the petrous temporal bone. (b) Contrast-enhanced CT shows foci of calcification. (c) Axial T2W and (d) axial T1W show predominantly uniform signal intensity of the tumor. (e) Contrast-enhanced T1W shows heterogeneous pattern of enhancement. (f) Apparent diffusion coefficient map shows high intralesional signal, indicating no restriction of diffusion.

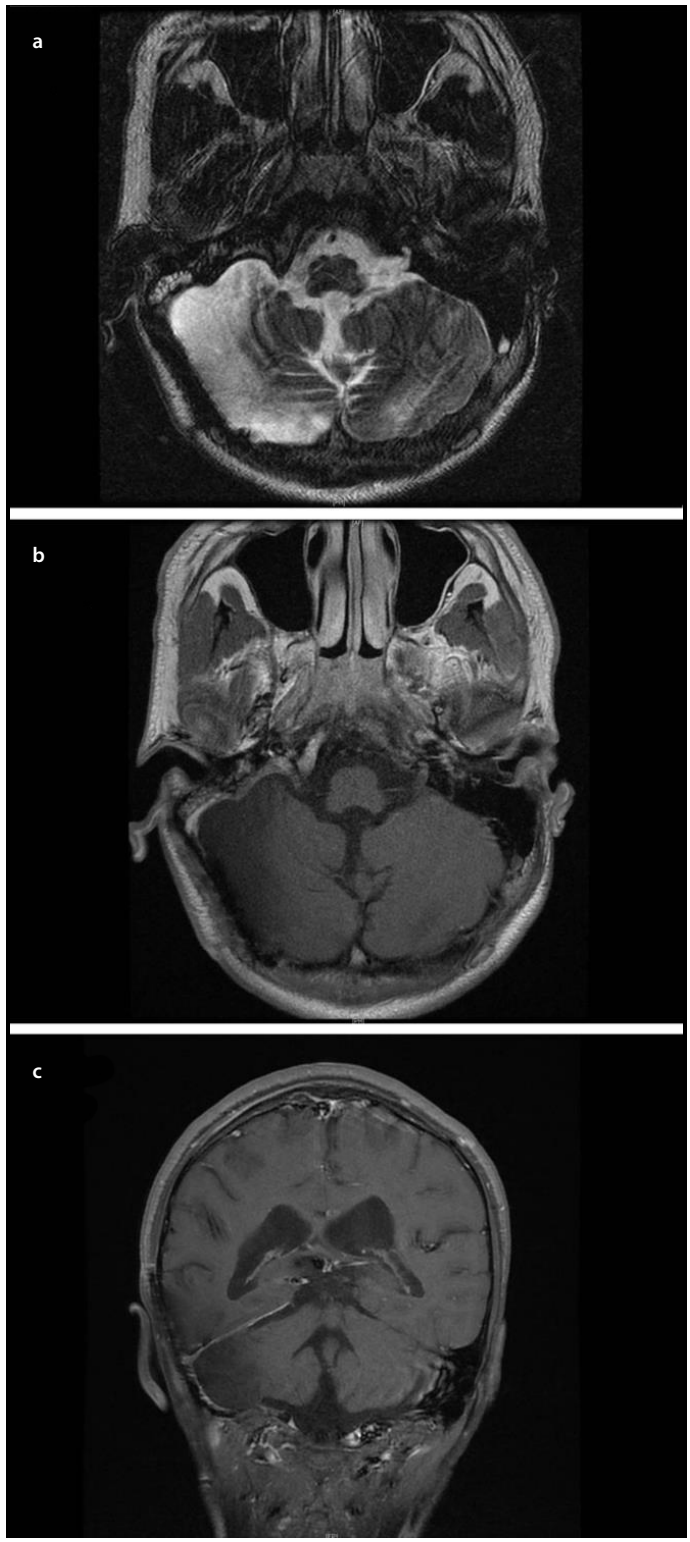

Figure 2. a-c. Two-year follow-up MRI without any recurrent disease. (a) Axial T2W; (b) axial contrast-enhanced T1; (c) coronal contrast-enhanced T1W.

no involvement of the otic capsule, which was left intact. Macroscopic clearance of the tumor was achieved. The patient's hospital stay was 12 days. No intra or perioperative complications were observed.

Macroscopically, the biopsy sample comprised gelatinous tissue. Microscopically, a neoplasm was identified with a multinodular architecture, comprising a small uniform ovoid to spindled cells with scanty eosinophilic finely vacuolated cytoplasm and small regular hyperchromatic nuclei (Figure 2). One mitosis was identified in 10 standard high-power fields (objective ×40 ~/1.6mm^2). The matrix was myxoid, and the cells formed cords or strands. Small foci of necrosis were identified. There was focal chondroid differentiation. Meningothelial cells, Physaliferous cells, rhabdoid cells, duct differentiation, and anaplastic/dedifferentiation were absent. The absence of nuclear pleomorphism favored a benign process. The immunohistochemistry is demonstrated in Table 1.

Chordoma was excluded due to negative brachyury immunohistochemistry. A non-midline myxoid chondrosarcoma of extraskeletal soft tissue or bone origin was not, and further opinions and molecular analyses were sought.

Additional FISH studies showed no evidence of gene rearrangement for EWSR1, NR4A3, EWS, FUS, SMARCB1, and PHF-1. These features were consistent with a primary intracranial myoepithelial lesion, i.e., parachordoma.

As a result of total tumor excision and the final histology and multidisciplinary team (MDT) discussion, no adjuvant therapy was given.

At a 4-month follow-up, the patient was recovering well, with examination showing a partial left IV nerve palsy, minor hearing impairment, and some unsteadiness. At a 2-year follow-up, the MRI showed no residual or recurrent tumor or metastases (Figure 3).

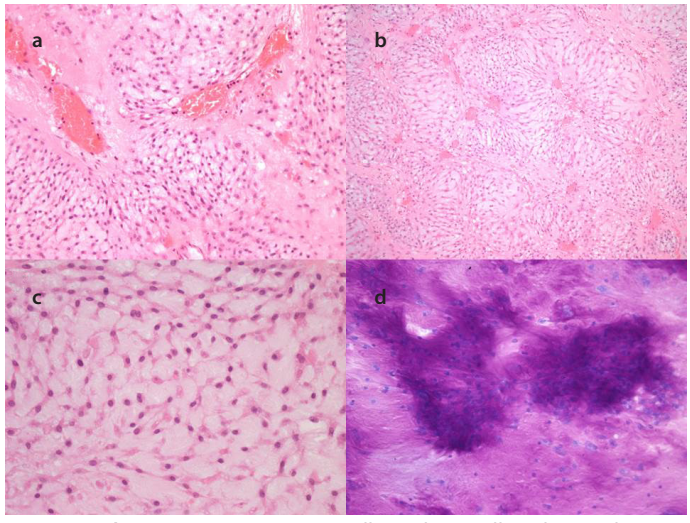

Figure 3. a-d. (a) Smear preparation: small neoplastic cells with metachromatic mucin Toluidine Blue objective ×20 bar=100µm; (b) Lobulated neoplasm H&E objective ×10 bar=200µm; (c) Lobulated neoplasm H&E objective ×20 bar=100µm; (d) Plump and spindled cells with regular oval nuclei, eosinophilic cytoplasm, fine vacuolation, and abundant extracellular clear mucin H&E objective ×40 bar=50µm.

DISCUSSION

Parachordoma has been described under other names, leading to confusion, all of which imply a similarity to chordoma. It is often misdiagnosed or used as a synonym for other soft tissue tumors, such as chondrosarcoma, extra-axial chordoma (EAC), or even low-grade sarcoma [3, 4]. Some authors have described true brachyury positive peripheral chordomas that are separate from parachordoma/myoepithelioma. This is significant because EAC confirmed with brachyury immunoreactivity has a tendency to grow and recur with local bone destruction, whereas parachordoma is considered less aggressive with better outcomes [2, 5]. However, it has recently been reported that the histology, immunochemistry, and cytogenic profiles of parachordoma are significantly characteristic enough to allow differentiation from chordoma, extraskeletal myxoid chondrosarcoma, and probably even soft tissue myoepithelioma [3, 5, 6,].

Although imaging is important to evaluate the anatomy of the tumor, diagnostic characteristics of parachordoma are largely nonspecific. Based solely on findings obtained by imaging, diagnosis will likely be difficult, as demonstrated by our case. Final diagnosis will therefore be determined by pathological findings [6].

The World Health Organization classification of tumors of soft tissue and bone Fletcher (2013) includes parachordoma in the chapter on myoepithelioma/myoepithelial carcinoma/mixed tumor of soft tissue. The label of parachordoma is applied when the vacuolation of cells is identified [7]. Myoepitheliomas are essentially benign but should be excised with a good margin, as recurrence is reported in approximately 20% of cases [4,14]. Adjuvant therapy is not indicated, except in those cases with metastasis [9]. Myoepithelial carcinomas are very rare but are indicated by nuclear atypia, high mitotic rate, and extensive necrosis: They predominate in the pediatric population. Myoepithelial carcinomas recur and metastasize in about half of these histologically aggressive cases [7].

The most common sites of origin of parachordomas are the lower limb, followed by the upper limb and then abdomen. Head and neck parachordomas are much rarer, with only three cases reported in the literature: intracranial, in the parietal skull, and in the nares. Of these three, none reported metastases or recurrence. However, the follow-up data from these cases were limited to 1 year, 15 months, and 4 months, respectively [4, 6, 10, 11, 14].

The literature contains cases of parachordoma that have metastasized, but despite the eight cases reported to date, including at least five fatalities, it is still considered rare in parachordoma. Of the 14 cases of recurrence, the recurrence rate ranged from 1 month to 12 years, with an average time after resection of 3.8 years for the parachordoma to recur [5, 6, 10-14].

CONCLUSION

Parachordoma/myoepithelioma is a rare neoplasm resembling chordoma, but differing clinically, morphologically, and immunohistochemically. This is important because outcomes and management are different. Local excision is adequate for parachordoma. Nevertheless, fatalities have been reported due to rare cases of metastases. A complete resection with clear margins is essential. Late recurrence is possible, so patients must be followed up with imaging at regular intervals.

Informed Consent: Informed consent was obtained from the patient who participated in this study.

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