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Case Report

A rare case of chondroblastoma of the jugular foramen: Imaging and pathological features ☆

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ABSTRACT

Chondroblastoma is a rare benign tumor that typically arises in the epiphyses or apophyses of long bones in patients with immature skeletal structures. Occasionally, it presents in atypical sites such as the bones of the foot and craniofacial region. This report details the case of an 18-year-old female who experienced neck pain and was found to have a lesion in the left jugular foramen. Imaging revealed typical lytic and expansive bone destruction with invasion into the adjacent internal jugular vein. Initially misdiagnosed as a glomus jugulare tumor, definitive diagnosis of chondroblastoma was established postoperatively via histopathology. This case underscores the diagnostic challenges posed by chondroblastomas at atypical sites. Through detailed imaging, histopathological analysis, and literature review, this study aims to enhance the awareness of clinicians and radiologists regarding chondroblastoma and provides insights and references for the diagnosis and management of similar cases in the future.

Introduction

Chondroblastoma was originally reported by Kolodny [1] in 1927, subsequently described by Codman [2] in 1931, and recognized as a distinct entity by Jaffe and Lichtenstein in 1942 as chondroblastoma [3]. This cartilage-derived tumor accounts for approximately 1% [4] of all bone tumors and can occur at any age, though it is most prevalent among children and young adults aged 10-25 years, primarily presenting with pain. Research indicates that while chondroblastoma typically develops in the epiphyses of long bones, such as the femur, tibia, and humerus [5], it can also manifest in atypical locations with secondary ossification centers, including the bones of the foot [6], hand phalanges [7], scapulae [8], and craniofacial bones [9]. These atypical occurrences are attributed to the tumor's association with secondary ossification centers. Although chondroblastoma can often be suspected preoperatively based on

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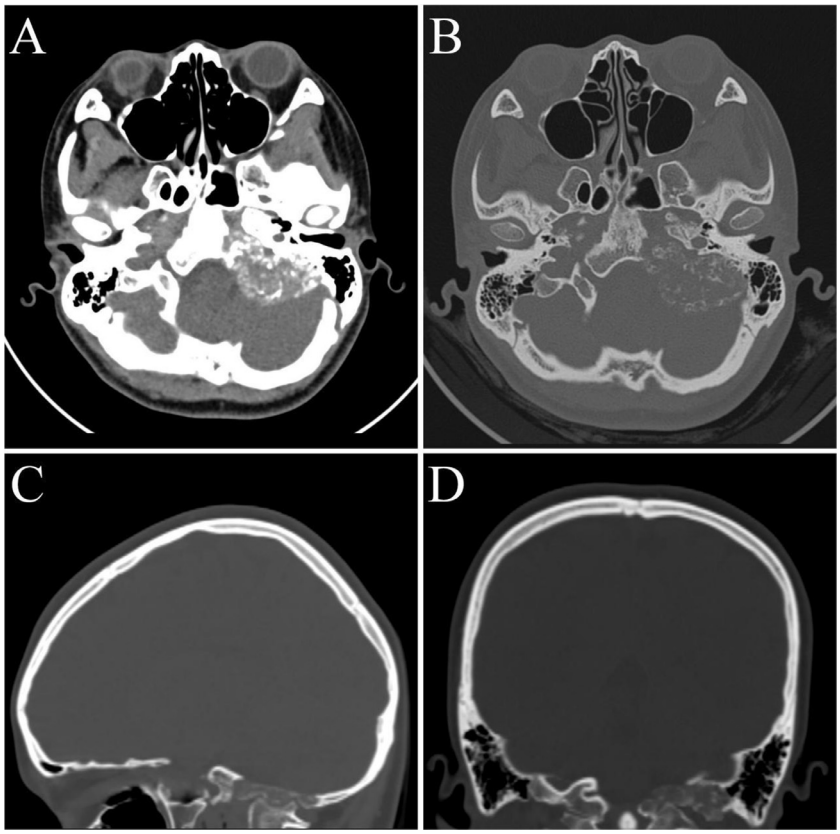

Fig. 1 – CT findings. (A-D) Three-dimensional CT images reveal multiple bone disruptions at the base of the left posterior cranial fossa, including the clivus, petrous apex, vertebrae, temporal bone, and hypoglossal canal. These areas show varying degrees of involvement, uneven density, unclear borders, and measure approximately 3.2 × 4.4 cm.

age, affected sites, and imaging characteristics, diagnosing the tumor in uncommon sites remains challenging due to its rarity and the general unfamiliarity within the clinical and radiological communities. This paper reports a case of chondroblastoma initially misdiagnosed as a glomus jugulare tumor, aiming to elucidate the clinical, CT, and MR features, along with pathological findings, and to provide a comprehensive literature review to distinguish chondroblastoma from other lesions and enhance diagnostic precision.

Case report

This case involves an 18-year-old female who presented with intermittent neck pain lasting 6 months, which was relieved by rest. A physical examination revealed slight swelling on the left side of the neck, with increased pain and dizziness upon neck rotation. The patient was alert, with normal vision and hearing. Initial cranial and thoracic CT scans (Fig. 1) showed extensive osteolytic bone destruction around the base of the left posterior cranial fossa but no periosteal reaction or thoracic abnormalities. Subsequent cranial MRI (Fig. 2) identified extensive bone destruction and mass formation in the left jugular foramen area, featuring low T1WI intensity and mixed high and low T2WI intensity. The lesion involved the left clivus, left petrous apex, left vertebrae, left temporal bone (mastoid process), and left hypoglossal canal. T1WI enhancement scans revealed marked heterogeneous enhancement of the lesion and localized stenosis of the left jugular vein, with some areas of unclear imaging. The adjacent left cerebellar hemisphere exhibited mild compressive changes, with no abnormal intensities in the cerebral parenchyma. Contrast-enhanced MR venography (CE-MRV) confirmed invasion of the left internal jugular vein (Fig. 3). After a multidisciplinary consultation based on the clinical and imaging findings, the diagnosis was assumed to be a glomus jugulare tumor, and surgical intervention was recommended. However, the patient and her family declined surgery due to the high associated risks and opted to be discharged from the hospital.

Three months later, the patient returned to our hospital due to aggravated and intolerable neck pain, reporting that neither rest nor oral administration of ibuprofen provided relief. After MRI examinations again (Fig. 4), we found no clear tumor progression, and the imaging findings were similar to those at the initial consultation. The patient underwent tracheal intubation under general anesthesia and proceeded with “left jugular foramen tumor resection + cerebrospinal fluid leakage repair through a left postauricular C-type incision”. During surgery, partial bone destruction was noted in the left mastoid root and occipital condyle. The mass was soft and locally calcified with a rich blood supply, invading the left

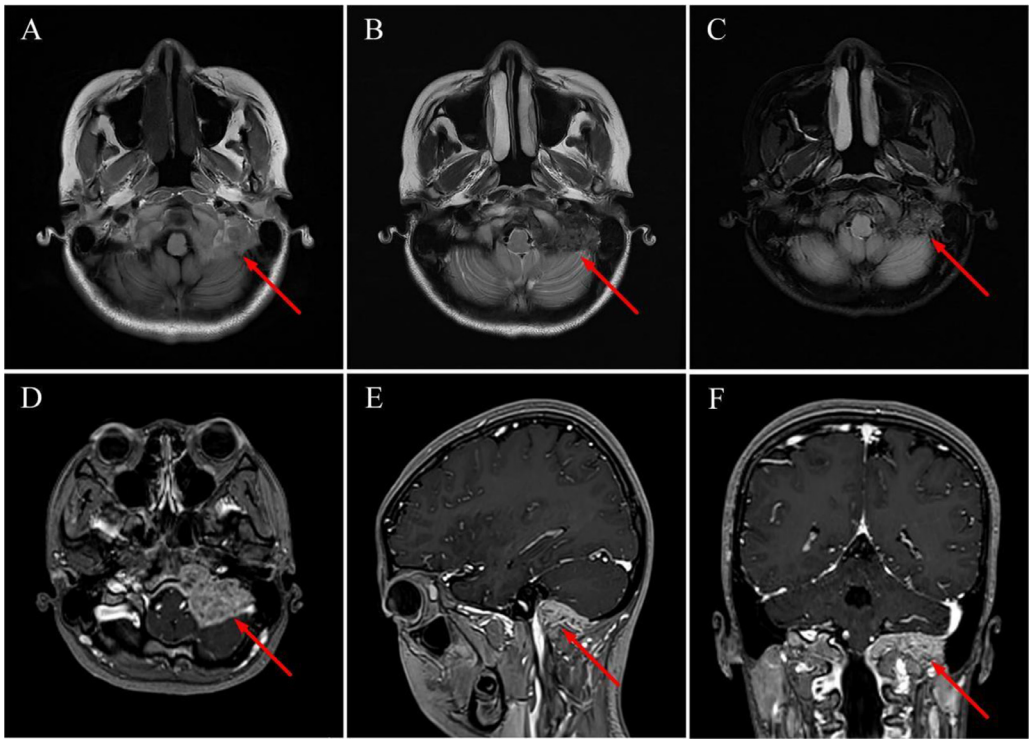

Fig. 2 – MRI findings. T1WI and T2WI depict an irregular mass in the left jugular foramen region with adjacent bone intensity alterations. The lesion exhibits low and slightly low intensities, and there is compression of the adjacent left cerebellar hemisphere (A, B). T2 fat suppression sequences reveal low intensity within the lesion with hyperintense spots (C); T1WI enhancement scans demonstrate significant heterogeneous enhancement of the lesion, which encircles and invades the left internal jugular vein, accompanied by multiple bone destructions. Importantly, the lesion does not invade the adjacent brain parenchyma (D-F).

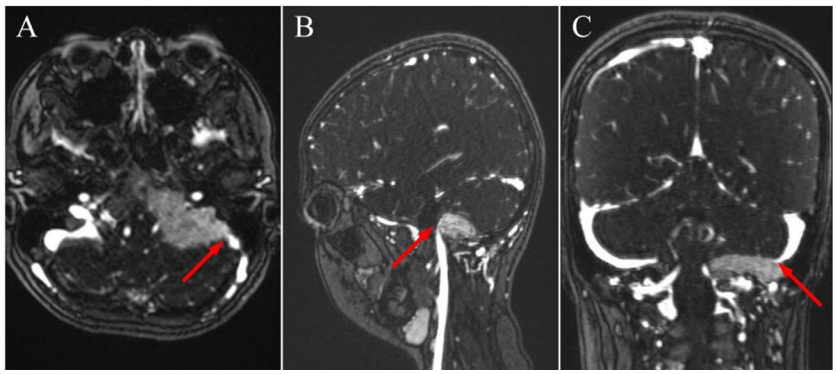

Fig. 3 – CE-MRV findings. (A-C) Display the mass occupying the left jugular foramen and invading the left internal jugular vein in axial, sagittal, and coronal views, respectively, causing narrowing and alteration of its course.

internal jugular vein and extending to the posterior pharyngeal wall along the lateral wall of the external auditory canal. The lesion was excised as completely as possible. Pathological examination confirmed the tumor as chondroblastoma, microscopically characterized by chondroblasts and osteoblasts in a cartilage-like stroma (Fig. 5). The chondroblasts were medium-sized, round or oval, with abundant reddish cytoplasm and large, deeply stained nuclei centrally located, generally rounded with an inconspicuous nucleolus and no visible nuclear division. The osteoblasts were irregularly distributed with characteristic calcium salt deposits around the tumor cells, forming chicken-wire calcifications. Immunohistochemistry showed P63(+), SATB2(-), Ki-67(3%+), CD68(osteoblast-like giant cells+), H3. 3G34W(-), CD163(monocyte-like cells+), S-100(+). The patient recovered well postoperatively, with no evidence of recurrence or metastasis on follow-up imaging.

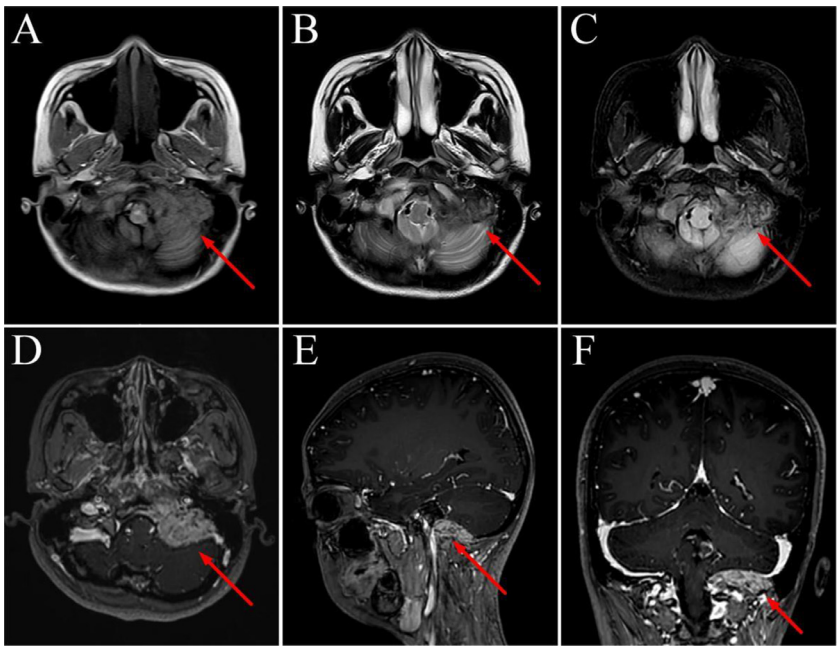

Fig. 4 – Preoperative MRI findings from 1 week prior. T1WI, T2WI and T2 fat suppression sequences (A-C); T1WI enhancement (D-F).

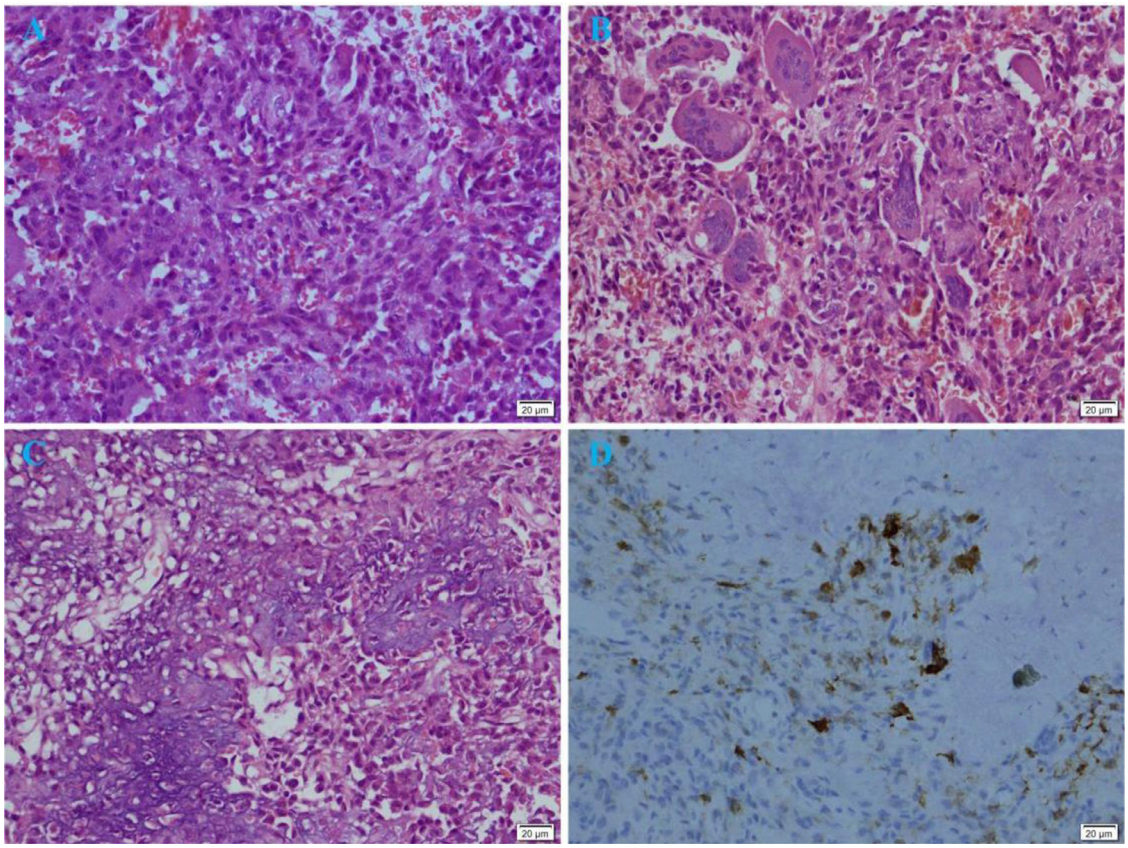

Fig. 5 – Pathologic findings (X400). (A) Numerous chondroblasts arranged in a paving-stone pattern, displaying weakly eosinophilic cytoplasm and longitudinal nuclear grooves. (B) Irregularly distributed multinucleated giant cells and chondrocyte-like stroma. (C) Characteristic chicken wire-like calcification around the tumor cells. (D) Tumor cells are S-100 positive.

Discussion

Overview

Chondroblastoma is a rare benign cartilage-derived tumor, primarily arising in the epiphyses of long bones in young patients, with a slightly higher incidence in males than females [4]. It infrequently occurs in atypical sites such as the hands, feet, and skull, and is even rarer in the jugular venous foramen. A multicenter analysis of 199 patients with chondroblastoma found that pain was the most common symptom, followed by joint effusion and limited movement [10]. Due to its complex anatomical location and similar imaging characteristics to other diseases, particularly tumors in the jugular foramen area [11] (e.g., glomus jugulare tumor), diagnosing chondroblastoma in the jugular foramen is especially challenging. Accurate diagnosis requires a thorough combination of clinical evaluation, imaging, and pathological analysis.

Imaging features

Chondroblastoma exhibits several distinctive features on X-ray examinations: The first is the site; the tumor typically originates in the epiphyses, appearing ovoid or round, often less than 5 cm in diameter, and may display cystic features. The second is boundaries; the tumor typically has well-defined borders with sclerotic margins and may exhibit expansile changes, as seen in this case. The third is calcification [12]; CT may be necessary to detect calcification foci amid varying calcification degrees. The fourth is periosteal reaction; chondroblastomas usually show no periosteal reaction, being confined within a sclerotic bone shell. However, periosteal reaction can occur in the adjacent diaphysis or epiphysis if the tumor extends and causes bone destruction. The typical CT appearance mirrors that of X-rays, showing clear boundary osteolytic bone destruction with circular and curvilinear calcifications within the lesion [13]. MRI reveals that specific chondroblastoma intensities are absent, with T1WI and T2WI intensities varying depending on the lesion's components such as cartilaginous matrix, cellular structures, calcifications, hemosiderin, and aneurysmal bone cyst-like areas [4]. Calcifications appear with low intensity on both T1WI and T2WI, accompanied by inflammatory changes like bone marrow edema, soft tissue edema, and synovitis [14].

Pathological manifestations

Typical microscopic tumor tissues consist of chondroblasts, multinucleated giant cells, and chondrocyte-like stroma [4]. Chondroblasts, as the primary component, features cells densely arranged in a paving-stone pattern, with medium-sized round or oval shapes, clear or mildly eosinophilic cytoplasm, distinct cell boundaries, a larger quasi-circular nucleus, an inconspicuous nucleolus, and longitudinal nuclear grooves. Multinucleated giant cells are irregularly distributed without specificity. Calcification, often associated with the tumor, displays a chicken wire-like pattern almost exclusive to chondroblastoma and holds diagnostic significance [15]. Immunohistochemistry typically shows S-100 positivity in most chondroblastoma patients, indicating chondrogenic differentiation, and CD68 expression in multinucleated giant cells, serving as an adjunctive marker for diagnosis.

Differential diagnosis

Chondroblastoma in atypical sites, such as the skull base and jugular foramen, should be differentiated from several conditions: (1) Glomus jugulare tumor: Appears as a soft, movable pulsatile mass by CT imaging, typically showing circular or quasi-circular low-density areas in the jugular foramen with rare calcification. MRI reveals low T1 and high T2 intensity, characterized by vascular enhancement on enhancement scans. CE-MRA often shows compression or stenosis of the internal jugular vein caused by the lesion. In contrast, chondroblastoma presents as expansive and osteolytic lesions with well-defined borders and typical chicken wire-like calcification, lacking vascular enhancement features. (2) Aneurysmal bone cyst: Approximately 30% of chondroblastomas are secondary to aneurysmal bone cysts. These typically present with a "soap bubble" or multilocular appearance on MRI. MRI with contrast typically demonstrates internal septations that may contain characteristic fluid-fluid levels, signifying layering of solid blood components within cystic areas of the lesion [16]. (3) This tumor shares similar imaging features with chondroblastoma and presents a variable number of osteoblast-like giant cells microscopically [17]. However, osteoblastoma usually occurs in individuals over 20 years old, displays no sclerotic margins on x-ray and CT, lacks a cartilage matrix or calcification among tumor cells, and is S-100 negative on immunohistochemical staining. (4) Chondromyxoid fibroma: A rare chondrogenic benign tumor that rarely involves the epiphysis and exhibits prominent lobular growth, minimal calcification, and microscopically small lobules of varying sizes with sparse cells in the centers and predominantly mucinous stroma [18], contrasting with the typical distribution of chondroblasts in a paving-stone pattern without significant mucoid degeneration. (5) Clear cell chondrosarcoma: Difficult to distinguish from chondroblastoma by imaging alone as it can also occur in the epiphysis, presenting well-defined expansive osteolytic lesions 18. Histologically, it is differentiated by larger tumor cells, broad and clear cytoplasm, significant cytoplasmic glycogen content by PAS staining, and prevalence in adults over 30 years of age.

Treatment

Although chondroblastoma is histologically benign, it can develop local recurrence and distant metastasis, with lung metastasis [19] being the most common. Surgical curettage combined with bone grafting is currently considered an effective treatment for chondroblastoma, and complete removal of the lesion is the only reliable method to prevent recurrence. However, completely removing chondroblastomas located in the jugular foramen is challenging due to the complex anatomy of the site, posing a significant risk of recurrence. Therefore, it is essential for patients to undergo long-term postoperative follow-up and imaging examina-

tions, even though no signs of recurrence or metastasis have been detected postoperatively.

Conclusion

In conclusion, diagnosing chondroblastoma in atypical locations, such as the jugular foramen, presents significant challenges. A comprehensive approach involving clinical evaluation, imaging, and pathology is essential for accurate diagnosis, and this case underscores the need for awareness among clinicians and radiologists.

Patient consent

Written informed consent was obtained from a legally authorized representative for anonymized patient information to be published in this article.

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