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Aggressive Giant Cell Granuloma of the Ethmoid: Diagnostic Challenges and Management Strategies

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1. Abstract

Giant Cell Granuloma (GCG) is a rare, benign but locally aggressive tumor-like lesion that predominantly affects the maxilla and mandible. Its occurrence in the ethmoid bone is extremely rare, with only a few cases documented in the literature. Due to its unusual location, ethmoidal GCG can pose diagnostic challenges and may mimic other osteolytic lesions.

We report the case of a 20-year-old Moroccan woman with no prior medical history who presented with a progressive, painless swelling in the right internal canthal region. Imaging studies, including Computed Tomography (CT) and Magnetic Resonance Imaging (MRI), revealed a well-defined osteolytic lesion affecting the ethmoid bone. A differential diagnosis included other giant cell-rich lesions such as aneurysmal bone cyst, brown tumor of hyperparathyroidism, and osteoclastoma. Histopathological examination confirmed the diagnosis of aggressive giant cell granuloma. The patient underwent complete surgical excision *via*an endoscopic endonasal approach. The postoperative course was uneventful, and no recurrence was observed after six months of follow-up.

Ethmoidal giant cell granuloma is a rare entity that should be considered in the differential diagnosis of osteolytic lesions in the paranasal sinuses. Early diagnosis and complete surgical excision are essential to prevent recurrence and ensure favorable outcomes. This case highlights the importance of histopathological confirmation and appropriate surgical management in rare presentations of GCG.

2. Keywords: Giant cell granuloma; Ethmoid bone; Aggressive lesion; Endoscopic surgery; Case report

3. Introduction

Giant cell granuloma (GCG) is a rare, benign but potentially aggressive fibro-osseous lesion characterized by the proliferation of multinucleated giant cells within a vascularized fibrous stroma. Histologically, it is marked by the presence of hemorrhagic foci, hemosiderin deposits, and areas of osseous metaplasia, which can sometimes lead to diagnostic confusion with other giant cell-rich lesions, such as aneurysmal bone cysts, brown tumors of hyperparathyroidism, and osteoclastomas.

GCG is most commonly observed in the mandible and maxilla, primarily affecting young adults, with a higher prevalence in women. Although the exact etiology remains unclear, some studies suggest that trauma, inflammation, or hormonal influences may contribute to its pathogenesis. Despite its relatively indolent nature, some cases exhibit aggressive behavior, characterized by rapid growth, cortical bone destruction, and a tendency for recurrence after surgical resection.

While GCG is well-documented in the gnathic bones, its occurrence in the craniofacial skeleton, particularly in the ethmoid bone, is exceedingly rare, with only a few cases reported in the literature. The involvement of the ethmoid poses unique diagnostic and therapeutic challenges due to its proximity to vital structures such as the orbit, skull base, and paranasal sinuses. Clinical presentation often depends on the extent of the lesion, with symptoms ranging from facial swelling and nasal obstruction to orbital involvement in advanced cases.

Herein, we present the case of a 20-year-old woman diagnosed with an extensive and aggressive ethmoidal giant cell granuloma. We describe the clinical presentation, imaging characteristics, histopathological findings, and the surgical approach employed for its management. This report aims to contribute to the limited body of literature on ethmoidal GCG and emphasize the importance of early recognition and appropriate treatment strategies for this rare entity.

4. Case Presentation

A 20-year-old woman presented to our department with a progressively enlarging right internal canthal swelling that had been evolving for nine months. The swelling was associated with intermittent right nasal obstruction and later complicated by chronic epiphora, progressive visual impairment, and cutaneous fistulization.

Physical examination revealed a firm, fixed, reddish, painless mass extending to the infraorbital region, displacing the right globe downward and outward, causing proptosis (Figure 1). Anterior rhinoscopy showed a fleshy mass filling the right middle meatus, appearing to be continuous with the middle turbinate, adherent to the inferior turbinate and nasal septum, with associated synechiae. A deviated nasal septum was also noted.

Orbital and cerebral CT revealed a heterogeneously enhancing soft tissue mass centered in the right orbit-ethmoidal region, measuring 45 × 49 × 33 mm. The lesion demonstrated osseous involvement of the medial orbital wall, intraorbital extension with displacement of the medial rectus muscle and globe without intraocular invasion, and mild proptosis. Additionally, the mass extended into the nasal cavity, displacing the nasal septum contralaterally and obstructing the frontonasal duct and maxillary sinus ostium. Superior-internal bone erosion was also noted (Figure 2 and 2A).

Microscopic analysis of the biopsy specimen showed multinucleated giant cells scattered within a fibrous stroma, with hemorrhagic and osteoid metaplastic foci. Immunohistochemical analysis demonstrated CD68 and CD163 expression in mononuclear cells, confirming the diagnosis of giant cell granuloma.

The patient underwent tumor resection via a combined approach: external para-canthal approach and endonasal endoscopic approach, including partial middle turbinectomy, Draf IIa frontal sinusotomy, and anterior and posterior ethmoidectomy. Reconstruction was performed using abdominal fat and Surgicel.

The postoperative period was uneventful, with no complications or recurrence observed at six months (Figure 3).

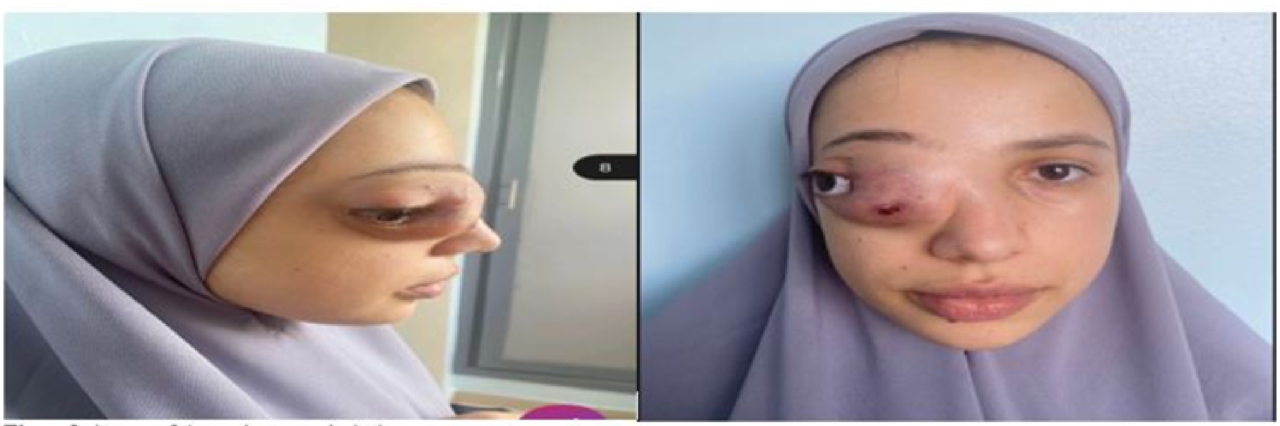

Figure 1:Image of the patient on admission.

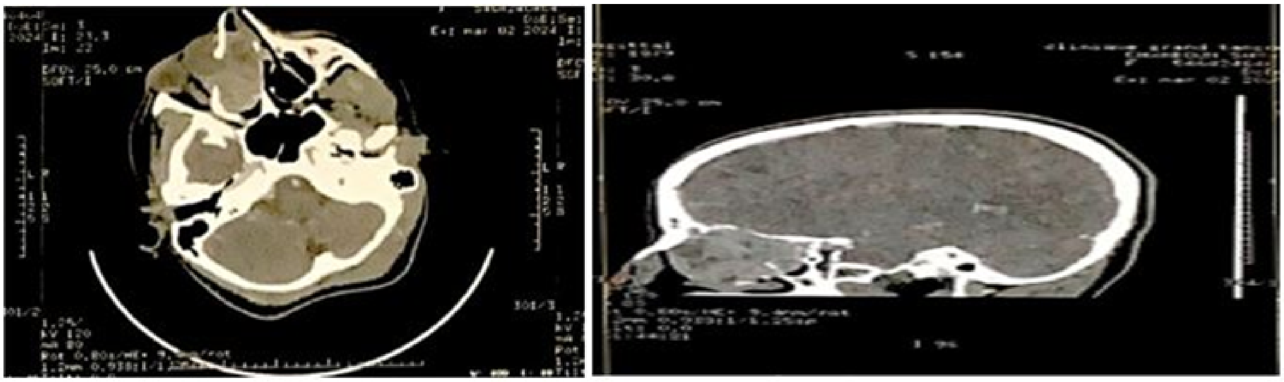

Figure 2: CT image showing an axial and sagittal section through the tumor process.

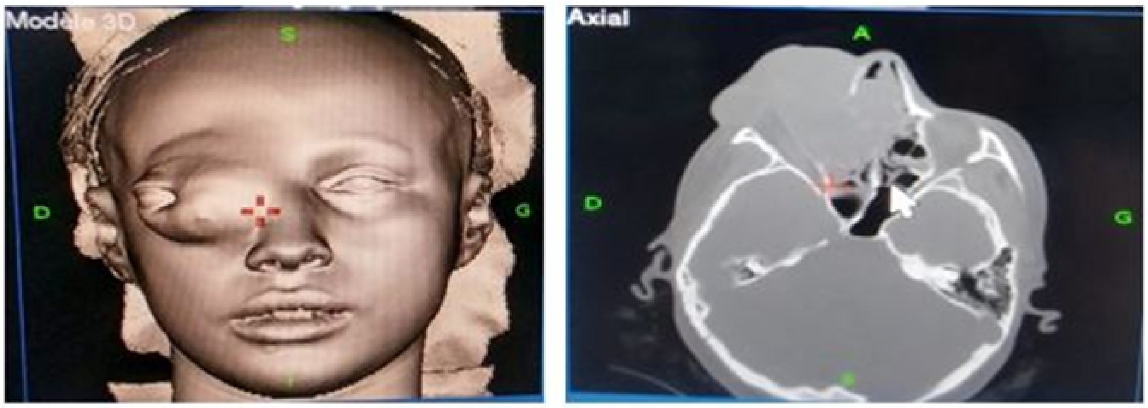

Figure2A: Image taken from the neuronavigation program showing the tumor process in axillary section and in 3D.

Figure 3: Image of the patient in the immediate post-operative period on the right and after 6 months on the left.

5. Discussion

GCG accounts for approximately 7% of benign jawbone lesions and predominantly affects young adults, with a female- to-male ratio of 1.5:1 [3,5]. It is classified into two subtypes:Non-aggressive GCG: Slow-growing, asymptomatic, well- circumscribed. Aggressive GCG: Rapid growth, pain, significant osseous destruction, and high recurrence rates (10% to 15%).Criteria for aggressive GCG include rapid growth, lesion size >5 cm, root resorption, and extensive bone destruction[6].

Historically, GCG has been considered a reactive lesion arising from an exaggerated response to local trauma, intraosseous hemorrhage, or chronic inflammation. However, some aggressive cases exhibit features suggestive of a neoplastic process, including somatic mutations (e.g., H3F3A mutations in giant cell tumors) [6].

GCG is characterized by: Multinucleated giant cells resembling osteoclasts, responsible for bone resorption.Fibroblastic and histiocytic stroma with vascular proliferation,CD68 and CD163 positivity on immunohistochemistry,frequent hemorrhagic foci and osseous metaplasia, contributing to imaging heterogeneity.

The clinical presentation depends on the lesions location and extent:

Orbital/nasal mass causing facial deformity,Progressive nasal obstruction if extending into the nasal cavity,chronic epiphora due to nasolacrimal duct compression,proptosis and diplopia with orbital involvement,pain though often absent in early stages.

CT Findings: Well-demarcated or ill-defined lytic lesion, expansile with bone erosion and heterogeneous contrast enhancement[6-8].

MRI Findings: T1 and T2 hypointensity, with heterogeneous T2 hyperintense areas.Variable gadolinium enhancement.Signal heterogeneity due to hemorrhagic changes.In our case, CT revealed an expansive 45 × 49 × 33 mm orbit- ethmoidal lesion with intraorbital extension and medial wall erosion, leading to globe displacement and proptosis[9,10].

Complete excision is the primary treatment, with: Endoscopic endonasal approach (preferred) [9]. External approaches (transfacial, para-canthal, craniotomy) for extensive orbital involvement.

In our case, a combined para-canthal and endoscopic approach was employed, followed by defect reconstruction with abdominal fat and Surgicel.

6. Conclusion

Ethmoidal giant cell granuloma is exceptionally rare. Diagnosis requires a comprehensive clinical, radiological, and histopathological evaluation. Surgical resection, preferably via an endoscopic approach, remains the mainstay of treatment. Long-term follow-up is essential due to the risk of recurrence.

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