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Case Report

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Giant Cell Tumor of the Inferior Turbinate in a 12-Year-Old Child: First Case Report

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Giant cell tumors (GCT) are highly rare neoplasias, accounting for less than 5% of all soft tissue tumors. Only 2% of GCTs occur in the head and neck region and they predominately affect the maxillary and sphenoid sinus.[1] To date, only 3 cases of nasal cavity GCTs were reported in literature, all in female adults.[2-4] GCTs show a locally expansive growth pattern and rarely metastasize. Due to its rarity, no standardized diagnostic and therapeutic recommendations are available so clinical management is highly individual depending on tumor localization, tumor size, the presence of distant or local metastases, and patient age.

Description

A 12-year-old girl was referred to our clinic with a 3-month history of right-sided nasal obstruction and nasal discharge as well as recurrent right-sided epistaxis. She did not report a loss of weight, fever, visual impairment, or pain. A treatment with decongestant nasal spray and oral amoxicillin for 7 days initiated by her family doctor could not relieve her symptoms. Nasal endoscopy showed a subtotal obstruction of the posterior part of the right nasal cavity with an endonasal mass originating from the inferior turbinate and covered by intact mucosa. Next, magnetic resonance imaging (MRI) with contrast and (**Figure 1A** and **B**) computed tomography imaging (**Figure 1C** and **D**) were initiated and showed an oval-shaped tumor with partial destruction of the inferior turbinate, nasal septum, and hard palate. The tumor showed an extension of $26 \times 21 \times 16$ mm and a moderate peripheral contrast enhancement. Peripheral blood laboratory analyses showed no suspicious findings. A biopsy was taken from the endonasal mass under general anesthesia where the tumor presented as highly indurated expansion of the posterior part of the inferior turbinate. Histopathology showed a solid lesion composed of multinucleated giant cells and mononuclear cells, surrounded by lamellar bone and fibrotic interstitial tissue with increased vascularization and spindle cell-like fibroblasts (**Figure 2C**). The giant cells stained positive for CD68 and negative for p53, and the mononuclear cells showed no evidence of H3.3GD34 mutation. Finally, the diagnosis of a GCT was made and after a case discussion in our pediatric tumor board, navigation-guided endonasal-endoscopic tumor resection was planned.

The tumor was completely resected with partial resection of the nasal septum and the inferior turbinate using sharp instruments and a shaver with a safety margin of 10 mm (**Figure 2A**). To achieve an R0 situation, the bony resection borders at the nasal septum, inferior turbinate, and hard palate were thinned out with a diamond burr (**Figure 2B**). Intraoperative frozen-section histology revealed tumor-free resection margins. No postoperative complications were observed and the patient could leave the hospital after 48 hours. A control MRI 4 months after surgery showed no signs of tumor recurrence (**Figure 2D**).

Written informed consent for patient information and images was provided by the patient's legally authorized representatives. The scientific use of the patient's tissue and clinical data was approved by the Saarland Medicines' Association ethical review board.

Discussion

GCTs represent rare soft tissue tumors that mainly affect the epiphyseal regions of the long bones in young adults.[5] The nasal cavity is an extremely rare site of presentation

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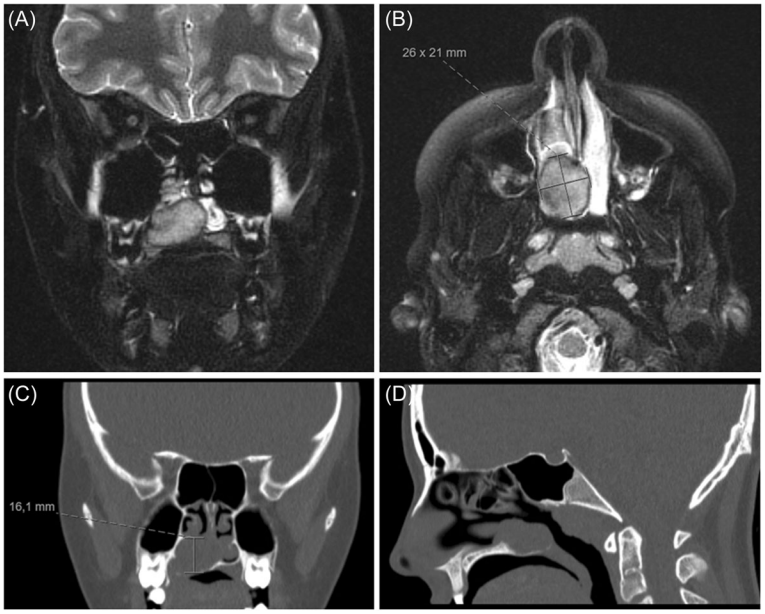

Figure 1. Preoperative MRI and CT imaging. (A) Coronar view of a T2 weighted MRI scan showing a solid tumor of the right-sided inferior turbinate with expansion to and thinning of the hard palate and expulsion of the nasal septum; (B) transversal view of a T2 weighted MRI scan showing a tumor extension of 26 × 21 mm; (C) coronar view of a CT scan with signs of locally expansive tumor growth; and (D) sagittal reconstruction of a CT scan showing a markedly thinning of the bone of the hard palate. CT, computed tomography; MRI, magnetic resonance imaging.

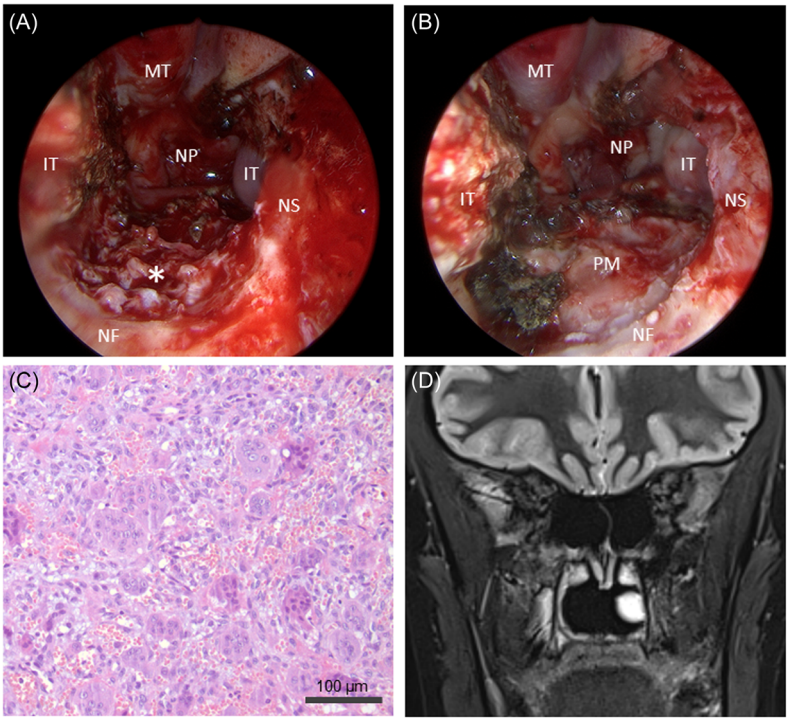

Figure 2. Transnasal endoscopic osteoclastoma resection and histopathological findings. (A) Surgical field after partial resection of the left-sided IT and the posterior part of the NS with tumor remnants at the nasal floor (*). (B) Completed tumor resection with an exposition of the PM after thinning of the hard palate with a diamond burr. (C) Histopathology shows multinucleated giant cells surrounded by a dense vascular network and mononucleated cells (H&E staining). (D) MRI scan 4 months postsurgery. H&E, hematoxylin and eosin; IT, inferior turbinate; MRI, magnetic resonance imaging; MT, middle turbinate; NF, healthy bone of the nasal floor anterior to the tumor site; NP, nasopharynx; NS, nasal septum; PM, palatine mucosa.

with only 3 cases reported in literature. Here, we present the first case of a GCT of nasal inferior turbinate. In the majority of reported head and neck cases, complete surgical resection was feasible and to date represents the therapeutic gold standard. In cases of incomplete resection or in patients where complete resection is not achievable, radiotherapy may be considered. Anti-RANKL antibody denosumab is a therapeutic option for metastasized and unresectable tumors as well as for neoadjuvant treatment of patients for whom surgery may result in significant morbidity.[5]

As GCT is a highly rare diagnosis in children especially in the head and neck region several differential diagnoses have to be considered including among others juvenile angiofibroma, giant cell granuloma, and fibrous dysplasia. Due to the rarity of the disease, no evidence-based therapeutic guidelines are available so disease management is highly individual and requires an interdisciplinary team. As up to 50% of patients develop locoregional tumor recurrence,[1] a long-term follow-up including MRI controls is mandatory.

Conclusion

Our case illustrates the case of a highly rare soft tissue tumor in children with a unique localization at the inferior turbinate that should be considered as a differential diagnosis in young patients with unilateral nasal obstruction and recurrent epistaxis.

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Author Contributions

Maximilian Linxweiler, Institutional Review Board (IRB) approval, surgery, drafting the article, revisions; **Marc Remke**, clinical management, review of the manuscript; **Beate K. Straub**, histopathological analyses, review of the manuscript; **Bernhard Schick**, IRB approval, surgery, review of the manuscript.

Disclosures

Competing interests: The authors declare that there is no conflict of interest.

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