Osteochondromyxoma of the nasal cavity: case report and review of the literature

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European Journal of Medical Case Reports
Volume 2(1):12-15

https://doi.org/10.24911/ejmcr/1/37
Discover Publishing Group

ABSTRACT

Background: Osteochondromyxoma is an extremely rare benign tumor. Few cases have been described in the literature. Patients with Carney complex may present with this tumor.

Case Presentation: We report a case of a nasal osteochondromyxoma in a 21-year-old female patient who presented with progressive nasal congestion, anosmia, and episodes of self-limiting epistaxis. Magnetic resonance imaging and computerized axial tomography scans were performed which demonstrated a large tumor occupying the nasal cavity. Endoscopic sinus surgery was performed resulting in complete resection of the tumor. No recurrence has been observed at 24 months following endoscopic sinus surgery. The patient has also recovered olfaction.

Conclusion: Although osteochondromyxomas are extremely rare, it should be considered as a differential diagnosis for nasal tumors and surgical resection should be completed. A multidisciplinary assessment is recommended as it is a major criterion for the diagnosis of Carney complex.

Keywords: Osteochondromyxoma, Carney's complex, nasal tumor, paranasal sinuses, case report.

Received: 26 Oct 2017 **Accepted:** 26 Nov 2017

Type of Article: CASE REPORT

Funding: None

Declaration of conflicting interests: None

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Background

Osteochondromyxoma is an extremely rare bone tumor. It appears to affect paranasal sinuses and long bones [1]. The World Health Organization has included this tumor in the benign chondrogenic bone tumor category with code 9211/0 [2,3]. To the extent of our knowledge, sporadic cases have nonetheless been described since 1955 [4].

In 1985, Carney et al. [5] described patients presenting with myxomatous masses, spotty pigmented lesions of the skin, and endocrine disorders, which would later be known as Carney complex. Major criteria for diagnosing this complex include myxomas (cardiac, mucocutaneous, breast), skin lesions (spotty pigmentation, blue nevus, epitheloid blue nevus), altered endocrine activity (pigmented nodular adrenocortical disease, acromegaly), large-cell calcifying Sertoli cell tumor, thyroid carcinoma (or multiple hypoechoic nodules), psammomatous melanotic schwannoma, breast ductal adenomas, and osteochondromyxoma. The patient must present with two of the criteria to be diagnosed with the complex [6]. It is believed that mutations in the PRKAR1A gene are the source of the complex, and demonstration of such or an affected first-degree relative, are also considered as a diagnostic criterion [7].

In 2015, 700 cases of Carney complex had been described worldwide and approximately 2% presented with an osteochondromyxoma [6,7]. However, a case of osteochondromyxoma of the maxilla without Carney complex has recently been described [8]. Cases found in the literature prior to 1985 exist, yet precision of concomitant diagnostic criteria that would correspond to Carney complex is lacking.

We hereby describe a case of osteochondromyxoma of the nasal region which appears to be unrelated to Carney complex. Informed consent to publish and present this case was obtained from the patient.

Because of the rare nature of this tumor, an extensive review of the literature was performed, yielding 12 cases of osteochondromyxoma. The earliest publication found corresponds to a sacroiliac tumor [4]. Cases of skull base [9], spine [10], chest wall [11], rib [12], ethmoid, inferior turbinate, tibia, and radius [6] have been described. Most recently, a case of the anterior wall of the maxillary sinus has been reported [8].

Of these cases, a diagnosis of Carney complex was determined for six patients; however, the complex was described in 1985 and, therefore, no definite assumptions can be made as many of the cases are published prior to this date. Mountricha et al. [8] describe a case in 2009 that was not associated with Carney complex.

Case Presentation

A 21-year-old woman was seen at the otolaryngology clinic with an 8-month history of right nasal fossa congestion which became bilateral within 2 months. The patient referred episodes of unilateral self-limiting epistaxis

which occurred approximately once per month, anosmia and ageusia. She later developed hyponasal speech and persistent unilateral tearing of the right eye. She also described oropharyngeal dysphagia. Her past medical history was uneventful; she has six siblings, none of which are known to present any medical condition. The patient denied pain, asthenia, fever, or loss of appetite.

On physical examination, a large whitish tumor was observed fully obliterating the right nasal cavity (Figure 1). A deviated septum with convexity to the left was also observed, which was pronounced.

An MRI of the sinuses revealed a slightly heterogeneous and large expansive lesion with well-defined margins which occupied the nasal cavities and extended toward the ethmoidal cells. A CT scan of the sinuses showed an expansive lesion extending toward the ethmoidal cells and expanding the medial walls of the maxillary sinuses bilaterally (Figure 2). Thinning of peripheral bone as well as the absence of the bony nasal septum was observed. Slight convex bowing of the palate was also seen on the sagittal plane. No signs of metastasis were detected. Ophthalmological evaluation revealed conserved visual acuity, and neurosurgery evaluated the hypothalamic – pituitary – adrenal axis, which was unaffected.

An incisional biopsy was performed and showed a mesenchymal tumor with chondroid differentiation. The patient underwent an endoscopic sinus surgery with complete removal of the tumor. The posteroinferior portion of the nasal septum appeared to be the site of origin of the tumor. The surgery was well tolerated by the patient, with no surgical complications. The specimen resected had a gelatinous consistency and the histological sections for light microscopy showed polygonal and bipolar tumor cells with small nuclei and no obvious atypia. The tumor appeared to be hypocellular exhibiting a nodular disposition and included disorganized cells which were separated by an abundant chondromyxoid matrix. Osteoid formation was also observed. The histologic diagnosis was of a low-grade chondromyxoid lesion with features favoring an osteochondromyxoma (Figure 3).

The postoperative evolution was uneventful. A CT scan of the sinuses was taken 2 weeks and 8 months follow-

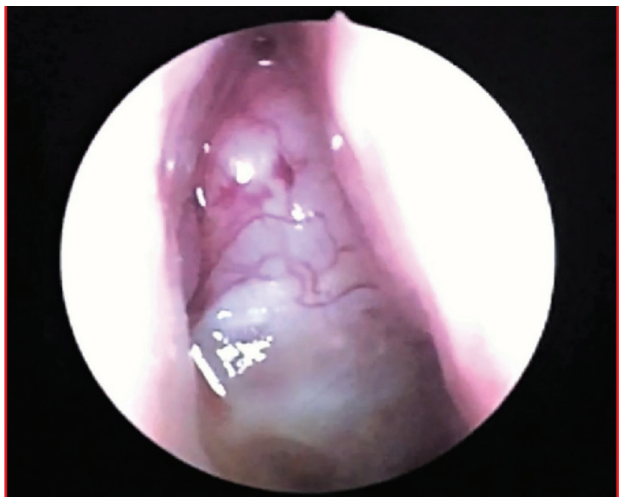

Figure 1: *Nasal endoscopic view of the right nasal cavity which demonstrated obstruction by a large whitish tumor with prominent blood vessels.*

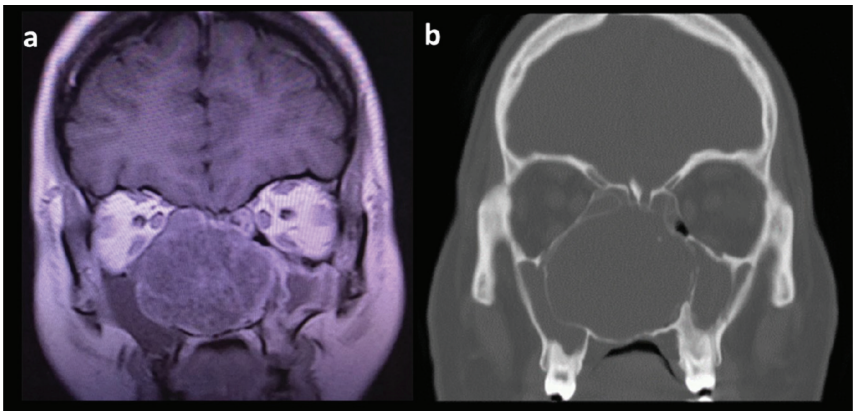

Figure 2: ***(a)*** *MRI,T1-weighted coronal plane depicts hypointense lesion with peripheral enhancement.* ***(b)*** *Coronal section CT scan with bone window demonstrating the expansive lesion with thinning of peripheral bone.*

ing the surgery (Figure 4). A large residual cavity without signs of recurrence can be observed. Clinically, the patient recovered olfaction and taste, and did not refer to any symptoms of recurrence. Because of the diagnosis and its possible association to Carney complex, the patient was evaluated by the cardiology, dermatology, gynecology, and endocrinology departments. An echocardiography and a thyroid ultrasound were performed, neither test revealed suspicious findings. A transabdominal pelvic ultrasound was also performed, and a unilateral ovarian cyst was detected. Blood test results did not show any hematological, biochemical, or endocrinological disorders.

Discussion

The prevalence of osteochondromyxomas is extremely low. Most cases reported have been in association to Carney complex; it is one of the 12 major diagnostic criteria [13]; however, it is uncommonly found in these patients with approximately 2% presenting with this tumor [6,7]. It is usually painless and presents symptoms based on its mass effect. It is a benign lesion but as it grows, it can present invasive features and expand or destroy bone [14]. In the case described here, the patient presented with a large mass in the nasal cavity which expanded and destroyed ethmoidal cells as well as part of the bony nasal septum, and also expanded the medial walls of the maxillary sinuses laterally. The mass effect can also be appreciated with the bowing of the hard palate.

This patient with a nasal osteochondromyxoma does not meet the diagnostic criteria for Carney complex at this point in time. Long-term follow-up is important as the patient can develop another diagnostic criterion in the future.

Conclusion

We have described a case of osteochondromyxoma of the nasal cavity. Although a rare tumor, osteochondromyxoma should be included as a differential diagnosis when evaluating tumors of the nasal cavity and paranasal sinuses. An accurate diagnosis is imperative because of its possible association with Carney complex. It is recommended that patients presenting with this tumor be evaluated by a multidisciplinary team in order to discard other potential features of Carney complex.

Acknowledgement

None

List of Abbreviation

None

Consent for publication

Informed consent was obtained from the patient to publish this case in a medical journal.

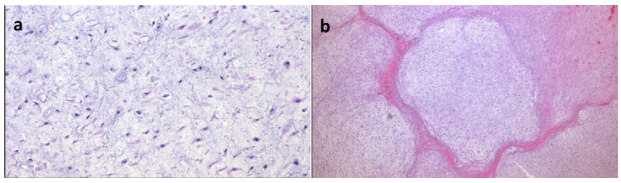

Figure 3. (a) *Polygonal and bipolar tumor cells with small nuclei were observed and lack of atypia (hematoxylin-eosin stain, original magnification ×400).* ***(b)*** *Nodular disposition and included disorganized cells which were separated by abundant chondromyxoid matrix (hematoxylin-eosin stain, original magnification ×400).*

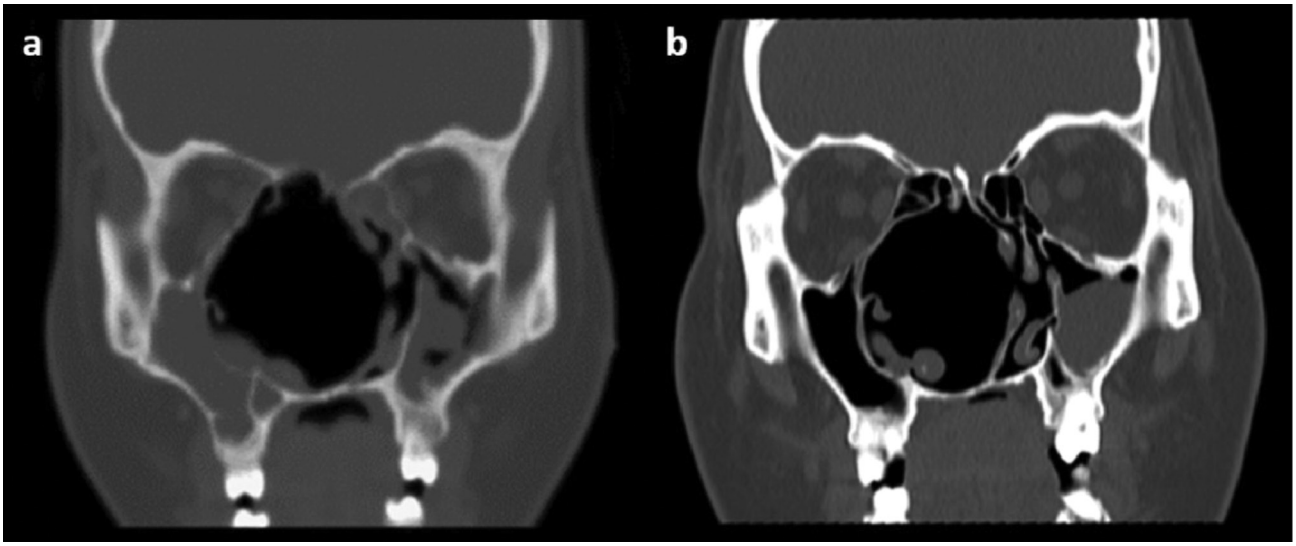

Figure 4. *Postoperative coronal CT imaging at* ***(a)*** *2 weeks and* ***(b)*** *8 months. Large residual cavity without any signs of recurrence.*

Ethical approval

Ethical approval was sought from the ethics committee at the Complejo Asistencial Dr. S del Rio Hospital.

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Summary of the case

Patient (gender, age)	1	Female, 21 years old
Final Diagnosis	2	Osteochondromyxoma of the nasal cavity
Symptoms	3	Nasal congestion, epistaxis, anosmia, ageusia, hyponasal speech, persistent unilateral tearing of the right eye, oropharyngeal dysphagia
Medications	4	None
Clinical Procedure	5	Endoscopic sinus surgery with complete removal of the tumor
Specialty	6	Otorhinolaryngology