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Case Report
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Osteochondromyxoma of the Temporomandibular Joint: Case Report of a Rare Criterion of the Carney Complex

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Abstract

Rationale: Osteochondromyxoma is an extremely rare benign tumor. Few cases have been described in the literature. Patients with Carney's complex can present with this tumor.

Patient concerns: A 59-years-old woman, with a 2-month history of multinodular goiter. Who presented for 1 year, pain in the Temporomandibular Joint (TMJ), as well as a limitation of the mouth opening with the appearance of a swelling in the preauricular region evolving for 1 year.

Diagnosis: A CT scan revealed a tumor centered on the right TMJ. An MRI confirmed the diagnosis of an intra-articular tumor.

Treatment and outcomes: Surgical excision was performed; the histological study of the tumor confirmed the diagnosis of the osteochondromyxoma.

Take-away lessons: Carney complex is a rare genetically heterogeneous syndrome of multiple endocrine neoplasia and lentiginosis that affects a number of organs. Osteochondromyxoma is a rare diagnostic criterion for this complex. The location of this tumor in the TMJ joint was never reported in the literature.

Keywords: Osteochondromyxoma; Temporomandibular joint; Carney complex; Bone tumor

Introduction

Osteochondromyxoma (OMX) is an extremely rare benign tumor. Few cases have been described in the literature. Patients with Carney's complex may present with this tumor [1,2].

We report a unique case of an osteochondromyxoma of the Temporomandibular Joint (TMJ) in a 59-year-old-patient with a history of multinodular goiter. Through this case we under line a very rare criterion of The Carney complex. Also, we report an exceptional location of the osteochondromyxoma. This tumor is a rare entity poorly documented in the literature, and there are no pathognomonic symptoms or imaging, which makes the diagnosis of this tumor challenging. Surgery is the main treatment and the diagnosis is confirmed by the histological study of the specimen.

Case Presentation

A 59-year-old-female with a chief complaint of pain in the temporomandibular joint, progressive swelling in the preauricular region and trismus, all this evolving for 1 year.

The patient has a medical history of multinodular goiter with a follow up in the ENT department. Extra-oral examination revealed a small mass in the preauricular region, fixed, non-tender, firm in consistency and the skin in that region appeared normal. Intra-oral examination revealed a limitation of the mouth opening (inter incisive distance measured was 2 cm) (Figure 1A), the rest of the intra oral examination was normal. Cervical examination revealed no cervical lymphadenopathy.

General examination revealed multiple spotty skin pigmentations on the face and the neck of the patient (Figure 1B).

Craniofacial computed tomography revealed a heterogeneous tumor centered on the right TMJ (Figure 2). MRI confirmed the diagnosis of a 35 mm long axis intra-articular tumor without endocranial extension.

The preoperative blood investigations were within normal limits, chest X-ray and echocardiogram showed no abnormality.

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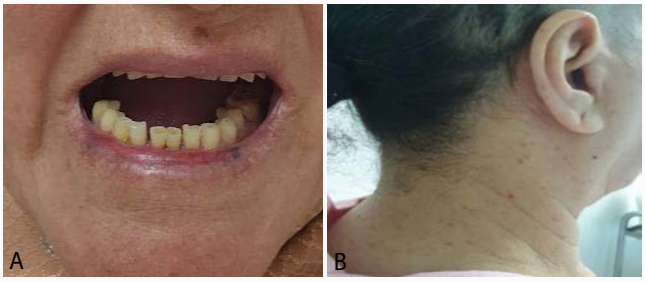


Figure 1: Preoperative pictures showing. A: Limitation of the mouth opening, B: Multiple spotty pigmentations (lentigines).

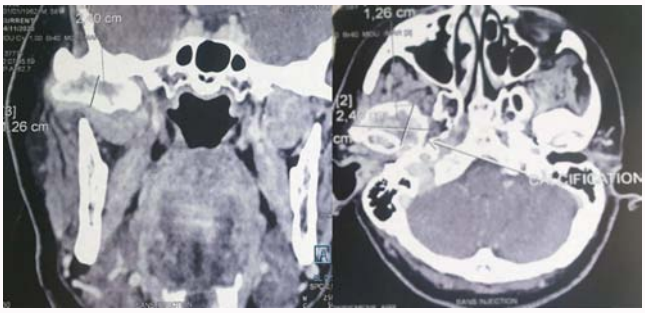


Figure 2: Craniofacial CT showing an intra-articular tumor.

Figure 3: Intraoperative picture showing, the approach and osteosynthesis of the zygomatic arch made after tumor resection.

Surgical excision was performed. The approach was made by a preauricular incision extended to the temporal region. Once on the zygomatic arc, we did an osteotomy of the arch so we can have access to the tumor (Figure 3). Total excision of the mass was performed. The zygomatic arch was fixed in its place by 2 microplates (Figure 3).

The postoperative follow-up was simple. The mouth opening was at 4cm and the pain disappeared (Figure 4).

Histologically, the tumor showed sheets and lobules of bland cells in myxomatous, cartilaginous, osseous, and hyaline fibrous matrix. HE X10 the tumor cells showed low to moderate cellularity without atypia (Figure 5A). HE X20 the diagnosis of osteochondromyxoma

Figure 4: Pictures of the mouth opening. A: Preoperative, B: Postoperative.

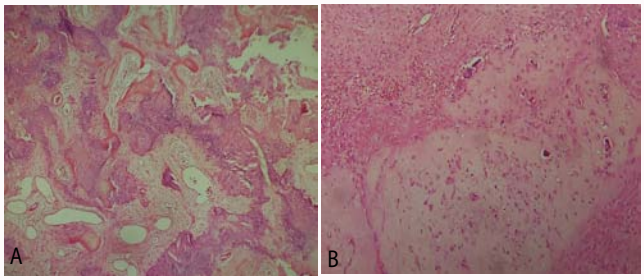


Figure 5: Histopathology of the patient showing. A: Sheets and lobules of bland cells in myxomatous, cartilaginous, osseous, and hyaline fibrous matrix. HEX10. B: The tumor cells showed low to moderate cellularity without atypia. HEX20.

was made (Figure 5B).

The presence of the osteochondromyxoma, the multinodular goiter and the spotty skin pigmentations confirmed the diagnosis of Carney complex.

Patient was referred to the endocrinologist, cardiologist and to genetic counseling for further investigations and treatment. No local recurrence was observed after 1 year follow up.

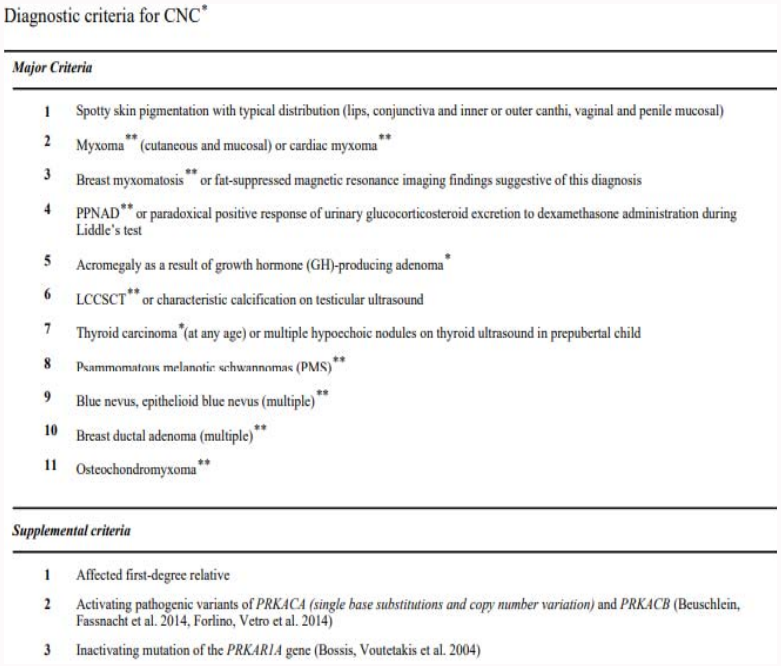
Discussion

Carney Complex (CNC) is a rare, autosomal dominant, genetically heterogeneous syndrome of multiple endocrine neoplasia and lentiginosis that affects a number of organs [1,2].

Described for the first time in 1985 by Carney et al. [1], the Carney complex is a multiple endocrine neoplasia associating cutaneous-mucous tumors (cutaneous-mucous myxomas, blue nevi or epithelioids, lentiginosis) or visceral (cardiac myxomas, mammary fibroadenomas, testicular, ovarian, thyroid tumors). It includes also, psammomatous melanotic schwannomas, large-cell calcifying Sertoli cell tumors, growth hormone-secreting pituitary adenomas, breast ductal adenomas and osteochondromyxomas [2,3].

The Carney complex is considered to be associated with a mutation of the PRKAR1A gene on chromosome 17q22-24 and another gene called CNC2 on chromosome 2p16 [4]. Molecular genetic studies of CNC have shown that it is linked to the regulatory subunit type I alpha of protein kinase A (PRKAR1A) gene located on 17q22–24, referred to as CNC1. CNC1 encodes PRKAR, which plays an important role in the cAMP signaling pathway. In addition, the CNC2 gene located on 2p16 was also detected in CNC, but its role needs to be further studied [4].

The diagnosis of CNC is based on the presence of at least two of major criterion, confirmed by histology (Figure 6). Also, the presence



Diagnostic criteria for CNC*

	Major Criteria
1	Spotty skin pigmentation with typical distribution (lips, conjunctiva and inner or outer canthi, vaginal and penile mucosal)
2	Myxoma** (cutaneous and mucosal) or cardiac myxoma**
3	Breast myxomatosis** or fat-suppressed magnetic resonance imaging findings suggestive of this diagnosis
4	PPNAD** or paradoxical positive response of urinary glucocorticosteroid excretion to dexamethasone administration during Liddle's test
5	Acromegaly as a result of growth hormone (GH)-producing adenoma*
6	LCCSCT** or characteristic calcification on testicular ultrasound
7	Thyroid carcinoma*(at any age) or multiple hypoechoic nodules on thyroid ultrasound in prepubertal child
8	Psammomatous melanotic schwannomas (PMS)**
9	Blue nevus, epithelioid blue nevus (multiple)**
10	Breast ductal adenoma (multiple)**
11	Osteochondromyxoma**

	Supplemental criteria
1	Affected first-degree relative
2	Activating pathogenic variants of *PRKACA (single base substitutions and copy number variation)* and *PRKACB* (Beuschlein, Fassnacht et al. 2014, Forlino, Vetro et al. 2014)
3	Inactivating mutation of the *PRKAR1A* gene (Bossis, Voutetakis et al. 2004)

Figure 6: Carney complex major and supplemental criterion [5].

of one supplemental criterion and one major criterion is sufficient to retain the diagnosis of Carney complex [5].

OMX is a rare diagnostic criterion for this complex. OMX of bone is an extremely rare tumor that is always associated with lentigines and other unusual disorders, and it has in fact been called "Carney bone tumor" since it is typically associated with Carney complex [3]. Most commonly this tumor occurs in long bones, paranasal sinuses and bones of the nose [2]. In 2015, 700 cases of Carney complex were described and approximately 2% presented with osteochondromyxoma [2,6]. In another study only 1% of Carney complex patients were diagnosed with an OMX [3]. In the literature and to the extent of our knowledge, no case of OMX of the temporomandibular joint associated or not with the Carney complex has been described.

There are no specific symptoms for this tumor, it is usually painless and presents with symptoms depending on its site and size and its mass effect. It is a benign lesion but as it grows, it can present invasive features and expand or destroy bone.

Imaging analysis can play an important role in any diagnosis of a bone tumor. OMX imaging is variable, it can be destructive, mineralized and can take different other images depending on its location and extend [6]. The radiological study made by Wei Yu et al. [7] and based on evidence presented in this study. They described a ring sign that can contribute to the imaging diagnosis and differential diagnosis of OMX in CNC.

Histologically, the tumor is usually composed of a mixture of mesenchymal cells, basophilic myxoid material, and mucopolysaccharide material. The degree of cellularity is inversely proportional to the amount of myxoid material and matrix [8].

Complete surgical excision of the OMX is curative, however, local recurrence is very common if the resection was incomplete [8,9].

Our patient presented an osteochondromyxoma of the right TMJ joint, confirmed by the histological study, a recent history of multinodular goiter and spotty skin pigmentations. Considering these 3 criteria, the diagnosis of Carney complex was made. The patient was referred to the endocrinologist, cardiologist and for genetic counseling to investigate the other criteria.

Through this case, we reported a very uncommon criterion of the Carney complex and we also presented an interesting surgical approach of the TMJ joint. The osteotomy of the zygomatic arc was the key to an open access to the tumor.

Conclusion

Although it is a rare tumor, once the diagnosis of osteochondromyxoma is retained and due to its possible association with the Carney complex. The patient must be treated by a multidisciplinary team in order to rule out other potential characteristics of the Carney complex.

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