

Up-to date review and case report

Clinical and radiological features of Jacob's disease. A case report involving an osteochondroma of the coronoid process

Chaabani Imen^{1,*}, Mziou Zouha², Zrig Ahmed³, Chaabouni Dorra⁴, Khochtali Habib⁵, Ben Alaya Touhami⁶

¹ Chaabani Imen, Assistant Professor, Department of Radiology, University Dental Clinic, 5000 Monastir, Tunisia

² Mziou Zouha, Assistant Professor, Department of Maxillo-Facial and Plastic Surgery, Sahloul University Hospital, 4000 Sousse, Tunisia

³ Zrig Ahmed, Assistant Professor, Department of Radiology, Fatouma Bourguiba University Hospital, 5000 Monastir, Tunisia

⁴ Chaabouni Dorra, Resident, Department of Radiology, University Dental Clinic, 5000 Monastir, Tunisia

⁵ Khochtali Habib, Professor and Chief of Maxillo-Facial and Plastic Surgery Department, Sahloul University Hospital, 4000 Sousse, Tunisia

⁶ Ben Alaya Touhami, Professor and Chief of the Radiology Department, University Dental Clinic, 5000 Monastir, Tunisia

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Abstract – Introduction: A large number of disorders affecting the masticatory system can cause mouth opening restrictions. Among them is Jacob's disease, characterized by restricted jaw movements and caused by pressure of the mandible coronoid process, which is longer than normal in size, on the posterior aspect of the zygomatic arch, together establishing new joint formation. **Observation:** We present a case report of a 29-year-old male patient presenting limited mouth opening. Inter-incisal maximum mouth opening was 5 mm. On panoramic radiograph an elongated coronoid process of the mandible was evident. Computed tomography (CT) scans showed the relationship between the exophytic mass and the inner surface of the zygomatic arch. An extra-oral coronoidectomy was performed. A mouth opening of 55 mm was achieved intra-operatively. The post-operative period was without complications. The histopathological diagnosis was osteochondroma. **Discussion:** The clinical, radiological and histopathological characteristics and surgical approach to Jacob's disease are discussed. **Conclusion:** In a patient with a limitation of mouth opening, and without any temporo-mandibular joint disease, an examination of the coronoid process is required to identify hypertrophy of the coronoid process and to diagnose Jacob's disease.

Mots clés :
maladie de Jacob /
articulation
temporo-mandibulaire /
processus coronoïde

Résumé – Aspects cliniques et radiologiques de la maladie de Jacob : à propos d'un ostéochondrome du processus coronoïde. Introduction : De nombreux troubles du système masticoire peuvent limiter les mouvements mandibulaires. La maladie de Jacob est une maladie assez rare, caractérisée par une limitation de l'ouverture buccale, causée par la pression du processus coronoïde hypertrophié sur la face postérieure de l'arcade zygomatique, établissant ainsi une nouvelle surface articulaire. **Observation :** Nous présentons le cas d'un patient âgé de 29 ans consultant pour une limitation de l'ouverture buccale à 5 mm, le panoramique n'avait pas montré d'anomalies des structures articulaires temporo-mandibulaires. Il a mis en évidence une élongation du processus coronoïde. L'examen tomographique a précisé la relation entre l'hypertrophie du processus coronoïde et la surface interne de l'arcade zygomatique. Une coronoidectomie par voie extra-orale a été réalisée. Une ouverture buccale de 55 mm a été récupérée en peropératoire, en postopératoire et lors des contrôles réguliers, aucune complication n'a été relevée. Le diagnostic anatomopathologique était en faveur d'un ostéochondrome du processus coronoïde. **Discussion :** Nous discutons dans cet article les particularités cliniques, radiologiques et histopathologiques ainsi que l'approche chirurgicale de cette maladie rare. **Conclusion :** Chez un patient présentant une limitation de l'ouverture buccale et en l'absence de pathologies des structures articulaires temporo-mandibulaires, l'examen du processus coronoïde s'impose à la recherche d'une hypertrophie en faveur du diagnostic de la maladie de Jacob.

* Correspondence: chaabanimen@gmail.com

Introduction

Jacob's disease is one of the numerous causes of reduction of mouth opening. It is not frequently reported. It was first reported by Langenbeck in 1853 but it was Jacob, in 1899, who first described osteochondroma of the coronoid process forming a pseudoarthrosis between the coronoid process and the zygomatic arch. Later, very few cases have been reported [1]. Patients usually complain of restricted and painful mouth opening. Therefore, these patients may be treated for a misdiagnosis of temporomandibular joint (TMJ) disorders [2]. The treatment of a Jacob's disease is surgical, with an intraoral or extraoral approach. We report an unusual case of a patient who presented with severe mouth opening limitations. Our report is intended to highlight the classical clinical and radiological features of a rarely reported condition of a Jacob's disease.

Case presentation

A 29-year-old man presented with a history of a 10-years progressive and painless limitation of mouth opening. The patient's brother does not have a similar complaint.

The maximum opening was 5 mm, with deviation to the left, and a minimally prominent left zygoma. There was no apparent facial asymmetry (Fig. 1). All the other aspects of the patient's history, as well as the physical and laboratory examinations were within normal limits. He had, also, no previous history of trauma in the facial area. The originality of this case is that the patient, when referred by his doctor, was not aware of the reduction in his mouth opening before consultation, although, on examination, he appeared to be well-developed and healthy. At first, the patient was diagnosed as having bilateral temporomandibular joint (TMJ) disorder. Orthopantomogram (OPG) and tomography of TMJ were conducted. A panoramic radiograph revealed no gross abnormalities, with the exception of an enlargement of the left coronoid process

(Fig. 2). Both examinations showed normal joints bilaterally, but without condylar movement associated with mouth opening. However, the anatomic features of the lesion could not be seen clearly from these views. Then, in addition to plain film radiographs and to have a clear idea about this hypertrophy of the coronoid process and its relationships with head-adjacent structures, a computed tomography (CT) imaging was performed. This examination revealed a well-corticated exophytic protuberance projecting anteriorly and superiorly from the hypertrophied left coronoid process. In fact, axial computerised tomography (CT) and coronal CT scans revealed the presence of a mushroom-shaped radiodense mass at the apex of the coronoid, the zygomatic arch was curved laterally by this bony outgrowth. It also revealed a remodelling of the inner aspect of the zygomatic arch, resulting in a pseudoarticulation (Fig. 3).



Fig. 1. View of patient showing restricted mouth opening, with deviation of the mandible to the left and left zygomatic arch expansion. *Fig.1. Patient présentant une limitation de l'ouverture buccale, une latéro-déviation mandibulaire et une expansion de l'arcade zygomatic gauche.*

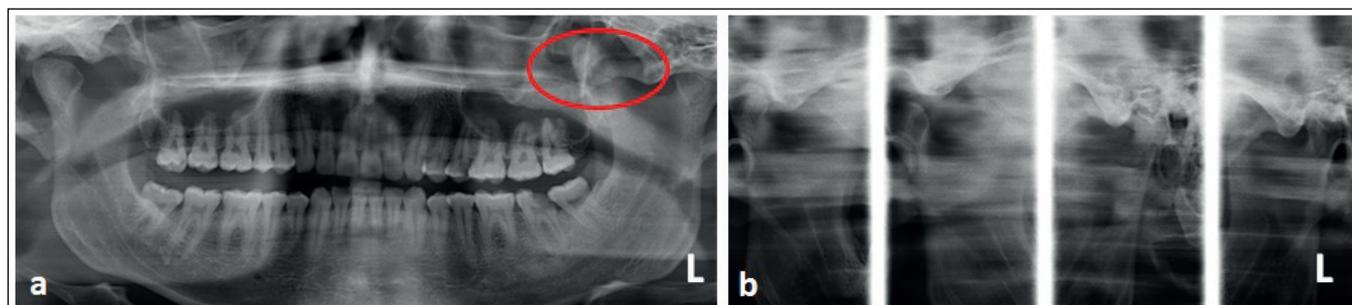


Fig. 2. a. Orthopantomography: hyperplasia of the left coronoid process. b. Sagittal Tomography of temporomandibular joint (mouth closed, mouth open): Bilateral limited condylar movement, especially on the left.

Fig.2.a. Radiographie panoramique : hypertrophie du processus coronoïde gauche. b. Tomographies sagittales des articulations temporo-mandibulaires (bouche fermée, bouche ouverte) : limitation bilatérale des mouvements condyliens, accentuée à gauche.

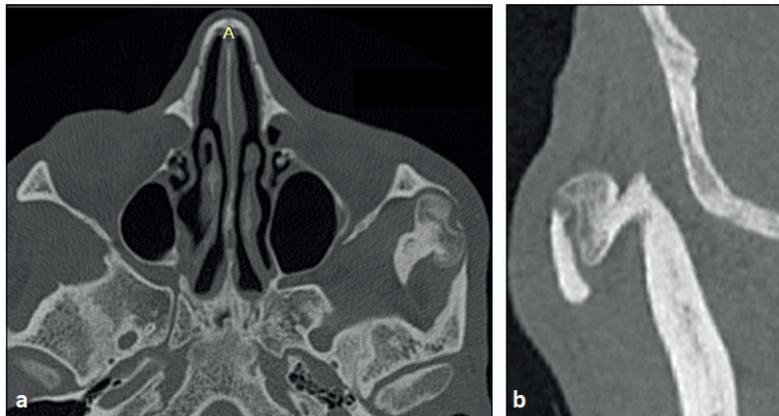


Fig. 3. a. Axial CT section showing a mushroom-shaped osteochondroma of the left coronoid process and the remodelling of the inner surface of the left zygomatic arch. b. Coronal CT section: unilateral hyperplasia of the left coronoid process.

Fig.3.a. Coupe axiale tomodensitométrique : hypertrophie ostéocartilagineuse coiffant le processus coronoïde gauche, à noter le remodelage de la surface interne de l'arcade zygomatique. b. Coupe coronale tomodensitométrique : hypertrophie unilatérale du processus coronoïde gauche refoulant en dehors le processus zygomatique et les tissus mous en regard.

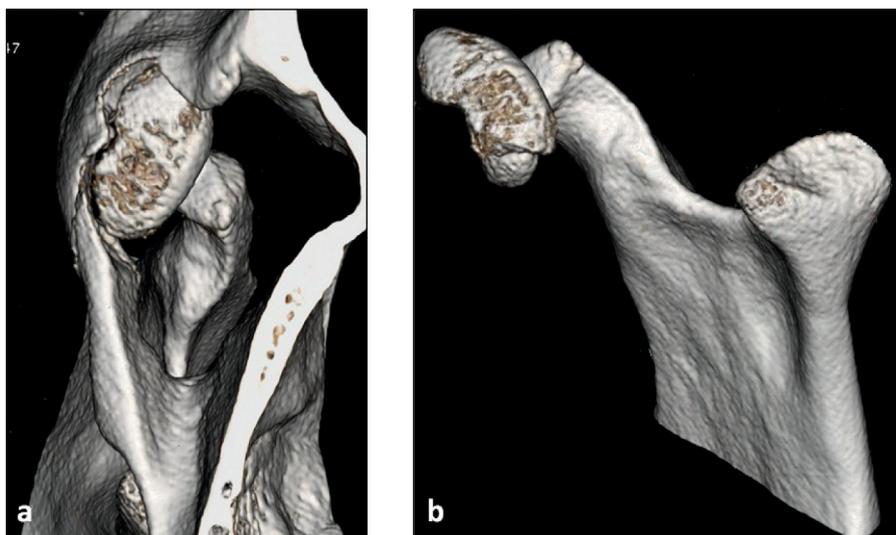


Fig. 4. a. Three dimensional CT image, showing the articulation (pseudoarthrosis) formed between the zygoma and the sessile mass, arising from the coronoid process. b. Three dimensional CT image of the vertical mandible ramus, showing mushroom-shaped osteochondroma of the left coronoid process.

Fig.4.a. Reconstruction tridimensionnelle : pseudo-articulation entre le processus zygomatique et l'hypertrophie coronoïdienne. b. Reconstruction tridimensionnelle de la branche mandibulaire : hypertrophie ostéocartilagineuse du processus coronoïde gauche.

Moreover, 3D reconstruction showed the relationship between the exophytic mass and the inner surface of the zygomatic arch, making a new joint formation especially clear at the upper view of the 3D CT scan (Fig. 4). The patient was then scheduled for excision of the exophytic mass by extraoral approach (Fig. 5). A mouth opening of 55 mm was achieved intra-operatively. The patient was recommended to have post-operative physiotherapy and jaw exercises. Histologically, the diagnosis of osteochondroma of the coronoids was carried out.

In fact, pathologic examination showed typical histologic characteristics with cancellous bone capped by hyaline cartilage which was undergoing endochondral ossification (Fig. 6).

Discussion

Coronoid process enlargement is a condition that can result from exostosis, osteoma, osteochondroma, chondroma,

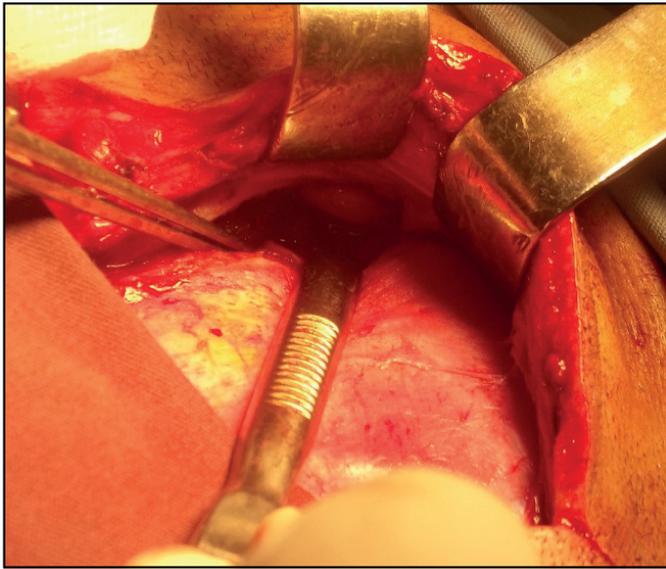


Fig. 5. Resection of the osteochondroma of the left coronoid process by an external coronal approach.

Fig.5. Résection de l'ostéochondrome du processus coronoïde gauche avec une approche chirurgicale extra-orale coronale.

hyperplasia and developmental anomalous [1]. First described in 1899, Jacob's disease can eventually lead to the formation of a pseudojoint between the zygoma and the coronoid process. In most of the patients with Jacob's disease, the involved coronoid segment was diagnosed histologically as osteochondroma, whereas hyperplasia was diagnosed in a few patients. To establish this diagnosis, we need to show a direct contact between the hyperplastic coronoid process and the posterior wall of the maxilla or the zygomatic arch and the joint surfaces at this location [2]. The etiology of the complaint is unknown, although several theories have been postulated, including hyperactivity of the temporal muscle, dysfunction of the temporomandibular joint, endocrine stimuli, traumatism and even genetic and family factors [3, 4]. Jacob's disease is most frequent in young patients with a mean age of 27 years (age range: 16-62 years) with male predominance. It is usually unilateral in occurrence with predilection for involvement of the left coronoid process [5]. These results were in accordance with those of our case report.

The mandibular hypomobility can lead to secondary problems such as airway problems, malnutrition and growth retardation, negative impact on speech development, limited access to oral hygiene and dental care, and muscle atrophy [6].

In this case report, the patient had never been subjected to any dental treatment due to mouth opening limitation. Because of its insidious clinical onset, Jacob's disease is often overlooked and misdiagnosed as a TMJ disorder, which could lead to mistreatment [7]. For this reason, we carried out a panoramic radiograph and tomography of TMJ. The panoramic radiograph showed an abnormal elongated left coronoid process,

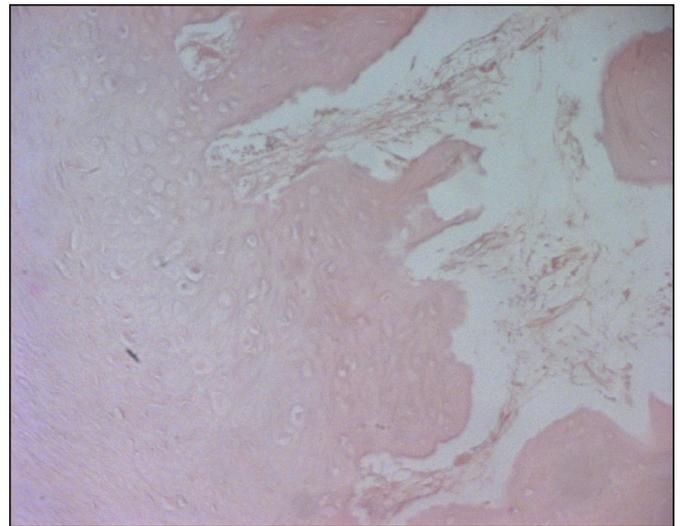


Fig. 6. Photomicrograph showing cancellous bone capped by hyaline cartilage with an endochondral ossification (Hematoxylin and eosin stain × 200).

Fig.6. Photomicrographie : os spongieux coiffé par un cartilage hyalin comportant une ossification onchondrale (hématoxyline et éosine × 200).

the tomography of TMJ showed a bilateral limited condylar movement especially at left. However, the anatomic features of the lesion could not be seen clearly from these views. So, in addition to plain film radiographs, recently computed tomography has been a beneficial imaging modality. CT has an important role in diagnosis and is useful for an adequate surgical planning by allowing assessment of the size of impingement of the coronoid processes. On axial CT images, protrusion of hypertrophic segment to temporal fossa and articulation of this segment with the inner aspect of zygomatic arch can be clearly seen. 3D CT shows the elongated coronoid process passed above the zygomatic arch and joint formation. Furthermore, 3D CT provides some additional measurement information for surgeons. Therefore, a 3D CT scan is very useful and revealing as it demonstrates the hyperplastic coronoid process, the joint surfaces and the changes in the inner aspect of the zygomatic arch. Joint formation can occur in two different models: (1) impingement of the coronoid process on the concavity formed at the zygomatic arch, (2) concavity on a coronoid process caused by the new bone formation on the medial surface of zygoma. The type of joint formation might determine a surgical approach [2]. The treatment of coronoid hyperplasia, essentially a mechanical problem, is primarily surgical without reconstruction. In fact, the treatment's goal will be to recover acceptable mouth opening ranges.

Different approaches have been advocated to treat this condition. Most of the previously reported cases of coronoid hyperplasia and Jacob's disease had been treated through an intraoral approach which avoids the surgical complications

inherent in the extraoral approaches such as facial motor nerve injury and facial scarring, although limitations of this approach are well recognized.

Extraoral approaches also have been described, this approach should be used in the following situations: 1) When the size and position of the lesion prevent removal by an intraoral approach. This can easily be determined from the CT scan; 2) In cases with concomitant involvement of the TMJ; 3) In bilateral cases. In this patient, an external coronal approach was indicated due to the limitation of mouth opening and because the coronoid process was large enough to be trapped above the arch, with the advantage of better visualization and esthetic scar within the line of hair [8-10].

Despite the immediate gain in jaw mobility, physical therapy is often necessary to help and maintain an effective mouth opening. Our patient's follow-up showed he was healing well and had improved mouth opening.

Conflicts of interests: none declared

Bibliography

1. Yesildag A, Yariktas M, Doner F, Aydin G, Munduz M, Topal U. Osteochondroma of the Coronoid Process and Joint Formation with Zygomatic Arch (Jacob Disease): Report of a Case. *Eur J Dent* 2010;4:91-94.
2. Akan H, Mehreliyeva N. The value of three-dimensional computed tomography in diagnosis and management of Jacob's disease. *Dentomaxillofac Radiol* 2006;35:55-59.
3. Fernández Ferro M, Fernández Sanroman J, Sandoval Gutierrez J, Costas Lopez A, Lopez de Sanchez A, Etayo Perez A. Treatment of bilateral hyperplasia of the coronoid process of the mandible. Presentation of a case and review of the literature. *Med Oral Patol Oral Cir Bucal* 2008;13:595-598.
4. D'Ambrosio N, Kellman RM, Karimi S. Osteochondroma of the coronoid process (Jacob's disease): an unusual cause of restricted jaw motion. *Am J Otolaryngol* 2011;32:52-54.
5. Ajila V, Hegde S, Gopakumar R, Babu GS. Imaging and Histopathological Features of Jacob's Disease: A Case Study. *Head Neck Pathol* 2012;6:51-53.
6. Costa YM, Porporatti AL, Stuginski-Barbosa J, Cassano DS, Bonjardim LR, Conti PC. Coronoid Process Hyperplasia: An unusual Cause of mandibular Hypomobility. *Braz Dent J* 2012;23:252-255.
7. Stringer DE, Chatelain KB, Tandon R. Surgical Treatment of Jacob's Disease: A Case Report Involving an Osteochondroma of the Coronoid Process. *Case Rep Surg* 2013:1-3.
8. Hernandez-Alfaro F, Escuder O, Marco V. Joint formation between an osteochondroma of the coronoid process and the zygomatic arch (Jacob Disease): report of case and review of literature. *J Oral Maxillofac Surg* 2000;58:227-232.
9. Villanueva J, González A, Cornejo M, Núñez C, Encina S. Osteochondroma of the coronoid process of the mandible. Report of a case and review of the literature. *Med Oral Patol Oral Cir Bucal* 2006;11:E289-91.
10. Thota G, Cill JE, Krajekian J, Dattilo DJ. Bilateral pseudojoints of the coronoid process (Jacob Disease): Report of a case and review of the literature. *Oral Maxillofac Surg* 2009;67:2521-2524.