http://dx.doi.org/10.1590/1981-86372021000562019012

| CLINICAL

Jacob's Disease: case report

Doença de Jacob: relato de caso

Marilia de Oliveira Coelho Dutra **LEAL**[1] https://orcid.org/0000-0003-1164-9983

Leonard Duarte **MOREIRA**[2] https://orcid.org/0000-0003-3720-7382

Fabrício **PASSADOR-SANTOS**[3] https://orcid.org/0000-0003-2777-8640

Andressa Borges **SOARES**[3] https://orcid.org/0000-0001-6112-3096

Rubens Gonçalves **TEIXEIRA**[2] https://orcid.org/0000-0003-0814-0940

Cláudio Roberto Pacheco **JODAS**[2] https://orcid.org/0000-0002-9750-2714

ABSTRACT

Jacob's disease or osteochondroma is a rare disease that presents as a pseudo-joint between the coronoid process of the mandible and the posterior surface of the zygomatic bone. The present case report is of an 11-year-old female patient with a maximum mouth opening of 2mm. When evaluating computed tomography with three-dimensional reconstruction, a mushroom-shaped increase in the coronoid process of the mandible on the left side was observed, intimately connected to the zygomatic bone (inferoposterior region of the body of the zygoma), which suggests a pseudoarticulation. Surgical removal of the tumor mass was performed under general anesthesia via intraoral access and is called coronoidectomy. Histopathological, the presence of hyaline cartilage was observed, a condition that is pathognomonic for Jacob's Disease. Jacob's disease case reports are still rare in the literature, with mushroom shape and histopathological confirmation due to the presence of hyaline cartilage.

Indexing terms: Hyaline cartilage. Mandible. Osteochondroma

RESUMO

A Doença de Jacob ou osteocondroma é uma doença rara que se apresenta como uma pseudoarticulação entre o processo coronóide da mandíbula e a superfície posterior do osso zigomático. O presente relato de caso é de um paciente, do sexo feminino, com 11 anos de idade e com 2mm de abertura máxima de boca. Ao se avaliar exame de tomografia computadorizada com reconstrução tridimensional foi observado um aumento volumétrico no processo coronóide da mandíbula do lado esquerdo em formato de cogumelo, intimamente ligado ao osso zigomático (região inferoposterior do corpo do zigoma), o que sugere uma pseudoarticulação. A remoção cirúrgica da massa tumoral foi realizada sob anestesia geral por acesso intraoral e é denominada coronoidectomia. Histopatologicamente, observou-se presença de cartilagem hialina, condição é que patognomônica da Doença de Jacob. Ainda são raros os relatos de casos de Doença de Jacob na literatura, com formato de cogumelo e confirmação histopatológica devido à presença de cartilagem hialina.

Termos de indexação: *Cartilagem hialina. Mandíbula. Osteocondroma.*

[1] Universidade Estadual de Campinas, Faculdade de Odontologia de Piracicaba, Departamento de Patologia. Av. Limeira, 901, Areião, 13414-018, Piracicaba, SP, Brasil. Correspondence to: MOCD Leal. E-mail: <marilialeal@hotmail.com>.

[2] Faculdade São Leopoldo Mandic, Instituto de Pesquisas São Leopoldo Mandic, Departamento de Cirurgias Bucomaxilofaciais. Rua Dr. José Rocha Junqueira, 13, 13045-755, Ponte Preta, Campinas, SP, Brasil.

[3] Faculdade São Leopoldo Mandic, Instituto de Pesquisas São Leopoldo Mandic, Departamento de Patologia. Campinas, SP, Brasil.

How to cite this article

Leal MOCD, Moreira LD, Passador-Santos F, Soares AB, Teixeira RG, Jodas CRP. Jacob's Disease: case report. RGO, Rev Gaúch Odontol. 2021;69:e20210056. http://dx.doi.org/10.1590/1981-86372021000562019012

INTRODUCTION

Jacob's Disease or osteochondroma of the coronoid process of the mandible is a rare tumor in the maxillofacial complex. It was first reported by Langeback in 1853, as an increase of coronoid process of the mandible and, in 1899 [1], it was described for the first time by Oscar Jacob [2], as a pseudoarticulation, mushroom shap, between the coronoid process and the arch/body of the zygomatic bone (osteochondroma).

Due to difficulty of detection in early stages of development and similarity of symptoms with other joint disorders, this pathology often is erroneously considered as a temporomandibular disorder [3]. Pain is uncommon and mainly affects young patients [4].

From the first documentation of Jacob's Disease until 2010, only 50 cases have been reported in literature, although only 39 had histological evidence of pseudoarticulation [5,6]. Reports point to male prevalence (63%), mean age of 30 years-old (5-73 years) [5] and slight predilection for the left side [1]. The first clinical sign and main presenting characteristic is a significantly reduced mouth opening [5]. Another possible symptom is face asymmetry due to decreased muscle volume, especially of the masseter muscle.

Diagnosis requires imaging exams, and Computed Tomography (CT) is considered the "gold standard", along with three-dimensional reconstruction in computer-assisted surgical planning. Radiographs, especially panoramic, are also used [2,7,8].

Coronoidectomy treatment reinstates mouth opening and surgical accesses can be extra and/or intraoral, depending on each case. In addition to the surgical team, treatment continuation (rehabilitation) requires multidisciplinary care, including speech therapists and physical therapists [7,8].

CASE REPORT

In 2014, a black female patient was the victim of an automobile accident and had a fracture of the left coronoid process. She had not been submitted to fracture reduction at the time.

In 2016, two years after facial trauma, the patient was 14 years-old and attended maxillofacial surgery service at Faculdade São Leopoldo Mandic, in Campinas/SP, complaining of severe limitation of mouth opening (2mm) and great difficulty eating (figure 1). Face CT Scan showed a volumetric increase, intra and extraoral, in the coronoid process of the mandible that generated a region of contact with the zygomatic bone – posterior surface – on the left side (figures 2 and 3). Comorbidity or painful symptoms had not been associated.

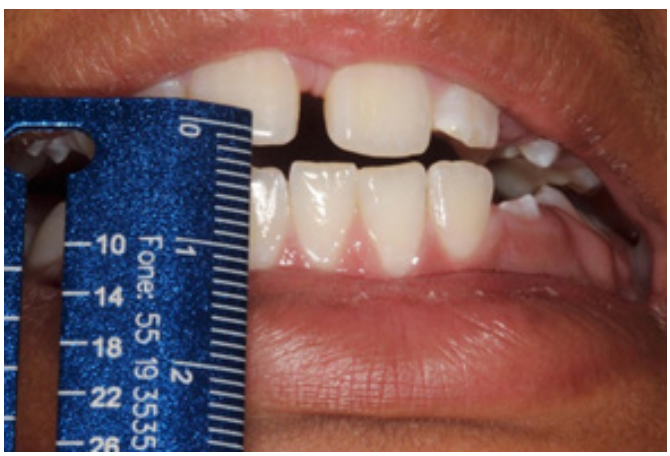

Figure 1. Mouth opening limitation (2mm).

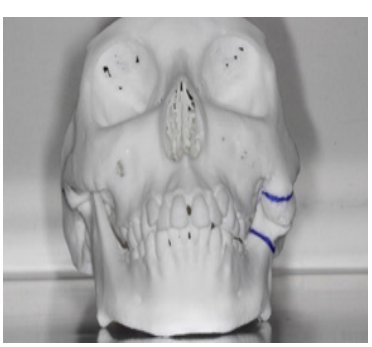

Figure 2. Reconstruction shows swelling in the zygomatic bone and coronoid process (front view).

Figure 3. Reconstruction shows swelling in the zygomatic bone and coronoid process (left lateral view).

The following preoperative tests were required: CT, face radiographs (P.A of mandible and lateral oblique of mandible left side), medical laboratory exams and pre-operative anesthesia visit.

More specific tests had not been necessary. The following diagnosis hypotheses have been suggested: ankylosis of the coronoid process and Jacob's Disease.

Surgery was performed in Ouro Verde Hospital, at Campinas/SP, under general anesthesia, through nasotracheal intubation (by nasofibroscopy due to the impossibility of orotracheal intubation due to limited mouth opening). Access was intraoral, incision along the bottom of the vestibule groove in left side of the maxilla (from the zygomatic pillar to the coronoid process and mandible branch). After tissue divulsion, the lesion area was exposed, which was evaluated using an endoscope, and a fusion was observed between structures of the zygomatic bone and an osteochondroma of the coronoid process (Figure 4A). Two parallel osteotomies lines were performed on each side of the lesion, with 1.5 cm of distance between them and the lesion that was removed (figure 4B). Consequently, an immediate release of mouth opening occurred (from 2mm to 25mm postsurgical) (figure 5). CT Scans were taken immediately after the surgery. Medication prescribed: Clindamycin 300mg (12/12h) oral, Nimesulida 40mg (08/08h) oral and Dipyrone 500mg (06/06h) oral, accompanied by physiotherapy and facial drainage. The patient had a satisfactory recovery and was discharged 48 hours after surgery.

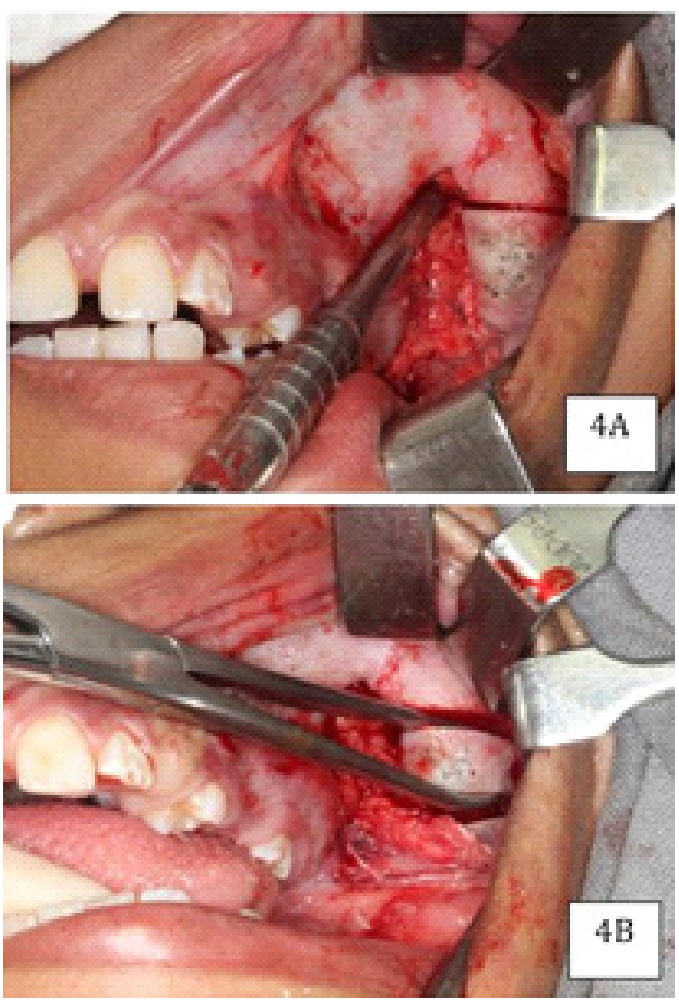

Figure 4. A) Fusion between structures of the zygomatic bone and the osteochondroma of the coronoid process; B) Two parallel osteotomies, 1.5 cm apart to remove the lesion.

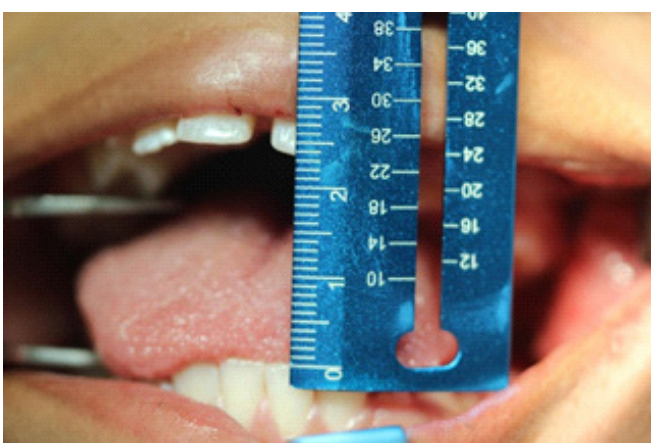

Figure 5. 25mm mouth opening in trans-surgery.

A sample of four fragments was histologically analyzed, the largest fragment measured 3.0 x 2.0 x 1.0 cm and smallest 1.0 x 0.5 x 0.5 cm. The sample consists of fibrous tissue in continuity with cartilaginous tissue, irregularly shaped, brownish in color and stone consistency. Histological analysis revealed fragments of trabecular bone tissue fused and permeated with hyaline cartilaginous tissue. Between the trabeculae of tissue bone, hematopoietic tissue was noted (figure 6). In some areas fibrous connective tissue was present in close contact with the trabecular bone tissue and cartilage hyaline (figure 6, detail).

DISCUSSION

Increase of the coronoid process can occur due to osteochondromas, hyperplasia, exostoses, chondromas,

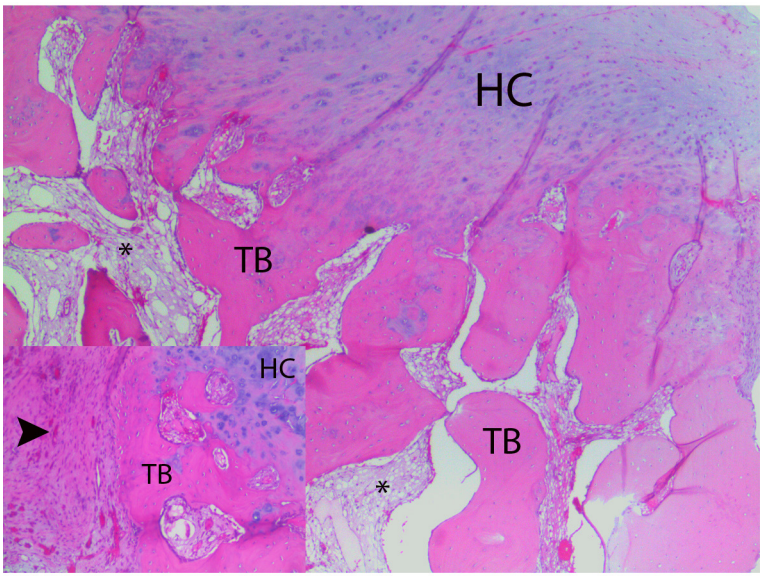

Figure 6. Histological features of Jacob's disease. Coronoid process shows trabecular bone (TB) mixed with hyaline cartilage (HC). The hematopoietic bone marrow (asterisk) is seen between the trabecular bones (H&E X 40 original magnification). Detail shows another area of the lesion with dense connective tissue (arrowhead) in contact with trabecular bone (TB) and hyaline cartilage (HC) (H&E X 100 original magnification). Hematoxylin and Eosin (H&E).

osteomas and/or growth abnormalities [9]. Osteochondroma is the most common benign bone tumor (exostoses) with cartilaginous components. They are rare in the maxillofacial region and at the coronoid process and their development usually occurs in the 20- to 40-year age group. The most common clinical features are gradual reduction of mouth opening, which leads to trismos and facial asymmetry, due to the increased of volume in zygomatic region [2].

The adhesion between the coronoid process and the zygomatic bone can be caused by firearms projectiles, fractures of zygomatic process treated or not, associated (or not) with fractures of the coronoid process, chemical burns, mandibular fractures, infections involving infratemporal space, local surgical complications and intracapsular ankylosis extensions [10]. The process of Ankylosis is still unknown.

A direct trauma of the zygomatic arch causes more damage to extra articular tissue as compared to the temporomandibular joint (TMJ) itself. TMJ generally suffers most from indirect damage, for example trauma in chin region [11], as compared with direct traumas. A direct trauma into the zygomatic arch can cause it to fracture and a create a delimited hematoma, which without appropriate physiotherapy can result in an ankylosis of the arch with the coronoid process [12].

There is no consensus on the etiopathogenesis of ankylosis, but different factors are postulated as possible causes, including temporal muscle hyperactivity, previous trauma, chronic disk displacement of the ipsilateral TMJ, indoctrinate stimuli and genetic changes [4]. Coronoid hyperplasia is a similar condition and probably occurs due to trauma or changes in the movement pattern of the temporal muscle. One hypothesis about pseudo joint formation is an increase in the coronoid process that causes compression, resorption and remodeling next to zygomatic bone [6].

The final diagnosis (gold standard) had been determined by histopathology, when presence of bone hyperplasia is confirmed, of cartilage and of the synovial capsule forming a new joint between the zygomatic bone and the coronoid process [4].

This type of osteochondroma is relatively rare in the human body as a whole and mainly occurs in the facial maxillary skeletal structure. Because most ossifications of this area are of intramembranous origin, and osteochondroma formation has endochondral origin [13], Jacob's Disease is rare and results from a pseudoarticulation between the zygomatic arch surface and the coronoid process due to a constant contact between these two surfaces [14].

This case report corroborates literature findings, as they were found in histopathological analysis: hyaline

cartilage, bone, and fibrous elements. All compatible with pseudoarthrosis formation [1,15,16].

Symptomatic coronoid processes are rare, so Jacob's disease is often initially treated as a simple TMJ ankylosis. Inadequate care or a wrong diagnosis can lead to ankylosis development [14]. Panoramic radiography can show coronoid process widening. However, CT examination (with reconstruction) is the gold standard for a correct diagnosis [7,816], because it can show the bone growth in more detail, in addition to its relationship with the zygomatic bone. In the present case, CT reconstruction revealed a mushroom-shaped growth (maximum length 45 mm) over the left coronoid process and next to the medial wall of the zygomatic arch.

The growth of a mushroom-shaped tumor is usually noticed on routine radiography exams or during palpation of the area [13]. In early stages this tumor remains asymptomatic. However, patients report a progressive limited mouth opening as the tumor develops. Despite all the clinical and radiographic features suggesting Jacob's disease treatment, definitive diagnosis should be done by histopathologic examination of the removed tumor mass.

Surgeons usually proceed with intraoral access (49%) for coronoidectomy, though extraoral access is common as well (40%) and a few use intra and extraoral combined (11%). There are controversies about the approach to surgical access for ankylosis [6], however, intraoral access is the more adequate for aesthetics and has lower risk of facial nerve rupture [1]. Coronoidectomy is the treatment choice, there are reports in literature of a 2% recurrence rate using this technique [16]. Intraoral access was the choice in this case report, due to the patient's young age and reduced tumor dimensions, in addition the risk of scarring and facial nerve neuropraxia were also reduced.

CONCLUSION

The coronoid process must always be evaluated together with the TMJ examination in cases of limited mouth opening. Coronoidectomy is the treatment of choice for Jacob's Disease. Confirming the presence of hyaline cartilage during the histopathological examination is essential for confirming the diagnosis of Jacob's Disease.

Collaborators

MOCD Leal, review of the text and preparation of paper. LD Moreira, surgical team that conducted the case and preparation of paper. F PASSADOR-SANTOS and AB SORES, histopathological analysis and text review RG TEIXEIRA and CRP JODAS, surgical team that conducted the case and preparation of paper.

REFERENCES

1. Hernández-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(2):227-32. https://doi.org/10.1016/S0278-2391(00)90345-8
2. Aoki N, Okamura K, Niino D, Iwamoto O, Kusukawa J. Osteochondroma of the right coronoid process (Jacob disease): a case report. Cranio. 2013;31(1):66-9. https://doi.org/10.1179/crn.2013.010
3. Roychoudhury A, Gupta YK, Parkash H, Karak AK. Jacob disease: report of a case and review of the literature. J Oral Maxillofac Surg. 2002;60(6):699-703. https://doi.org/10.1053/joms.2002.33125
4. Coll-Anglada M, Acero-Sanz J, Vila-Masana I, Navarro-Cuéllar C, Ochandiano-Caycoia S, López de-Atalaya J, et al. Jacob's disease secondary to coronoid process osteochondroma. A case report. Med Oral Patol Oral Cir Bucal. 2011;16(6):e708-10. http://dx.doi.org/doi:10.4317/medoral.16820
5. Choi JG, Kim SY, Perez-Atayde AR, Padwa BL. Bilateral coronoid process hyperplasia with pseudocartilaginous joint formation: Jacob disease. J Oral Maxillofac Surg. 2013;71(2):316-21. https://doi.org/10.1016/j.joms.2012.05.029
6. Sreeramaneni SK, Chakravarthi PS, Krishna Prasad L, Raja Satish P, Beeram RK. Jacob's disease: report of a rare case and literature review. Int J Oral Maxillofac Surg. 2011;40(7):753-7. https://doi.org/10.1016/j.ijom.2011.02.011
7. Akan H, Mehreliyeva N. The value of three-dimensional computed tomography in diagnosis and management of Jacob's disease. Dentomaxillofac Radiol. 2006;35(1):55-9. https://doi.org/10.1259/dmfr/52275596
8. Escuder i de la Torre O, Vert Klok E, Marí i Roig A, Mommaerts MY, Pericot i Ayats J. Jacob's disease: report of two cases and review of the literature. J Craniomaxillofac Surg. 2001;29(6):372-6. https://doi.org/10.1054/jcms.2001.0247
9. 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(1):91-4.
10. Agarwal M, Gupta DK, Tiwari AD, Jakhar SK. Extra-articular ankylosis after zygoma fracture; A case report & review of literature. J Oral Biol Craniofac Res. 2013;3(2):105-7. https://doi.org/10.1016/j.jobcr.2013.05.003
11. Zhong SC, Xu ZJ, Zhang ZG, Zheng YH, Li TX, Su K. Bilateral coronoid hyperplasia (Jacob disease on right and elongation on left): report of a case and literature review. Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 2009;107(3):e64-7. https://doi.org/10.1016/j.tripleo.2008.10.017
12. Wenghoefer M, Martini M, Anwander T, Götz W, Reich R, Bergé SJ. [Hyperplasia of the coronoid process: diagnosis and

treatment]. Mund Kiefer Gesichtschir. 2006;10(6):409-14. https://doi.org/10.1007/s10006-006-0028-2

13. 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;2013:253740. https://doi.org/10.1155/2013/253740
14. Losa-Muñoz PM, Burgueño-García M, González-Martín-Moro J, Sánchez-Burgos R. Osteochondroma of coronoid process: a rare etiology of jacob disease. Craniomaxillofac Trauma Reconstr. 2014;7(4):306-9. https://doi.org/10.1055/s-0034-1378182
15. Thota G, Cillo JE, Krajekian J, Dattilo DJ. Bilateral pseudojoints of the coronoid process (Jacob disease): report of a case and review of the literature. J Oral Maxillofac Surg. 2009;67(11):2521-4. https://doi.org/10.1016/j.joms.2009.04.129
16. Fernández Ferro M, Fernández Sanromán J, Sandoval Gutierrez J, Costas López A, López de Sánchez A, Etayo Pérez 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(9):E595-8.

Received on: 8/8/2019
Final version resubmitted on: 11/4/2020
Approved on: 27/4/2020