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# Joint Formation Between an Osteochondroma of the Coronoid Process and the Zygomatic Arch (Jacob Disease): Report of Case and Review of Literature

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Enlargement of the coronoid process of the mandible was first described by Langenbeck in 1853,[1] and joint formation between the coronoid process and the zygoma was first described by Jacob in 1899.[2] Subsequently, enlargement of the coronoid process has been sporadically reported in the literature.[1,3-13]

This condition can be unilateral or bilateral. The latter is more frequent in young men, resembles the normal coronoid in shape, and is self-limited in growth.[11] The unilateral form usually grows progressively to form a mushroom-shaped enlargement of the process. Besides, in Jacob disease, a new joint forms between the coronoid process and the zygoma. The most consistent clinical feature of this condition is reduction of mouth opening. Treatment consists of coronoid resection through an intraoral or extraoral approach. Histologically, most of the lesions show a bony growth capped by cartilage. Numerous factors have been suggested in the pathogenesis of coronoid process enlargement,[3,4,6,8-10,14] but nothing has been suggested regarding the pathogenesis of the new joint.

Because of the history, which includes an insidious clinical onset, this condition has often been overlooked and treated initially as a temporomandibular joint (TMJ) disorder. We report a case of Jacob disease that illustrates the importance of a proper differential diagnosis when faced with a patient having restricted mouth opening.

## Review of the Literature

Only 8 reported cases of Jacob disease were found (Table 1). There was one in the English literature,[15] 5 in the French literature (including the original description),[2,16-19] 1 in the Belgian literature,[14] and 1 in the Czeck literature.[20] Of the 8 previous cases, there were 7 in males and 1 in a female, with a mean age of 28 years and a range of 13 to 62 years. Three of the cases were bilateral. All of them except the first were treated surgically by unilateral or bilateral coronoidectomy. One case[15] was approached extraorally (no more details given); the others were treated by an intraoral approach. Only 1 case was treated under regional anesthesia.[16]

## Report of Case

A 22-year-old man was referred to our department with a history of limitation of mouth opening that began 2 years before and was initially diagnosed by a dentist as a left TMJ disorder. The patient underwent 6 months of bite appliance

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**SUMMARY OF REPORTED CASES OF JACOB DISEASE**

| Case | Author | Year | Age (yr) | Treatment | Comments |
|---|---|---|---|---|---|
| 1 | Jacob[2] | 1899 | 62 | None | Postmortem finding |
| 2 | Hallam[15] | 1947 | 18 | Extraoral coronoidectomy | |
| 3 | Ginestet et al[18] | 1956 | 19 | Intraoral bilateral coronoidectomy | |
| 4 | Chemin et al[16] | 1958 | 20 | Intraoral surgery | Regional anesthesia |
| 5 | Van de Vijver[14] | 1962 | 18 | Intraoral bilateral coronoidectomy | Previous surgery of the TMJ |
| 6 | Dechaume et al[17] | 1964 | 13 | Intraoral coronoidectomy | |
| 7 | Goudot et al[19] | 1989 | 45 | Intraoral bilateral coronoidectomy | Unilateral |
| 8 | Rames and Orban[20] | 1990 | 36 | Intraoral bilateral coronoidectomy | |
| 9 | Hernández-Alfaro et al | 1996 | 22 | Coronal approach | Previous arthroscopy of the TMJ |

therapy, and subsequently 5 arthroscopic procedures were performed without any improvement in mouth opening. Finally the patient was referred with a diagnosis of left TMJ ankylosis for open TMJ surgery.

When first seen (Fig 1), the interincisal opening was 21 mm, with 4-mm deviation to the left. There was no pain or muscle tenderness. A slight facial assymetry was present because of the mandibular deviation on opening and to a discrete bulging in the left zygomatic region. On palpation, this area was nontender and moved slightly on attempted maximal opening. Oral examination indicated a class II malocclusion with severe crowding of the upper and lower anterior teeth. Protrusion was 5 mm, with deviation to the left. This finding was thought to be significant because protrusion is not possible with intra-articular TMJ ankylosis, but with extra-articular ankylosis the patient may protrude slightly. Left lateral excursion was 6 mm, and right lateral excursion was 3 mm. A panoramic radiograph disclosed an atrophic left condyle. Also, an enlarged and distorted left coronoid process was seen. A 3D computed tomography (CT) scan confirmed the panoramic radiographic findings and showed a mushroom-shaped left coronoid process extending superiorly and laterally, with impingement on the temporal surface of the zygoma and zygomatic arch. The left condyle appeared distorted and anteriorly displaced in the glenoid fossa, resembling one that had sustained a fracture. A diagnosis of benign bony enlargement of the left coronoid process was made (Fig 2A, B).

The patient was admitted to the hospital and, after blind awake nasoendotracheal intubation and general anesthesia had been accomplished, a coronal flap provided easy access to the left temporal fossa and TMJ. After releasing the insertion of the temporalis muscle, a fibrous pseudocapsule was found surrounding both the zygomatic arch and the hyperplastic coronoid process (Fig 3). To allow for easier removal of the mass, a temporary zygomatic arch ostectomy was made. The arch appeared thin, and there was a depression lined with a layer of cartilaginous tissue in the medial aspect. Fibrous bands surrounded the cavity (Fig 4). A low coronoidectomy was performed with an oscillating

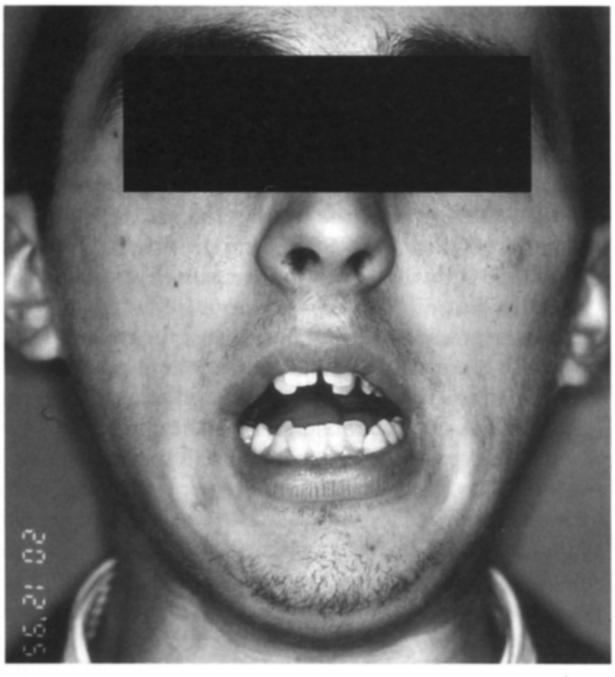

**FIGURE 1.** View of patient showing restricted mouth opening, with slight deviation of the mandible to the left. Note bulging in the left zygomatic region.

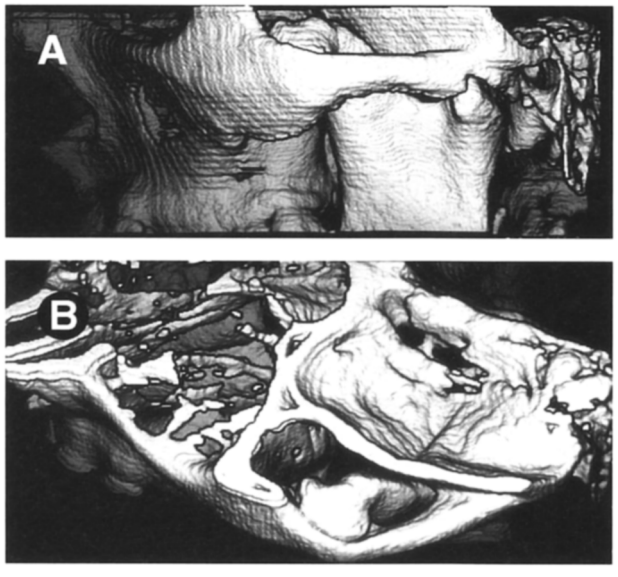


**FIGURE 2.** *A*, Lateral and *B*, axial 3D CT scan showing the relationship between the neoplastic growth and the inner aspect of the zygomatic arch.

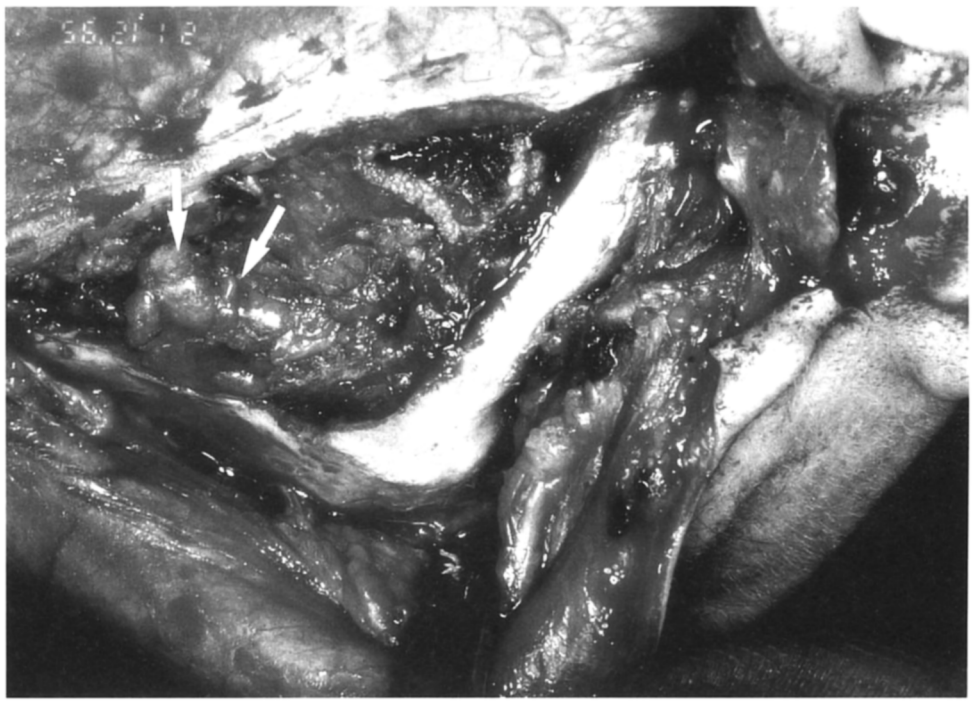

**FIGURE 3.** Intraoperative view after the coronal approach showing distortion of the arch and the fibromuscular tissue covering the mass (*arrows*).

saw. The process was then removed after releasing some fibrous insertions. This immediately allowed a 52-mm interincisal opening. The zygomatic arch was repositioned with 2 miniplates after removal of the fibrocartilaginous tissue and smoothing of the remaining irregular bony surface.

The TMJ was then investigated through an incision made in the capsule. The disc appeared distorted and perforated. Thus, a discectomy was done followed by an interpositional pedicled flap of temporalis muscle and fascia (Fig 5). A drain was inserted, and the flap was closed. Maximal interincisal opening was maintained with a rubber wedge left in place for 24 hours. Recovery after surgery was uneventful, and the patient was discharged 48 hours later. Thereafter, jaw stretching exercises maintained a stable interincisal opening of 47 mm 6 months postoperatively (Fig 6).

The coronoid specimen resembled a mandibular condyle

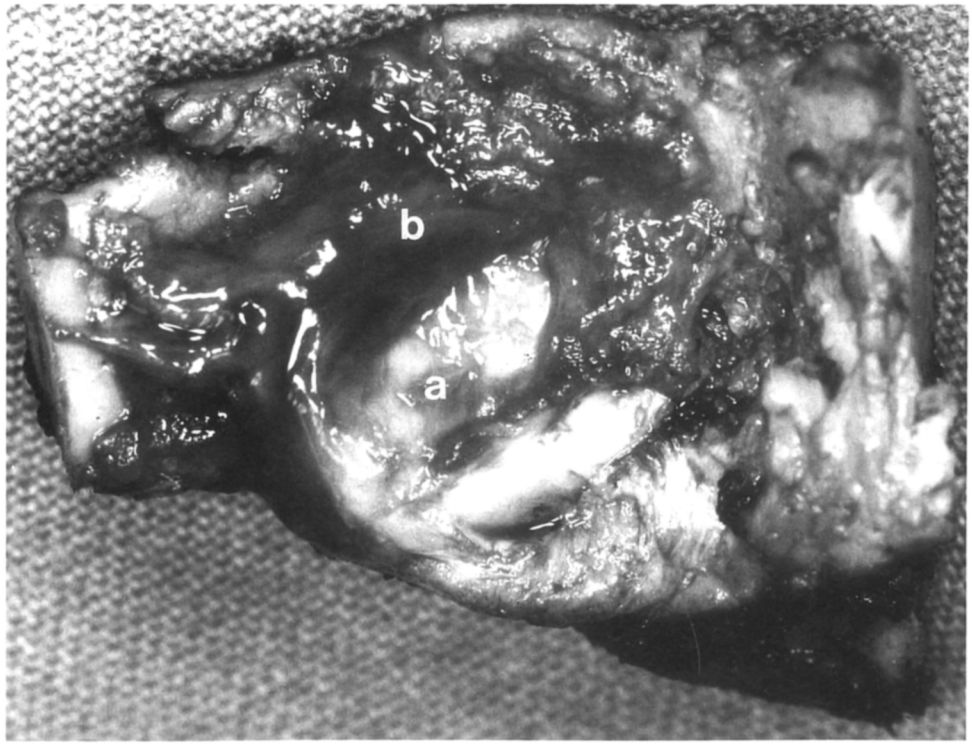


**FIGURE 4.** View of gross specimen. Note the well-delineated cavity lined with cartilaginous tissue (*a*) and surrounded by synovial remnants (*b*) on the inner aspect of the arch.

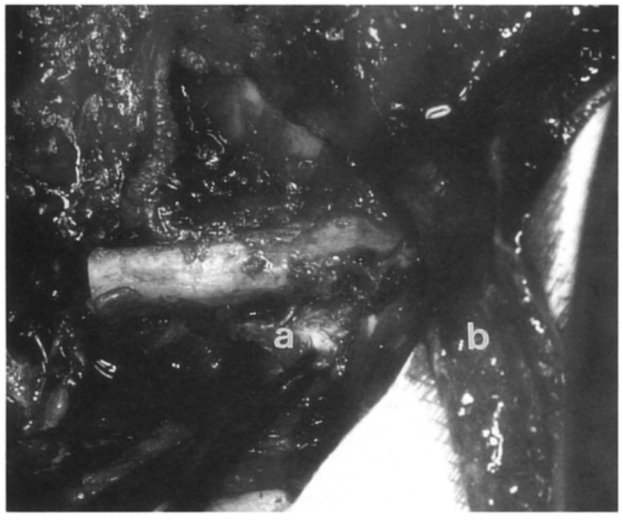


**FIGURE 5.** View of the left TMJ. Note the distorted fibrous disc (*a*) and temporalis muscle and fascia flap for reconstruction (*b*).

with fibromuscular insertions (Fig 7). Microscopically, the sections showed fibrous, cartilaginous, and bony elements irregularly arranged. A diagnosis of osteochondroma was made (Figs 8, 9). The cartilage lining the cavity of the zygomatic arch was disorganized and uncalcified. Synovial tissue was attached to both the hyperplastic coronoid process and the zygoma.

## Discussion

Symptomatic enlargement of the coronoid process is a rare condition. Since the first reported case by Langenbeck,[1] much confusion has existed regarding the nature and pathogenesis of this condition. Although there are not enough epidemiologic data regarding the prevalence of this process, asymptomatic cases are probably more frequent than previously thought.[6] Hönig et al[21] examined the panoramic radiographs of a randomly selected sample of 2,000 patients and found a prevalence of 0.5%. A much lower prevalence of the Jacob disease should be expected.

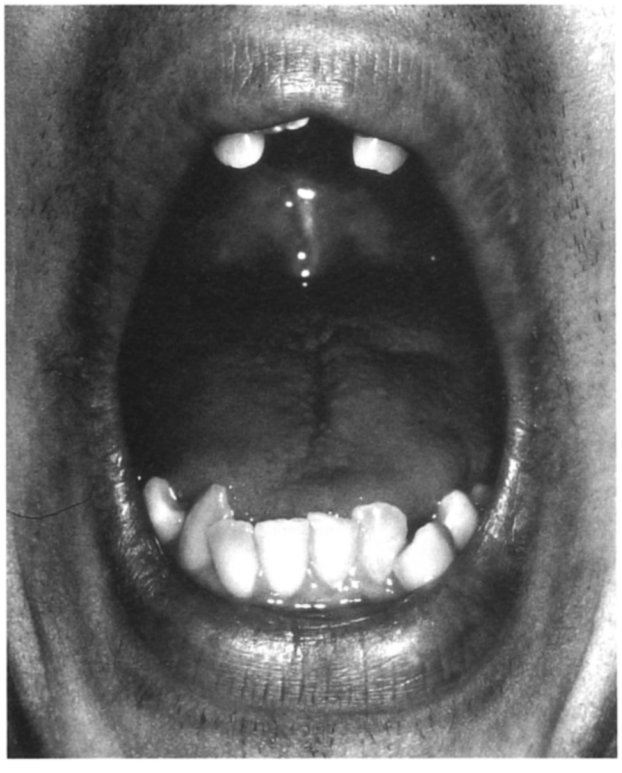

**FIGURE 6.** Normal opening 6 months after surgery.

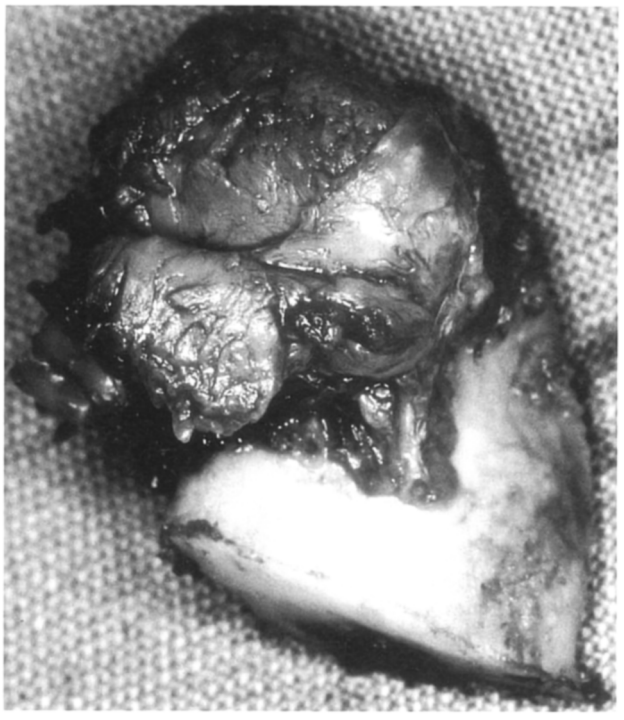

**FIGURE 7.** View of resected mushroom-shaped coronoid specimen.

Some have advocated trauma as a possible causative event in the development of the hyperplasia. The influence of functional alterations in the shape and structure of the coronoid process has been proposed by others.[7,9] Isberg et al[6] pointed out that hyperactivity of the temporalis muscle, which is often present together with internal derangements of the TMJ, is likely to promote coronoid hyperplasia through a reactive process in response to pull of the tendon. Nothing has been suggested regarding the pathogenesis of the new joint, and it is still a subject of discussion whether the Jacob disease is a particular variety of coronoid process hyperplasia or a completely different clinical entity.

This case was initially diagnosed as TMJ dysfunction and managed as such. However, several panoramic radiographs obtained at the onset of symptoms already showed coronoid enlargement. Therefore, in this case, TMJ dysfunction was most probably secondary to the surgical manipulation involved in the multiple arthroscopic procedures and to lack of function for several years.

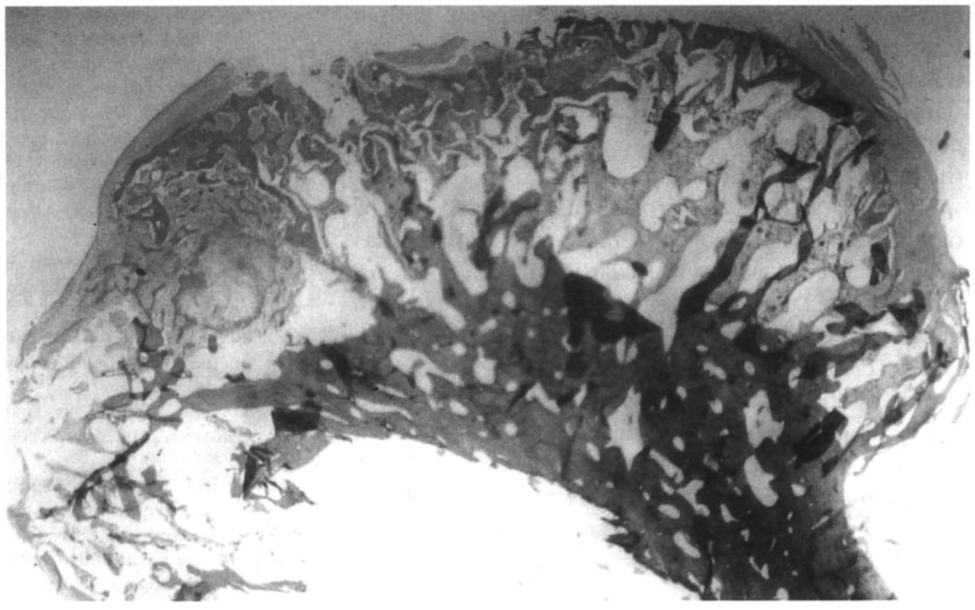

**FIGURE 8.** Low-power photomicrograph of the specimen (hematoxylin and eosin, original magnification ×16).

Diagnosis of this entity can be made easily from a panoramic radiograph and careful clinical examination. Although a Waters' radiograph is very useful in showing the coronoid hyperplasia, and its relation with the zygoma,[22] we found, as other authors did previously,[19,23] that 3D CT imaging is essential to complete the diagnosis and especially to plan the surgery. In this case, such imaging helped in deciding the surgical approach and confirmed the disturbed condition of the homolateral condyle.

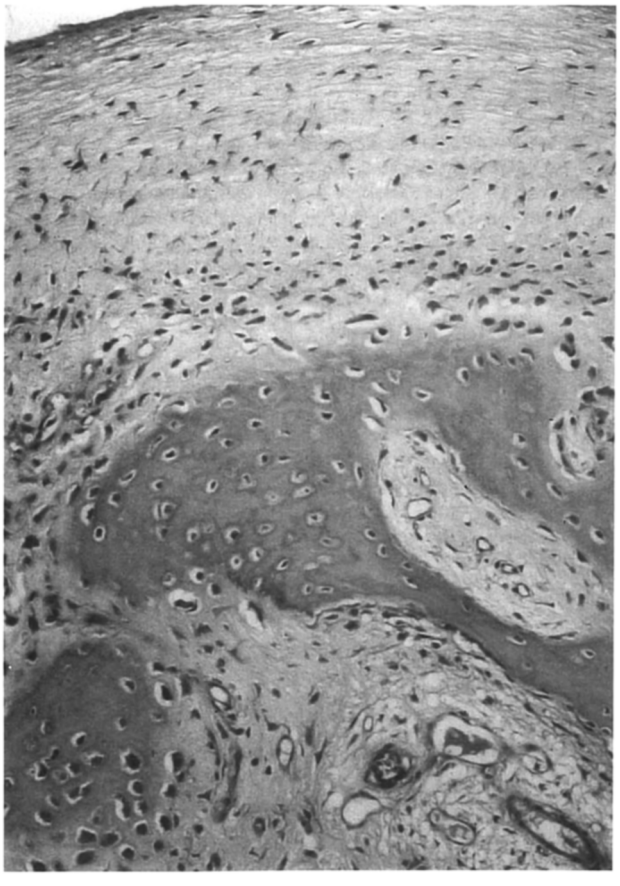

**FIGURE 9.** High-power microphotographs (hematoxylin and eosin, original magnification ×40) showing detail of Fig 8. Note the hyaline cartilage and the underlying bone with irregular trabeculae.

Different approaches have been advocated to treat this condition. Most of the previously reported cases of coronoid hyperplasia and Jacob disease had been treated through an intraoral approach, although limitations of this approach are well recognized.[8] Extraoral approaches also have been described. Ostrofsky and Lownie[22] treated 5 of 9 patients through a submandibular approach, advocating that it is safer when the full extent of the problem is unknown. Both the intraoral and the submandibular approach are insufficient in cases in which the coronoid is large enough to be trapped over the arch, as was the situation in this patient. Other reports have proposed a surgical approach directly over the arch.[9] This approach, beside leaving an aesthetically unacceptable scar, risks injury to the upper branches of the facial nerve and should be avoided. The coronal flap, recommended in previous reports,[21] offers an excellent approach to the region, while avoiding visible scars and allowing for complete visualization and treatment of this condition. Also, considering the amount of debridement that has to be done with the fibrous and muscular insertions on the coronoid, we think that 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, the coronal approach allowed removal of the lesion, thorough debridement of the

fibrous adhesions, and TMJ reconstruction with a temporalis flap. Temporary removal of the arch has been shown to facilitate removal of the hyperplasic coronoid process.[17] In this case, it also allowed for complete removal of the adhesions and smoothing of the inner aspect of the zygomatic arch.

Since Shackelford and Brown[9] first reported 2 cases of enlargement of the coronoid process, there has been much confusion with regard to the basic nature of this entity. Differences in the proportion of cartilaginous and bony elements in the specimen have justified several histologic diagnoses, namely, osteochondroma, osteoma, cartilage-capped exostosis, and hyperplasia.[24] Osteochondromas are benign neoplasms developing most frequently between the ages of 10 and 30 years,[25] as in most of the patients with the Jacob disease. They probably arise from the periosteum, which forms areas of metaplastic cartilage.[12] The lesion in this case consisted of a mushroom-shaped process with fibrous, cartilaginous, and bony tissue, and it had the well-described cartilaginous cap.

The first report of this condition by Jacob[2] described involvement of the malar bone and bulging into the temporal fossa, thus reducing the space and allowing premature contact with the hyperplasic coronoid process. Few reports have mentioned impingement of the process on the inner aspect of the zygomatic arch.[24] In this case, impingement on the arch was accompanied by the presence of a concavity covered by cartilage. Both bony sides of the lesion were surrounded by a pseudocapsule consisting of fibrous and synovial tissue.

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