Imaging and Histopathological Features of Jacob's Disease: A Case Study

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Abstract A pathologically elongated coronoid process forming a joint with the zygomatic arch and bone is termed as “Jacob’s disease.” Herein, we present a case of Jacob’s disease, which has rarely been reported in the literature. Moreover, very few cases have been reported from the Indian subcontinent. A 28 years-old male reported to us with progressive restriction of mouth opening. After detailed imaging, coronoidectomy was performed through an intraoral approach. A mushroom-shaped gross specimen was obtained. Osteochondroma of the coronoid process was diagnosed based on the histopathologic examination of the excised tissue. Although Jacob’s disease is seldom considered in the list of differential diagnoses for restricted mouth opening, our case highlights the need for evaluation of the coronoid process, which is usually ignored during routine radiographic examination. The report also highlights the miniscule incidence of this condition in the available literature.

Keywords Jacob’s disease · Coronoid process · Mushroom shape · Osteochondroma · Computed tomography

Jacob’s disease (JD) is characterised by the formation of a new joint between a pathologically elongated coronoid

process and the body of the homolateral malar bone. Hyperplasia of the coronoid process 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 [1]. Since then, there have been sporadic reported cases in the literature [2]. A recent review revealed 35 cases of Jacob’s disease previously published in literature [3]. So far, only two cases of this rare condition have been reported from the Indian population [3, 4]. We report to the readers a rare case of Jacob’s disease with detailed imaging and histopathologic features.

Case Report

History

A 28 years-old male reported to our department with the complaint of restricted mouth opening over 2–3 years. The patient revealed a history of trauma leading to a left clavicular fracture and bruising over the same side of the face, 5 years prior to presentation. During the initial years, the patient reported intermittent left temporomandibular joint pain and clicking, which gradually decreased later, with simultaneous reduction in mouth opening. On clinical examination, mouth opening of 7 mm with left-sided deviation of the mandible was noticed.

Radiographic Features

A diagnostic panoramic radiograph revealed a normal condylar process, body of the mandible, and ramus. The left coronoid process showed a globular projection that had a mixed radiopaque-lucent feature. The mass was

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approximately 3 cm in diameter and had a radiopaque periphery with a radiolucent inner rim (Fig. 1). Water's view revealed a globular radiopaque projection from the left coronoid. Axial computerised tomography (CT) section revealed the presence of a mushroom-shaped radiodense mass at the apex of the coronoid. It also revealed outward bulging of the ipsilateral zygomatic arch (Fig. 2).

Diagnosis and Management

Coronoidectomy was performed via the intraoral approach under general anesthesia. The gross specimen appeared as a mushroom-shaped yellowish-white mass measuring about 3 cm in size (Fig. 3). On histopathologic evaluation, cartilaginous and osteoid tissue with few areas of fibrous tissue were observed. Cartilaginous tissue showed numerous chondrocytes arranged irregularly and separated by minimal matrix (Fig. 4). Based on the histopathologic features and the anatomical location, a final diagnosis of Jacob's disease was made. The patient was then followed up for a period of 1 year with no evidence of recurrence.

Discussion

Jacob's disease is the name given to a pseudojoint formation between the coronoid process and the zygomatic arch due to an osteochondroma of the coronoid process [5]. It accounts for 35.8% of benign bony tumours and 8.5% of bony tumours overall [6]. It can occur singly or as part of an autosomal dominant syndrome called osteochondromatosis [6]. Jacob's disease is most frequent in young patients with a mean age of 27 years (age range: 16–62 years) with male predominance [7]. It is usually unilateral in

Fig. 2 Axial CT section showing outward remodelling of the left zygomatic arch (*encircled in black*)

occurrence with predilection for involvement of the left coronoid process [7]. Bilateral cases are self-limiting but the unilateral cases enlarge progressively to form a mushroom-shaped enlargement of the coronoid process [4].

Various theories have been proposed regarding the etiology of coronoid hyperplasia, including trauma, hyperactivity of the temporalis muscle, dysfunction of the TMJ caused by chronic disc displacement, endocrine stimuli and genetic and familial factors [2, 8, 9]. It has been hypothesized that the enlarging osteochondroma compresses against the zygoma and either resorbs or remodels it

Fig. 1 Panoramic view showing radiopaque mass superior to the left coronoid process (*encircled in black*)

Fig. 3 The mushroom-shaped gross surgical specimen

Fig. 4 Pictomicrograph ($\times 10$, H&E) showing features of osteochondroma

causing a pseudo joint [4]. Progressive limitation of mouth opening is the most commonly reported symptom [3, 7, 8]. Deviation of the mandible to the affected side and facial asymmetry are other notable features that are frequently reported [3]. History of pain has been rarely reported in the available literature [2, 5], and the disease was often mistakenly treated as a temporomandibular joint dysfunction which lead to considerable delay in the diagnosis and treatment of this condition [5, 7, 10]. Panoramic radiography and paranasal sinus views have been conventionally used for the diagnosis of Jacob's disease. Recently, CT has been a beneficial imaging modality [10, 11].

Coronoidectomy is the treatment of choice with either an extraoral or intraoral approach. The former carries the risk of facial nerve damage; hence, the latter is preferred although accessibility is limited [5, 12]. Histopathologic features show differences in the proportion of cartilaginous

and bony elements in the specimen that lead to a spectrum of differential diagnoses, namely osteochondroma, osteoma, cartilage capped exostosis, or hyperplasia [2, 13].

Prognosis depends on the histopathologic nature of the lesion. It has been observed that the incidence of sarcomatous change is as low as 1% [11]. However, in conditions like osteochondromatosis, sarcomatous transformation can be as high as 11% [5, 11]. Our report is intended to highlight the classical clinical, radiological, and histopathologic features of a rarely reported condition.

References

1. Jacob O. Une cause rare de constriction permanente des machoires. Bull et Mem De la Societe Anatomique De Paris. 1899;1:917–9.
2. Coll-Anglada M, Acero-Sanz J, Vila-Masana I, Navarro-Cuellar C, Ochandiano-Caycoia S, de Atalaya J L, Navarro-Vila C. Jacob's disease secondary to coronoid process osteochondroma. A case report. Med Oral Patol Oral Cir Bucal. 2011;16(6):708–10.
3. Sreeramaneeni SK, Chakravarthi PS, Prasad LK, Satish PR, Beeram RK. Jacob's disease: report of a rare case and literature review. Int J Oral Maxillofac Surg. 2011;40:753–67.
4. Roychoudhury A, Gupta YK, Parkash H, Karak AK. Jacob disease: report of a case and review of the literature. J Oral Maxillofac Surg. 2002;60:699–703.
5. 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;67:2521–4.
6. Vezeau PJ, Fridrich KL, Vincent SD. Osteochondroma of the mandibular condyle: literature review and report of two atypical cases. J Oral Maxillofac Surg. 1995;53:954–63.
7. Capote A, Rodriguez FJ, Blasco A, Munoz MF. Jacob's disease associated with temporomandibular joint dysfunction: a case report. Med Oral Patol Oral Cir Bucal. 2005;10:210–4.
8. Ferro MF, Sanroman JF, Gutierrez JS, Lopez AC, de Sanchez AL, Perez AE. 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.
9. Isberg A, Isacson G, Nah KS. Mandibular coronoid process locking: a prospective study of frequency and association with internal derangement of the temporomandibular joint. Oral Surg Oral Med Oral Pathol. 1987;63:275–9.
10. Akan H, Mehreliyeva N. The value of three dimensional computed tomography in diagnosis and management of Jacob's disease. Dentomaxillofac Radiol. 2006;35:55–9.
11. Emekli U, Aslan A, Onel D, Cizmeci O, Demiryont M. Osteochondroma of the coronoid process (Jacob's disease). J Oral Maxillofac Surg. 2002;60:1354–6.
12. Kerscher A, Piette E, Tideman H, Wu PC. Osteochondroma of the coronoid process of the mandible. Report of a case and review of the literature. Oral Surg Oral Med Oral Pathol. 1993;75: 559–64.
13. Hernandez-Alfaro F, Escuder O, Marco V. Joint formation between an osteochondroma of the coronoid process and the zygomatic arch (Jacob disease): report of a case and review of literature. J Oral Maxillofac Surg. 2000;58:227–32.