OSTEOCHONDROMATOSIS AND THE CORONOID- ZYGOMATIC PSEUDOJOINT (JACOB’S DISEASE) AS AN UNUSUAL CAUSE OF MIDFACIAL ASYMMETRY AND JAW RESTRICTION

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ABSTRACT

A young male presenting with a history of progressive midfacial symmetry and limitation of mouth opening was found to have a unilateral coronoid hyperplasia forming an osteochondromatous pseudojoint with the expanded zygomatic arch (Jacob’s disease). This paper reviews the differential diagnoses of problems, especially in developing countries, that can cause restricted mouth opening, highlighting problems with conventional radiography and the role of CT assessment in the diagnosis of this rare condition.

Key Words: Jacob’s disease; pseudojoint; osteochondroma; coronoid enlargement; restricted opening; facial deformity.

INTRODUCTION

Jacob’s disease is an extremely rare condition characterized by osteochondroma caused coronoid hyperplasia and the formation of a pseudojoint between the hyperplastic coronoid and the inner aspect of the proportionally enlarged zygomatic arch. It is reported most frequently in young patients with a mean age of 27 a male dominance and a predilection for the left side. The large coronoid process exerts pressure that results in restricted jaw movements, mid-face deformity and possible malocclusion. Hypertrophy or fibrosis of the affected masticatory muscles may worsen the symptoms. While hyperplasia of the coronoid can occur bilaterally, in unilateral cases patients may present with facial asymmetry. Since this is a rare and slowly progressive disorder that can mimic a wide range of problems including the common TMJ dysfunction and may not clearly be visible on conventional radiographs, an early diagnosis can be missed resulting in greater morbidity.

CASE REPORT

A 15 years old male presented with a history of painless left sided facial asymmetry and progressive limitation of mouth opening increasing over the past year. A bony hard, non tender, asymmetric enlargement at the left zygomatic and mid-face region was obvious and the laterally flaring zygomatic arch curve was distinctly palpable. Fig. 1 Mouth opening was 22mm with left deviation. He had no significant medical history and neither he nor his parents recalled any specific early trauma to the site. He had no oral habits that could cause restricted opening. Other parameters were normal for his age. Intra-oral examination revealed no abnormality other than a more prominent left anterior ramus and the wider curve of the left arch in the upper buccal sulcus. There was no clear evidence of TMJ Dysfunction, one presumptive diagnosis.

Due to superimposition of bone shadows in the region the panoramic radiograph (OPG) Fig. 2 was not very helpful. The occipito-mental radiograph showed a larger left zygomatic arch curve and either a considerably enlarged left coronoid or ossification in the attached temporalis tendon but was unclear about its direction of growth. Fig. 3 In both radiographs both condyles were normal and there was no other bone pathology. A tentative diagnosis of coronoid related extra-articular ankylosis was made.
Axial and coronal computerized tomographic (CT) scans showed the joint like relationship of the enlarged coronoid against the medial cortex of the left zygoma with enlargement and remodeling of the bone and arch. Fig. 4 (a and b)

Fig 1: (a) Frontal and (b) superior views of patient showing left sided zygomatic enlargement.

Fig 2: OPG showing the enlarged left coronoid with possible bulbous tip (arrow)

Fig 3: Occipito-mental radiograph showing the enlarged left coronoid process. Compare the size differences (arrowed).

Fig 4: (a) axial and (b) coronal CT scan views showing the bulbous enlargement of the left coronoid and asymmetric expansion in the zygomatic region to accommodate the pseudojoint. A soft tissue 'capsule' like arrangement can be seen enveloping the 'joint'.

Fig 5: Resected left coronoid with laterally curved and smooth "articular" head. Note the remnants of the temporalis tendon attachment lower down the bone.
On 6 July 2006 the left pseudojoint coronoid was ‘disarticulated’ and removed intraorally through a superiorly extended ramus incision and dissecting through the enveloping muscles at the level of the zygomatic arch. The laterally flaring upper tip ended in a smooth, bulbous joint surface. Fig. 5 The wound was sutured over a drain tube left for 24 hours. Histopathology confirmed a fibrous tissue capped osteochondroma at the tip of the hyperplastic, curved, coronoid.

Assisted by vigorous exercise, post operative trismus settled over the next few weeks. Mouth opening was 41 mm four months later. The parents were advised that in view of his youth the zygomatic enlargement would remodel over the next few years. Any residual asymmetry could be addressed then. His face had improved considerably over 18 months review.

DISCUSSION

Osteochondromas are among the most common benign tumors of the skeleton but are rarely found in the craniofacial bones. In this situation the majority have arisen as male predominant, unilateral or less frequently bilateral, growths in the coronoids or coronoid process or with occasional reports in other sites.\(^1\,\text{2,3,4}\) They most commonly come to attention as a restriction in opening, or occlusal disturbances or as a progressive facial deformity.\(^4\,\text{5,6}\)

Coronoid hyperplasia was originally described by von Langenbeck in 1853. The hyperplastic coronoid process can impinge against the zygomatic arch restricting jaw movements.\(^8\) The formation of a pseudojoint between the enlarged coronoid process and the inner aspect of the zygomatic arch is an even rarer occurrence that was first described by anatomist Oscar Jacob in 1899.\(^1\,\text{2,9}\) The pathogenesis of the disease is still not clear although numerous factors have been suggested. In addition to excessive temporalis muscle pull and chronic TMJ joint dysfunction, endocrine stimulation, trauma, genetic and familial predisposition are possible etiological factors.\(^2\,\text{3}\)

Apart from local, relatively transient, masticator inflammatory disease and the fairly common problems of TMJ dysfunction,\(^2\) the differential diagnosis in patients with chronic restricted mouth opening includes muscle diseases, oro-pharyngeal neoplasms and coronoid hypoplasia.\(^2\) Rarely, post traumatic ankylosis can occur between the zygomatic arch and coronoid.\(^10\) In the Asian region this is not uncommon while Oral Submucous Fibrosis (OSF),\(^11\) and articular TMJ Ankylosis,\(^12\) are extremely important and fairly common causes of restricted mouth opening, especially in the younger age group. Also in this age group the painless slowly growing bony swelling and deformity may be an indicator of localized fibro-osseous disease which has a predilection for the maxilla / zygomatic region.

Panoramic radiography and Water’s occipito-mental view maybe the first steps in radiographic examination but the enlarged and deformed coronoid can be missed because of overlaps and the possible tendency to focus on the condyle as the cause of restriction. A joint like close relationship between the hyperplastic coronoid process and the posterior wall of the maxilla or zygomatic arch must be seen to establish the diagnosis of Jacob’s disease.\(^2\) Because of multiple overlaps conventional radiography may be inadequate for this purpose.

CT has an important role in diagnosis and is useful for adequate surgical planning by allowing assessment of the size of impingement by the coronoid processes.\(^2\) The protrusion of the hypertrophic segment into the temporal fossa and its articulation with the inner aspect of zygomatic arch can be clearly seen. Fig.3 3D CT may show the elongated coronoid process passing above the zygomatic arch and the joint formation. Joint formation may occur in two different models: (1) impingement of the enlarged and deformed coronoid process on a 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 the surgical approach.\(^2\) In our case, CT images suggested the first sequence.

Access to the coronoid process is usually intra-oral since this approach avoids external scars and, even in the mouth with limited opening, is usually possible. Submandibular ramus and coronal approaches,\(^3\) to
the zygomatic arch have been proposed as has a transzygomatic access. With a grossly enlarged and deformed coronoid/zygoma inaccessibility of the lesion may dictate an external approach.

CONCLUSION

In patients with limited mouth opening and malocclusion TMJ related pathologies tend to be considered first. Coronoid hyperplasia and the rarer possibility of Jacob’s disease should be considered in the differential diagnosis of limited opening and progressive malocclusion or facial asymmetry. CT is invaluable both in diagnosis and surgical planning.

REFERENCES


