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Osteochondroma of the coronoid process (Jacob's disease): an unusual cause of restricted jaw motion

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Abstract Osteochondromas are the most common benign bone tumor, most commonly found in the ends of long bones; however, they rarely involve facial bones, particularly the mandible. Osteochondromas involving the coronoid process have rarely been reported in the literature but pose a diagnostic dilemma. When large enough, osteochondromas of the mandibular coronoid process can form a joint with the zygomatic arch (Jacob's disease). This pseudoarticulation results in restricted jaw motion, which can clinically be mistaken for temporomandibular joint dysfunction. We report a case of a 39-year-old man with chronic restricted jaw motion undiagnosed for several years.

1. Introduction

Osteochondroma, or osteocartilaginous exostosis, is a cartilage-capped exophytic lesion that arises from the cortex of a bone. It constitutes 20% to 50% of all benign tumors and 10% to 15% of all bone tumors [1].

Although osteochondroma is considered the most common tumor of skeletal bones, it is relatively uncommon in the jaw. The rare osteochondroma of the mandible occurs at the condyle or the tip of the coronoid process. This cartilage-capped growth accounts for 35.8% of benign bony tumors and for 8.5% of bony tumors overall [2].

Enlargement of the coronoid process of the mandible was first described by Langenbeck in 1853, and joint formation between the coronoid process and the zygoma was first described by Jacob in 1899 [3-5]. The most consistent clinical feature of this condition is reduction in mouth opening.

A review of the literature reveals only 34 histologically proven cases of osteochondroma of the coronoid process of the mandible. The disease appears to involve males (73.5%) more often than females, with a mean age of 35 years [6]. The treatment is surgical, with an intraoral approach being the most preferred among most reported cases. None of the reported cases showed a recurrence [7].

2. Case report

A 39-year-old white man complained of progressive limitation in mouth opening for approximately 3 years. The patient was treated for temporomandibular joint (TMJ) dysfunction, without improvement. Physical examination revealed no palpable mandibular mass or facial asymmetry. For several years, the patient had undergone several magnetic resonance imaging (MRI) of the TMJ, revealing degenerative changes of the articular disk bilaterally, without evidence of abnormal displacement.

After clinical complaints of maxillary sinus pressure and suspected sinus disease, computed tomography (CT) examination of the paranasal sinuses demonstrated a large mushroom-shaped exostosis arising from the left coronoid process. The lateral wall of the maxillary sinus was deformed and showed a medially oriented concavity (Fig. 1). There was no significant anterior bowing of the zygoma, thus, resulting in no gross facial asymmetry. There was no evidence of bony destruction or erosion. The sagittal reformatted image demonstrated the relationship between the tumor and the zygomatic arch, resulting in a pseudoarticulation (Fig. 2).

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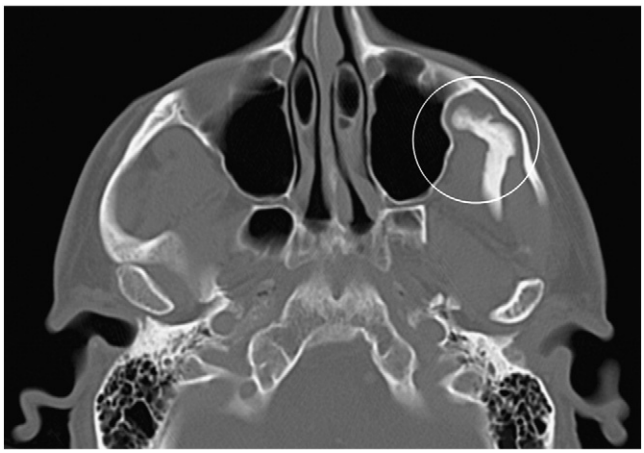

Fig. 1. An axial CT scan performed at the level of the zygoma demonstrates a large exophytic mass arising from the coronoid process of the left mandible (circle). The mass results in smooth, inward deviation of the lateral wall of the maxillary sinus, without evidence of erosion or destruction.

A preoperative radiologic diagnosis of coronoid osteochondroma was established, based on imaging characteristics. During the following month, the patient was referred to a head and neck surgeon. Using an intraoral approach, we endoscopically excised the coronoid mass and removed it from the masticator space.

Pathology revealed an osteochondroma, measuring 2.0 × 1.0 × 0.8 cm, with a cartilaginous cap measuring 0.3 cm in maximal thickness. Shortly after surgery, the patient reported much improved jaw motion and opening.

3. Discussion

Although osteochondroma is the most common tumor of skeletal bones, it is relatively uncommon in the jaw, occurring at the condyle or the tip of the coronoid process. This benign cartilage-capped growth is usually discovered incidentally on radiographic examination or on palpation of a protruding mass in the affected area [2]. Initial signs and symptoms include tightness within the joint area and gradual reduction in mouth opening. The slow development of trismus and painless facial mass typify the later stages [6].

Coronoid lesions of the mandible histologically confirmed as osteochondroma have been reported infrequently. A review of the literature reveals only 34 histologically proven cases [8].

Jacob's disease is a rare condition that affects the coronoid process; it was first described in 1899. Jacob's disease involves the development of an osteochondroma of the coronoid process and leads to the formation of a pseudo joint between the zygoma and the coronoid process. The zygoma remodels, forming a pseudo joint that results in decreased mouth opening. The growth enlarges to push out the cortices of the zygoma, thus, accounting for a bulge that may be seen at the malar prominence [6].

Because of its insidious clinical onset, Jacob's disease is often overlooked and misdiagnosed as a TMJ disorder [6]. Magnetic resonance examination of the TMJ is usually the imaging method chosen in patients with such symptoms. However, the coronoid processes are not evaluated because they are not included in the field of view in MRI of TMJ. For that reason, these patients may be treated for a misdiagnosis of TMJ disorders [9].

In addition to plain film radiographs, CT has become the standard in preoperative assessment and surgical planning [7]. To establish the diagnosis of Jacob's disease, we need to show a direct contact between hyperplastic coronoid process and posterior wall of maxilla or zygomatic arch and joint surfaces at this location [9]. Three-dimensional reformatted images also allow for the assessment of the length of the coronoid processes and changes on the inner aspect of the zygomatic arch.

The differential diagnosis for unilateral coronoid process enlargement includes relatively rare clinicopathologic entities. Endocrine stimulation, increased TMJ activity, trauma, and genetic and familial predisposition are mentioned as etiologic factors for coronoid hyperplasia [9]. Although rare, peripheral chondrosarcoma can arise from a sessile osteochondroma, especially if the cartilaginous cap, accurately demonstrated on MRI, exceeds 2 cm [2].

Osteochondroma of the mandibular condyle is also rare. Patients usually present with TMJ dysfunction and a palpable mass, with facial asymmetry [1]. Thirty-eight cases of osteochondroma of the mandibular condyle have been described in the English literature [10].

Different surgical approaches have been advocated to treat this Jacob's disease. Most of the previously reported cases of coronoid hyperplasia and Jacob's disease had been treated through an intraoral approach [3]. In the intraoral approach, an incision is made along the anterior border of the

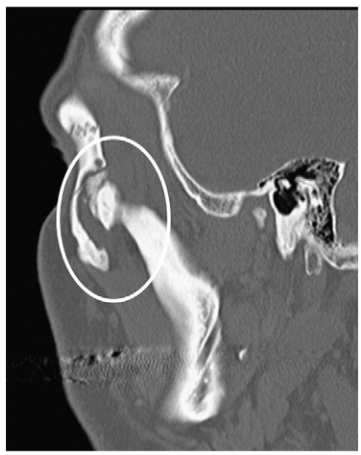

Fig. 2. A sagittal reformatted image demonstrates the articulation (pseudoarthrosis) formed between the zygoma and the sessile mass, arising from the coronoid process (circle). This pseudo "ball in socket" joint resulted in severe restriction in jaw motion.

ramus of the mandible. The periosteum is elevated, and the temporalis muscle is bluntly dissected from the coronoid process to provide exposure [8].

Until 1961, nearly all reported tumors were resected via a zygomaticofacial approach, with or without temporary severing of the zygomatic arch. The intraoral approach, first described in 1958 by Antoni, is safer and generally allows complete removal of the tumor [7].

We consider the intraoral approach of the coronoid process of the mandible as a safe and adequate treatment of osteochondroma in this area. None of the reported cases showed a recurrence after several years of follow-up.

4. Conclusion

An osteochondroma of the coronoid process can pose a diagnostic dilemma. Jacob's disease is defined as a condition consisting of new joint formation between the hyperplastic coronoid process of the mandible and the inner aspect of the zygomatic arch.

Because of the history, as in this case, which includes an insidious clinical onset, this condition has often been overlooked and treated initially as a TMJ disorder. CT has an important role in diagnosis and is useful for an adequate surgical planning.

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