CASE REPORT

Aplasia of the mandibular condyle associated with some orthopaedic abnormalities: a case report

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A rare case of aplasia of mandibular condyle associated with some other orthopaedic problems is presented. A 5-year-old boy attended our clinic with a chief complaint of facial asymmetry and chewing difficulty. The mandible was deviated to the left. The occlusion also showed a deflection to the left of the mandibular midline. He also had walking difficulty owing to a hip abnormality. Panoramic radiographic examination of the patient revealed that the left mandibular condyl was totally absent. The right condyle was unremarkable. His history revealed neither trauma nor any significant disease. Aplasia is a rare anomaly and means the insufficient development of the mandibular condyle. True agnesis of the mandibular condyle is extremely rare. Association of the manifestations of the patient with some orthopaedic problems makes this case interesting.

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Introduction

A 5-year-old boy presented to the Oral Diagnosis and Radiology Department at Ondokuz Mayıs University with a chief complaint of evident facial asymmetry and chewing difficulty (Figure 1).

The patient and his twin brother were born prematurely because their mother developed severe pre-eclampsia. The proband underwent treatment for a long period of time for septic arthritis on his left elbow joint, which was first detected at 1 month of age. Additionally, walking difficulty was detected when he was 22 months old and the family attended the orthopaedic clinic for this problem. Bilateral developmental hip dysplasia and bilateral aplasia of the collum femoris and caput femoris were diagnosed, but treatment was postponed until the completion of his development.

The patient's mother and father were normal; no consanguinity or blood incompatibility was reported between them. In addition, his family history was negative for craniofacial abnormalities and his twin brother was normal.

Extraoral examination revealed that the left side of the patient's face was hypoplastic and the pogonion was deviated to the left. The mandible was observed as deviating to the left when he opened his mouth. The left mandibular condyle was not palpable. The rest of the face showed no observable anomalies, and there were no defects of the auricles.

In occlusal examination, approximately 1 cm of deflection to the left side of the mandible was detected (Figure 2). Maximum mouth opening was within normal limits (approximately 4 cm). Both overjet and overbite were normal. His tooth development and tooth count were also normal.

Panoramic examination revealed that the left mandibular condyle was totally absent, and the ramus terminated relatively flatly at the level of the sigmoid notch (Figure 3). In three-dimensional (3D) CT reconstruction, flattening of the articular fossa and aplasia of the mandibular condyle were detected on the left side of the mandible. The right condyle was unremarkable (Figure 4). Anteroposterior skull radiography showed facial asymmetry (Figure 5), and lateral and anteroposterior cervical radiographs were also normal.

A pelvic radiograph revealed bilateral developmental hip hypoplasia, which was marked on the left

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Figure 1 Frontal view of the patient reveals that the left side of the face is hypoplastic and the pogonion deviates to the left

side. Bilateral aplasia of the collum femoris and caput femoris was also present. Additionally, the tuberculum majus were not detected bilaterally and the fossa acetabulum were indistinct. At this area, cortical and periosteal irregularities were noted. At the vertebral column, there was an arcus defect and spina bifida at sacral 1 (Figure 6).

A radiograph of the right arm showed lytic defects at the ulnar medial condyle and cortical irregularities at other joint surfaces because of septic arthritis (Figure 7).

Figure 2 In intraoral examination, approximately 1 cm deflection to the left side of the mandible is clearly remarkable. Maximum mouth opening is within normal limits (approximately 4 cm)

Discussion

Developmental anomalies of the mandibular condyle are generally classified in terms of aplasia, hypoplasia, hyperplasia and bifidity. Aplasia means the insufficient development of the mandibular condyle.¹⁻⁵

Mechanical trauma during active growth, inflammatory conditions like juvenile arthritis (JRA) and radiotherapy may be counted among the most common causes of condyle anomalies. In the case of trauma, it has to have happened before the age of 2 years.⁶ Parathyroid hormone-related protein deficiency also affects bone formation and chondrocyte differentiation, leading to condyle malformation.⁷ Some authors further affirm

Figure 3 Panoramic imaging reveals that while the left mandibular condyle is totally absent and the ramus is terminated relatively flat at the level of sigmoid notch at that side, the right condyle is unremarkable. Also, tooth development and tooth count is normal

Figure 4 In a CT image, flattening of the articular fossa and complete absence of the left mandibular condyle are detected on the left side of the mandible

that mandibular deficiency can occur without any defined aetiology.⁸ Our patient had no history of trauma.

Condylar aplasia may be present with some craniofacial syndromes such as hemifacial microsomia, Goldenhar, Treacher Collins, Hallerman–Streif, Morquio's and Proteus. As a rule, in each of these conditions some soft tissue manifestations accompany the condylar agenesis and/or condylar malformations.^{6,9}

Those are outlined in Table 1. In our case, there were neither common findings of other syndromes nor soft tissue manifestations.

Aplasia of the mandibular condyle without any other facial malformations is an extremely rare condition,⁴ but cases of non-syndromic mandibular condylar aplasia have been previously reported by Krogstad,⁶ Prowler and Glosman,¹⁰ Akihiko et al¹¹ and Santos

Figure 5 In an anteroposterior skull radiogram, evident facial asymmetry and the deflection of the mandible to the left side of the face was observed clearly

Figure 6 In pelvic radiography, bilateral aplasia of the collum femoris and caput femoris is revealed. Tuberculum majus are not detected bilaterally. In addition, at sacral 1, there is an arcus defect and spina bifida

Figure 7 Examination of plain film radiograph of the left arm reveals lytic defects at the ulnar medial condyle and cortical irregularities at other joint surfaces

et al.⁷ While Krogstad⁶ and Santos et al⁷ presented their cases as having an unknown aetiology, the case described by Prowler and Glassman¹⁰ was reported as most likely being of an acquired nature because the condition obviously started after the age of 6 years. Nonetheless, Akihiko et al¹¹ stated that they considered their case to be congenital. Poswillo and Robinson¹² suggested that mandibular condylar aplasia cases which are not accompanied by soft tissue anomalies must be of acquired origin. In this case, no anomaly was reported accompanying the condylar aplasia.

In our case, besides aplasia of the mandibular condyle, there were no other commonly found syndromes or soft tissue manifestations. Besides this, there was congenital unilateral femur head aplasia, yet to our knowledge, no case of concomitant presence of aplasia of the mandibular condyle and congenital aplasia of the femoral head has been reported previously.

Congenital condylar aplasia occurs embryologically as part of conditions originating in the first branchial arch.¹⁻⁵ In a newborn child, the temporomandibular joint is mobile. After the eruption of the first permanent dentition at the age of 7 years, the articular eminence begins to be prominent and its development continues at an accelerated pace until 12 years of age. If a condyle is present, an adaptive, well-defined fossa and articular eminence should be present. So, in the case of the absence of a condyle, those coherent structures should also be absent. Total absence of the condyle points to an earlier period of condyle formation.^{7,13} In our case, not even rudimentary presence of the left condyle was

Table 1 Syndromes and their skeletal and general manifestations⁹

Syndromes	Skeletal traits	Other traits
Hemifacial microsomia	Underdevelopment of the temporomandibular joint, mandibular ramus	Underdevelopment of ear and mastication muscles, hearing loss, cardiac anomalies
Goldenhar	Most severe form of microfacial microsomia	Eye tumours, fused spine
Treacher Collins	Mandibular hypoplasia, hypoplasia/aplasia of the zygomatic arch, coronoid process and condyle	Underdeveloped/malformed ears, ocular anomalies, hearing loss
Hallerman–Streiff	Dyscephalia and bird face, hypoplastic mandible/ramus, missing condyle	Cataract, microphthalmia, skin atrophy, dental anomalies
Morquio's	Abnormal development of bones, including the spine, a prominent lower face	Respiratory, cardiac, ocular and hepatic abnormalities, enamel hypoplasias
Proteus	Atypical growth of the bones, cranial hyperostosis	Thickening of skin, vascular anomalies, verrucous epidermal naevi

observed. This condition may constitute evidence that the defect originated during the pre-natal period of the patient's life.

Various treatment approaches have been proposed for treating condylar aplasia, and possibilities for influencing mandibular growth have been the topic of numerous clinical and experimental studies. A costochondral rib graft can be used to help establish an active growth centre. Surgery is often required, but the timing and regimen of this choice is still an issue to be resolved. It is advisable to postpone surgical treatment to a later time. Orthognathic treatment or a combination of such treatment and a graft can be successful.^{3,14,15} In addition, Krogstad⁶ reported that effective results were obtained through the application of a form of orthodontic activator which aimed to swing the

mandible to the unaffected side and promote formation of a mandibular condyle, albeit irregular in shape. After consultation with the departments of orthodontics and plastic surgery, surgical treatment was advised, but it was decided to conduct treatment after the completion of the patient's growth and development period. Up to the time of treatment, exercises aiming to the development of masticatory muscles were advised. The patient is now under follow up.

In conclusion, a case of unilateral aplasia of the mandibular condyle without any other facial malformations but together with bilateral congenital hypoplasia of the femur head is presented. To our knowledge, no case reporting concomitant presence of aplasia of the mandibular condyle and congenital aplasia of the femoral head has been reported previously.

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