The Development of a Giant Extraskeletal Osteochondroma in the Masticatory Space of the Mandible

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Abstract

Extraskeletal osteochondroma is an uncommon benign tumor that arises in soft tissues lacking continuity with an adjacent bone and joint. Due to similar histopathological findings, extraskeletal osteochondroma is often misdiagnosed for a conventional osteochondroma, the most common benign tumor that arises from the epiphyseal plates of long bones. The pathogenesis of extraskeletal osteochondroma is unclear, but metaplasia of synovial mesenchymal tissue is the best supported etiology. The most common sites of extraskeletal osteochondroma are the hands and feet, and it is rarely found in the maxillofacial area. We present a case of a two-year-old boy with a giant extraskeletal osteochondroma that developed in the masticatory space of the mandible along with a review of the relevant literature.

Key words: Osteochondroma, Extraskeletal origin, Mandible

Introduction

Osteochondromas are the most common benign bone tumors. Approximately 20% to 50% of benign bone tumors and 10% to 15% of all bone tumors are osteochondromas. They commonly arise from cartilage in the epiphyseal growth plates of long bones, usually in the lower extremities[1]. In the maxillofacial area, mandibular condyles and coronoid processes show dominant site predilection and are more likely to develop chondromas, osteomas, and osteochondromas because of their cartilaginous origin[2]. Although extraskeletal osteochondromas have similar histopathological features to common osteochondromas, they originate in the soft tissues without any continuity of parent bones. Extraskeletal osteochondromas most commonly arise in the digits of hands or feet and the knee. Although some reports of extraskeletal osteochondromas in the thigh, hip, buttock, and neck have been presented, extraskeletal osteochondromas arising in the maxillofacial region are very rare[3]. To the authors’ knowledge, there have been no case reports of extraskeletal osteochondromas in the masticatory space. In this article, we present a case of an extraskeletal osteochondroma arising in the masticatory space of the mandible.

Case Report

A two-year-old boy presented to the Department of Oral
and Maxillofacial Surgery, College of Dentistry, Seoul National University, Korea, with respiratory difficulty during sleep and mouth opening limitation.

Upon clinical examination, the patient had a palpable mass on the left submandibular area with submucosal bulging that deviated the uvula to the right side of the soft palatal arch, although occlusal deviation was not observed, the interincisal mouth opening was 10 mm.

Computed tomography (CT) revealed a large radio-opaque lesion in the left masticatory space. Coronal and axial CT of the head and neck showed a $4.5 \times 4.0 \times 3.5$ cm large calcifying mass confined in the left masticatory space that was distinguished from the parapharyngeal space by a thin fat layer. The lesion contained dense, calcifying bodies and was closely located to the medial surface of the mandibular body and was well demarcated by the soft tissue layer. The tumor thinned the mandibular ramus and continued to grow to besiege the pterygoid plate (Fig. 1).

Surgical resection was performed with a two way approach to facilitate the removal of the mass, which was separated into two pieces because of its large size. An intraoral incision was performed to expose the proximal half of the tumor that was close to the parapharyngeal space, while a submandibular incision was made for the distal half of the tumor (Fig. 2). A surgical chisel was used intraorally to separate and remove the upper masticatory part of the tumor close to the mandibular ramus. The rest of the tumor that was located in the lower portion was removed with a submandibular approach because further intraoral access risked damage to important nerves and

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**Fig. 1. Preoperative CT findings.**

(A) On the axial view, the multinodular calcifying mass was well-demarcated by the soft tissue layer, besieging the left pterygoid plate. (B) A coronal section showed the mass making the left mandibular ramus bend in the masticatory space.

**Fig. 2. Surgical approach for resection of the mass.**

(A) Removal of the proximal half of the tumor via intraoral approach. (B) Removal of the distal half of the tumor via submandibular approach.
vessels. The tumor was well enclosed by connective tissue and had no contact with the pterygoid plate, mandibular body, or ramus, and therefore was easily removed. The excised mass had bone-like hardness with a whitish smooth texture, encapsulated by well-defined connective tissue (Fig. 3).

A diagnosis of extraskeletal osteochondroma was confirmed based on histopathological findings. The specimen was composed of hyaline cartilage and trabecular bone. A fibrous capsule covered the cartilaginous tissues at the periphery of the tumor and hyaline cartilage gradually changed into trabecular bone with intervening endochondral ossification. At the central portion, well-developed trabecular bone and marrow appeared. Endochondral ossifications presented as multiple nodular types in the tumor and showed no directional characteristics of conventional osteochondromas that originated from epiphyseal plates of long bones (Fig. 4).

The mass was completely removed without any damage to the adjacent anatomic structures and the postoperative course was uneventful. The patient was asymptomatic after

Fig. 3. Removal of the tumor specimen. Tumor specimen was separated into two parts. Mass (A) was the proximal part of the tumor that was accessed via intraoral incision. Mass (B) was the distal part of the tumor that was removed through submandibular incision.

Fig. 4. Histopathological findings. (A) Multi-nodular endochondral ossification foci appeared without typical directional characteristics of conventional osteochondromas (haematoxylin and eosin, ×12). (B) Well-developed hypercellular chondrocytes and transparent hyaline cartilage (haematoxylin and eosin, ×200). (C) The tumor was covered with a fibrous capsule at the periphery and hyaline cartilage gradually changed into trabecular bone and marrow space with intervening endochondral ossification (haematoxylin and eosin, ×40).
surgery, with an improvement in mouth opening (35 mm) and no respiratory difficulty during sleep.

Discussion

Conventional osteochondroma, the most common benign bone tumor, is a developmental anomaly of bone rather than a true neoplasm. It results from the separation of a cartilaginous fragment of the epiphyseal growth plate of long tubular bones and has pathological characteristics of cortical and medullary bone contents with an overlying hyaline cartilage cap[1]. Extraskeletal osteochondroma occurs in the soft tissues and is very rare. It may be associated with the tendon sheath, joint capsule, or periosteum. Clinically, extraskeletal osteochondroma presents as a palpable, nonpainful, slow-growing mass. However, it can be secondarily symptomatic due to the involvement of adjacent nerves, vessels and joints that limit range of motion[3].

Extraskeletal osteochondromas are common in patients ranging from 13 to 75 years, but may occur at any age with a male predominance[4]. Reith et al.[5] suggested the following diagnostic criteria: 1) the lesion presents as a single, dominant mass, both radiographically and grossly; 2) the mass is composed of both bone and cartilage, organized in a manner similar to conventional osteochondromas; 3) the lesion is not intra-articular, which means it does not manifest within the synovial lining of a joint[4-6]. The most common sites are the infrapatellar fat pad and joint capsule of the knee (76%), followed by the foot (19%) and ankle (5%). Lesions can be treated with a simple excision and no recurrences have been reported[6].

Extraskeletal osteochondromas have histopathological features of well-demarcated and multinodular masses. The periphery of lesions is composed of cartilaginous tissues while bone tissues of a trabecular pattern comprise the central portion of the lesion. Endochondral ossification appears prominently in the interface between bone and cartilaginous tissues. The cartilaginous lesion may have a proliferation of hypercellular chondrocytes with cellular atypia. These histopathological features are similar to those of conventional osteochondromas[8].

The pathogenesis of extraskeletal osteochondromas is unknown, but they are thought to arise from the synovial tissues of the joints, tendon sheaths, or bursae in the hands and feet[3]. Metaplasia of mesenchymal cells is the most accepted proposed etiology of extraskeletal osteochondromas[7]. Lynn and Lee[8] suggested three different mechanisms of cartilage formation in soft tissues: 1) cells that can produce cartilage move from a parent bone to adjacent connective tissues; 2) precartilaginous tissue in a tendon above a joint can convert to an active phase; 3) metaplasia may occur when synovial cells differentiate into cartilage stimulated by unknown factors[9]. Metaplasia of adipose tissue, beginning as a lipoma and eventually changing to an extraskeletal osteochondroma, has been also suggested as an etiology[6]. Repeated trauma was also suggested as a cause of the occurrence of extraskeletal osteochondromas[10].

The differential diagnosis of extraskeletal osteochondromas includes variable calcifying benign masses such as conventional osteochondromas, synovial chondromatosis, myositis ossificans, and chondromas[3,6]. Extraskeletal osteochondromas frequently have been misdiagnosed as conventional osteochondromas because of their histopathological similarity, but conventional osteochondromas are osteocartilaginous exostoses that originate from long bones with a cartilaginous cap[1]. The cellular atypia of cartilage and abnormally enlarged hyperchromatic nuclei may cause misdiagnosis of extraskeletal osteochondromas for malignancy such as chondrosarcoma and synovial sarcoma[3]. However, differential diagnosis of extraskeletal osteochondromas should be established when the well-demarcated lesion of osteocartilaginous tissues exists in the para-articular soft tissues without any evidence of direct contact with adjacent bone and joint.

In the management of extraskeletal osteochondromas, periodic observation is recommended because patients usually have a slow-growing, asymptomatic mass. Suspicion of malignancy, pain, limited range of motion, and cosmetic problems make surgical resection reasonable[4]. Most of cases on mandible need surgical treatments, including surgical resection, condylectomy, and vertical ramus osteotomy, due to functional limitation and facial asymmetry[11]. Orthognathic surgery might be required for the correction of malocclusion and facial asymmetry[12].

In this case of extraskeletal osteochondroma, the lesion occurred in the masticatory space of the maxillofacial area.
and had no continuity with adjacent bone or the temporomandibular joint. Even though extraskeletal osteochondroma develops in various ages, congenital type needs early resection for the control of normal mandibular growth[12]. Surgical resection was also recommended, in this case, because of functional problems (respiratory difficulty, limitation of mouth opening) and a suspicious possible malignancy on the CT image that showed active growth. Histopathological examination excluded the possibility of malignancy and a well developed fibrous capsule facilitated removal of the mass without any surgical complications.

References