Oral fibromatosis : a case report and clinico-pathologic considerations for radiolucent lesion in the mandible

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Abstract

The fibromatoses are a broad group of fibrous proliferations. They have a biologic behavior and histopathologic pattern that is intermediate between those of benign fibrous lesions and fibrosarcoma. Because they are not common diseases, it is important to differentiate the fibromatoses from other similar diseases. In this report, we present a case of fibromatosis, and possible other diseases to be differentiated, with a review of literatures.

Case Presentation

A 17-year-old boy was referred to the department of oral and maxillofacial surgery, Yonsei university dental hospital, Seoul, Korea, with facial swelling on his left face. The patient had a history of feeling swelling 10 days ago. With the symptom considered to be of the third molar origin by a local oral surgeon, the boy had been given antibiotics, then signs were slightly improved. After the medication, the dentist found a radiolucent lesion in a radiographic evaluation. The patient's medical history was noncontributory, but had a history of direct trauma on the left mandibuar area by a bicycle, two years ago. He was not taking any medication and had no known drug allergies.

In extraoral examination, there was mild swelling of the cheek and in the region over the mandibular angle without tenderness to palpation. In intraoral examination, there was no remarkable swelling or any sign of infection.

A panoramic radiograph showed a large multilocular radiolucent lesion extending from the root of the lower left canine to the condylar neck. The inferior border of the mandible was intact (Fig. 1). A computed tomography (CT) scan was taken to assess the buccolingual extent of the lesion. On axial and coronal sections, there were a low attenuated soft tissue mass showing a pressure bony erosion of the mandible (Fig. 2, 3). In addition, thickening of the masseter, buccinator, and medial and lateral pterygoid muscles which lying adjacent to the lesion were noticed.

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Differential diagnosis

The patient's radiographic view showed a large radiolucent lesion, which led the mandible to erosive bony resorption. Tumors and tumor-like conditions of the mandible in young adolescents can attribute to several lesions of various origins which could affect this region. They can be classified as neoplastic (benign or malignant), traumatic, metabolic, infectious, or developmental in origin. The absence of infection sign in this case would exclude the infectious origin.

Taking all the above considerations into account, the differential diagnosis should include the following entities in order:

1. Traumatic bone cyst. The traumatic or simple bone cyst is a lesion with an uncertain pathogenesis. Most traumatic bone cysts of the jaws are encountered in patients between 10 and 20 years of age. They are

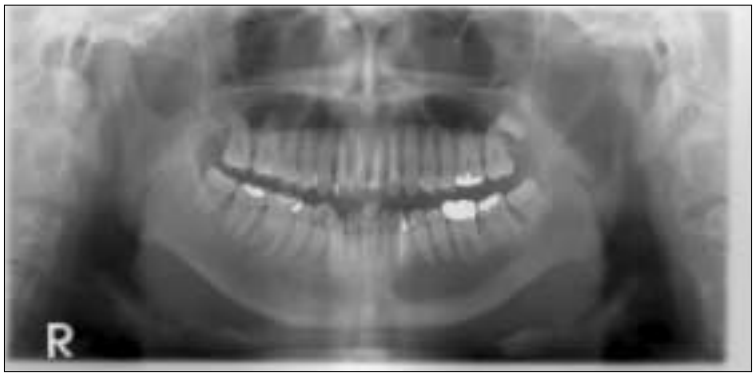

Fig. 1. Panoramic radiograph showing a large radiolucent lesion extending from symphysis to condylar neck.

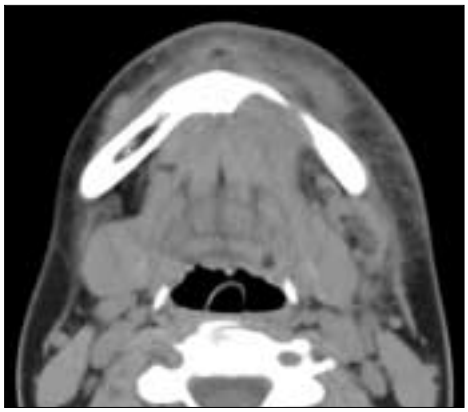

Fig. 2. Axial computed tomography scan showing a erosive lesion at the mandibular body.

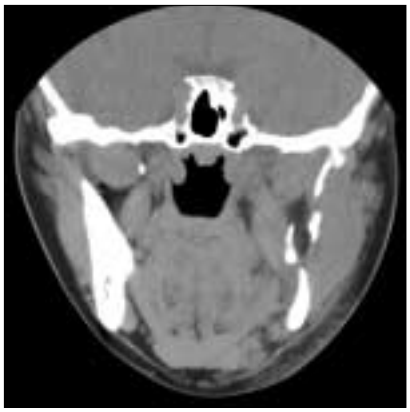

Fig. 3. Coronal computed tomography scan showing a erosive lesion at the ramal area of the mandible.

essentially restricted to the mandible, although there have been reports of the lesion in the maxilla. Bilateral simple bone cysts of the mandible are occasionally encountered. About 60% of the cases occur in males. Traumatic bone cysts are usually asymptomatic, however, about 20% of the cases shows a painless swelling. Radiographically when several teeth are involved in the lesion, the radiolucent defect often shows dome-like projections that scallop upward between the roots. Teeth that appear to be involved in the lesion are generally vital and do not show root resorption.

2. Aneurysmal bone cyst (ABC). The lesion is uncommon in the jaws. ABCs are most commonly seen in the shaft of a long bone or in the vetebral column in the patients younger than age 30. The mandible is more commonly involved than the maxilla, and the molar region is the most frequent site of involvement. Pain is a common feature, and patients often have a rapidly developing facial swelling. Radiographs show an unilocular or multilocular radiolucency with cortical expansion and thinning.
3. Neurofibroma. The neurofibroma is the most common type of peripheral nerve neoplasm. It arises from a mixture of cell types, including Schwann cells and perineural fibroblasts. Solitary tumors are most common in young adults and present as slow-growing, soft, painless lesions that vary in size from small nodules to larger masses. The skin is the most frequent location for neurofibromas, but lesions of the oral cavity are not uncommon. On rare occasions, the tumor can arise centrally within the bone, where it may produce a well-demarcated or poorly defined unilocular or multilocular radiolucency.
4. Fibromatosis. Soft tissue fibromatosis of the head and neck presents as a firm, painless mass, which may be

either rapid or insidious in growth. The lesion usually occurs in children or in young adults. The most common oral site is the paramandibular soft tissue region, although the lesion can occur almost anywhere. The tumor can grow to considerable size, resulting in significant facial disfigurement. Destruction of adjacent bone may be observed on radiographs.

5. Stafne defect. This condition represents a focal concavity of the cortical bone on the lingual surface of the mandible. In most cases, biopsy has revealed histologically normal salivary gland tissue, suggesting that these lesions represent developmental defects. However, a few of these defects contain muscle, fibrous connective tissue, blood vessels, fat, or lymphoid tissue. They present as an asymptomatic radiolucency below the mandibular canal in the posterior mandible, between the molar teeth and the angle of the mandible. Male predilection is observed. In few cases, the lesion has increased in size over time.
6. Fibrosarcoma of bone. Fibrosarcoma of bone is a malignant fibroblastic tumor that shows varying degrees of collagen production without formation of tumor bone, osteoid, or cartilage in the primary tumor or in any metastatic site. They may arise in the medullary portion of a bone or in a periosteal location. Fibrosacormas of bone may be encountered in patients over a wide age range (The average age is 40 years). About 15% of cases occur in the craniofacial bones, and the mandible is the predominant site. Radiographically, fibrosarcomas of bone present as lytic, destructive lesions. In patients with relatively slow-growing tumors, the radiolucent area may be fairly well defined, suggesting a benign process.
7. Desmoplastic fibroma. The desmoplastic fibroma of bone is a rare tumor that appears to be the osseous counterpart of soft tissue fibromatosis. The mandible is the fourth most commonly affected bone. Most examples of desmoplastic fibroma of bone are discovered in patients younger than 30 years of age. There is no sex predilection. Of the reported cases involving the jaws, 90% have occurred in the mandible, most often in the molar-angle-ascending ramus area. A painless swelling of the affected area is the most common initial complaint. Radiographically, the lesion presents as a unilocular or multilocular radiolucent area. If the lesion erodes through the cortex, an accompanying soft tissue mass will be present.
8. Ossifying fibroma. The ossifying fibroma is a well-demarcated and occasionally encapsulated neoplasm composed of fibrous tissue that contains varying amount of calcified tissue resembling bone, cementum, or both. They may occur over a wide range, but the greatest number of cases are encountered during third and fourth decades of life. There is a definite female predilection. It is much more common in the mandible, most often found in the premolar and molar region. Large tumors may cause a painless swelling, and paresthesia is rarely seen. Radiographycally, the lesion is oftenly well defined and unilocular. But some examples show a sclerotic border. Depending on the amount of calcified material produced in the tumor, it may appear completely radiolucent; more oftenly it shows varying degrees of radiopacity. Large lesions often cause expansion of the inferior cortex of the mandible.

Fig. 4. Histopathological examination showing dense fibrosis in the subepithelial connective tissue (hematoxylin-eosin, original magnification, × 40).

Subsequent Course

The tumor was excised via an intraoral and submandibular approach under general anesthesia.

Pathologic Diagnosis

Pathologic report was *'consistent with fibromatosis'*. (Fig. 4).

Discussion

The fibromatoses are a broad group of fibrous proliferations. The frequency with which fibromatoses involve

the head and neck is difficult to assess because of the pecularities of claasification used by different authors[1]. They have a biologic behavior and histopathologic pattern that is intermediate between those of benign fibrous lesions and fibrosarcoma. A number of different forms of fibromatoses are recognized throughout the body, and they often are named on the basis of their particular clinicopathologic features. In the soft tissue of the head and neck, these lesions are frequently called a juvenile aggressive fibromatosis or extra-abdominal desmoids. Similar lesions within the bone have been called desmoplastic fibromas[2].

The etiology of fibromatoses is still unclear. Some explanations have been suggested like hormonal cause[3], viral theory, chromosomal alterations[4], and abnormal expression of the c-sys oncogene and of platelet-derived growth factor(PDGF)[5].

Fibromatoses are uncommon soft tissue mass lesion that can occur in all anatomic sites and accountings for 6.9% of soft tissue tumors[6], and among these, only 5% of all fibromatoses are found in the head and neck region. Pathologically soft tissue fibromatoses are characterized by a cellular proliferation of spindle-shaped cells that are arranged in streaming fascicles and are associated with a variable amount of collagen. The lesion usually is poorly circumscribed and infiltrates the adjacent tissues. Hyperchromatism and pleomorphism of the cells should not be observed[2].

The correct treatment is excision (radical and compartmental, if possible) and continuous follow-up[7]. Difficulty with complete excision is reflectd in the relatively high recurrence rates of fibromatoses excised from the head and neck region. Fasching et al[8] showed that fibromatoses recurred at a very high rate when margins were positive but also frequently when margins were considered negative. Without complete excision, 12 of 14 tumors (25%) recurred. However, some literatures contain occassional reports of spontaneous regression and descriptions of arrested growth after incomplete resection without subsequent therapy[9]. Siegel et al[10] suggested a guideline for therapy. First, meticulous surgical excision is essential. Second, if tumor margins are known to be positive, postoperative adjunctive therapy should be considered. Finally, close follow-up is essential.

If the condition is inoperable[11] or tumor margins are known to be positive, postoperative adjunctive therapy, such as radiation therapy, should be considered. However, the possible benefits of radiation therapy need to be weighed against its potential for long- and short-term complications. Radiation therapy should be used with caution in the pediatric population because of the well-known effects of radiation on bone growth and the potential for the delayed development of a cancerous lesion[10].

If radiotherapy is contraindicated, chemotherapy is an alternative. Several pharmacologic interventions such as progesterone[12,13], antiestrogens, nonsteroidal anti-inflammatory drugs, warfarin, vitamin K, and ascorbiate[14]. The use of tamoxifen or testolactone is also suggested[15].

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