Well differentiated nonmetastasizing fibrosarcoma (aggressive fibromatosis) of mandible: a rare case report and a literature review

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Fibromatosis in the maxillofacial region is a very rare occurrence among diverse pathologic conditions, and because of the rarity of this tumor, definite treatment regimen is not established, which may be a contributing factor for a high recurrence rate. Fibromatosis may attain a large size and cause compression, infiltration, and destruction of adjacent structures. Such growth behavior presents severe management problems, especially in the head and neck region, where the presence of many vital structures within a small space makes the patient susceptible to the effects of the fibromatosis, likewise making complete excision difficult. We report the case of a 28-year-old female patient with aggressive desmoid fibromatosis involving the mandible, with a literature review of postoperative pharmacologic management to prevent recurrence. (Oral Surg Oral Med Oral Pathol Oral Radiol 2013;116:e98-e102)

Aggressive fibromatosis is a rare benign tumor that develops from the tissue of the musculoaponeurotic system and is characterized by local aggressive behavior.[1,2]

The terminology of the World Health Organization classification was used, using the term extraabdominal fibromatosis; however, other terms exist, including extraabdominal desmoid tumor, desmoid tumor, well differentiated nonmetastasizing fibrosarcoma, grade I fibrosarcoma, and aggressive fibromatosis. The latter term emphasizes the clinical nature of the disease process, indicating the aggressive behavior it may sometimes show.[3]

The lesion usually arises from the connective tissue of the muscle, the overlying fascia or aponeurosis.[3] Differentiating this entity from low-grade fibrosarcoma may be difficult.[2] However, it resembles a "borderline" lesion without metastases, though local recurrence rates vary from 20% to 70%.[4] Fibromatosis may attain a large size and cause compression, infiltration, and destruction of adjacent structures. Such growth behavior presents severe management problems, especially in the head and neck region, where the presence of many vital structures within a small space makes the patient susceptible to the effects of the fibromatosis, likewise making complete excision difficult.[3]

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CASE REPORT

A 28-year-old female patient reported to the Department or Oral and Maxillofacial Surgery in our hospital for swelling on the left side of face. The patient revealed a 6-week history of swelling that was gradually progressive in size and resulted in facial disfigurement, which was the patient's concern for seeking treatment.

Clinical evaluation revealed a single diffuse swelling in the left preauricular area measuring 4 × 5 cm in greatest dimensions with healthy overlying skin. The left-side ear lobule was obscured which was suggestive of swelling not related to the parotid gland. The swelling was nontender, firm in consistency, and fixed. There were no symptoms of lower lip paresthesia, and mouth opening was within normal limits.

Radiologic evaluation by orthopantamogram (Figure 1) revealed a single radiolucent lesion with well defined radiopaque border anteriorly and superiorly, with ill defined border posteriorly and inferiorly. Conventional computerized tomogram (Figure 2) revealed destruction of both buccal and lingual cortical plates along with lingual extension of lesion in pterygomandibular space. Aspiration of the lesion was nondiagnostic. Incisional biopsy was performed under local anesthesia. Histopathologic evaluation revealed the pathology as aggressive fibromatosis. In view of aggressive nature, high recurrence rate, and extensive extension, ramus resection along with its processes was planed under general anesthesia.

Under general anesthesia with all aseptic precaution, a modified incision was marked in the upper neck crease that was a combination of Risdon and Hind approaches with retroauricular extension. Disarticulation ramal resection was carried out along with disease-free soft tissue margin. After achieving hemostasis, reconstruction was done with recon-

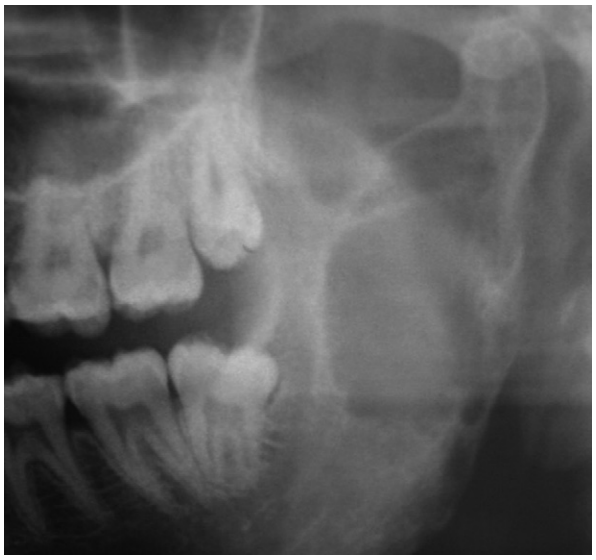

Fig. 1. Orthopantamogram.

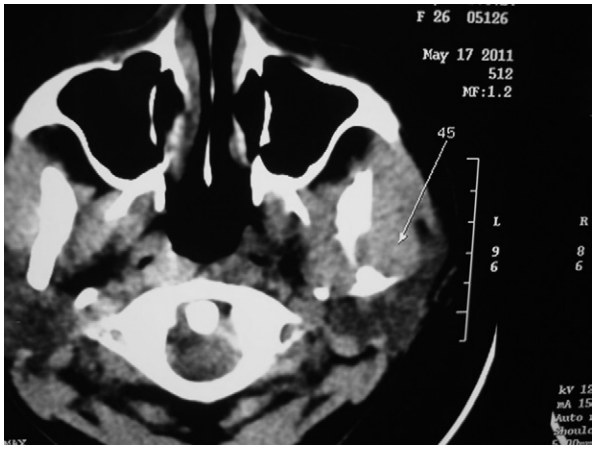

Fig. 2. Conventional computerized tomogram.

struction plate with condylar component. Postoperative recovery was uneventful. Excisional biopsy on histopathologic examination showed stromal condensation of fibroblast and homogenization of collagen fibers were prominent features. Stromal fibroblasts at places were arranged in the form of streaming fascicles (Figure 3), which supported our previous diagnosis.

In the postoperative period, the patient was started with antiestrogenic agent tamoxifen at 2 mg/kg/d and indomethacin at 75 mg/d to prevent recurrence. The patient is followed periodically.

DISCUSSION

Fibromatosis in maxillofacial region is a very rare occurrence among diverse pathologic conditions, and because of its rarity, definite treatment regimen has not been established, which may be a contributing factor for the high recurrence rate. Hoos et al.[6] have reported

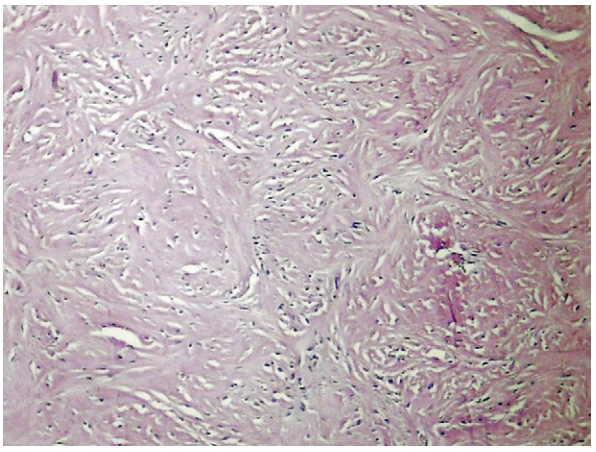

Fig. 3. Immunohistochemistry of soft tissue specimen.

a local recurrence rate in extraabdominal desmoids of 20%-77%. In the head and neck region, Hoos et al.[6] and Morioka et al.[7] have reported 46%-62% and 40%-70% recurrence rates, respectively. Enzinger and Shiraki[8] reported that younger patients and larger tumors have a higher propensity for recurrence. In view of the aggressive nature and high recurrence rate, surgical planning and postsurgical pharmacologic treatment play an important role.

Radiographic evaluation provides information about type of bone involvement, periosteal reaction, and its proximity to vital anatomic structure, which is major concern in the anatomically complex maxillofacial region. In aggressive fibromatosis, the cortex of the bone is rarely breached and the mass may be surrounded by a shell of cortical bone.[9,10] But in our case, conventional computerized tomographic evaluation revealed that the tumor mass had involved the ramus of the left mandible with bicortical perforation, which added to the complexity of management and a risk factor for recurrence.

Histopathologic examination can distinguish between the various forms of the disease, and above all it is important for differential diagnosis to rule out metastasizing tumors such as fibrosarcoma, fibro-osseous lesions such as monostotic form of fibrous dysplasia, Albright syndrome, cherubism, and other neoplasms such as desmoplastic fibroma, neurofibroma, and proliferative myositis. In our case, ruling out metastasis from breast tumor was also important, considering the age and sex of the patient.

The most significant differential diagnosis is fibrosarcoma, which is diagnosed when the tumor cells show atypical cytologic features and/or a significant number of mitotic figures (>1 per high-power microscopic field). This differentiation may be difficult, but it is essential because fibromatosis does not possess the metastasizing potential of fibrosarcoma.