Misdiagnosed fibrosarcoma of the mandible mimicking temporomandibular disorder: a rare condition

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The jawbones can be sites of various neoplastic conditions. Given the variety of processes affecting this particular anatomical area, formulation of a precise diagnosis can be challenging to clinicians. Limited jaw movement, pain, and facial asymmetry are common signs among patients, especially those with temporomandibular disorder (TMD). This paper reports a case of primary fibrosarcoma affecting the mandible and surrounding structures in a 14-year-old girl presenting signs and symptoms similar to TMD. Her condition was misdiagnosed, and she was treated for TMD over an extended period before the correct diagnosis was made for fibrosarcoma. The patient underwent surgical resection with postsurgical radiotherapy and chemotherapy and now is being followed up. Although malignant lesions are rare in the temporomandibular joint (TMJ) region, dentists are advised to be aware of the condition and to keep in mind that patients who are admitting for TMD can also possibly be affected from neoplasms. Hence, those patients have to be examined meticulously to avoid misdiagnosis and mistreatment. **(Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2007;104:e26-e29)**

Soft tissue sarcomas are rare in the oral and maxillofacial region and account for less than 1% of the cancers. At one time, fibrosarcoma was the most common soft tissue sarcoma. With the introduction of electron microscopy and immunohistochemistry, it became evident that many previously diagnosed fibrosarcomas were other spindle cell malignant lesions. Today, fibrosarcoma is defined as a malignant spindle cell tumor showing a herringbone or interlacing fascicular pattern without expression of other connective tissue cell markers.[1-3] Clinically, soft tissue sarcomas are difficult to diagnose. They may grow to a considerable size before causing symptoms, and these symptoms in the head and neck region may be masked by a concurrent temporomandibular disorder (TMD). Therefore, they may be overlooked for a considerable amount of time. Symptoms of neoplasms located in this anatomic region may mimic TMD.

This manuscript describes a case of fibrosarcoma of the mandible misdiagnosed as TMD.

CASE REPORT

A 14-year-old girl was referred to Ankara University, Faculty of Dentistry, Department of Oral Diagnosis and Radiology. One year previously, she had a complaint of difficulty in opening her mouth and malocclusion (due to primary contacts), especially on the right-side teeth. She also had pain that appeared in the right preauricular region.

After her symptoms had started, the patient was referred to a pediatric dentist and an orthodontist in a private clinic, who diagnosed the problem as TMD. Firstly, she was prescribed a myorelaxant and analgesic combination. She used prescribed medications, which did not change the symptoms. Again she appeared to the same clinic, and her doctor performed numerous occlusal adjustments and changed her filling in the mandibular left first molar. There was no improvement of symptoms after occlusal adjustments. She had significantly more difficulty in opening her mouth and an increase in pain intensity. The pain referred to the ear, eyes, forehead, and to the temple on the right side. Finally, after an unsuccessful treatment for some months, a bite splint (2 mm) was applied due to suspicion of disk displacement.

After 1 year, the patient consulted at Ankara University, Faculty of Dentistry, Department of Oral Diagnosis and Radiology. The patient's medical history was noncontributory. There was pain and crepitus on the right temporomandibular joint (TMJ) on palpation. However, paresthesia was not evidenced and sensory-neural examinations were normal. On intraoral examination, the right lateral pterygoid muscle was painful on palpation, and we detected lymphadenopathy in the

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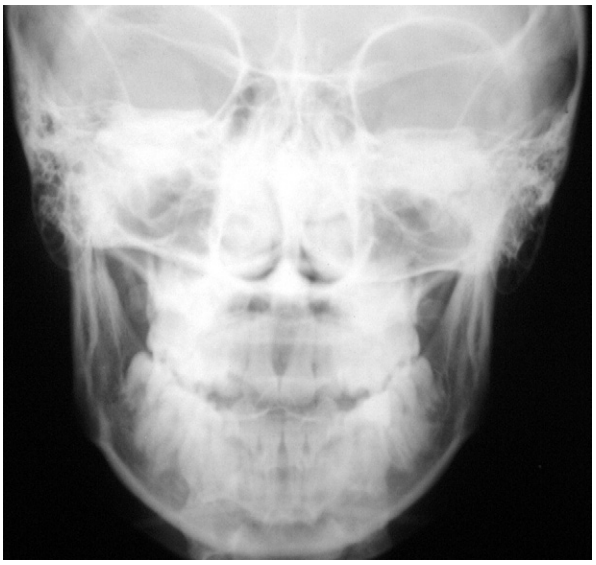

Fig. 1. Conventional posterior-anterior radiography. No evidence was found.

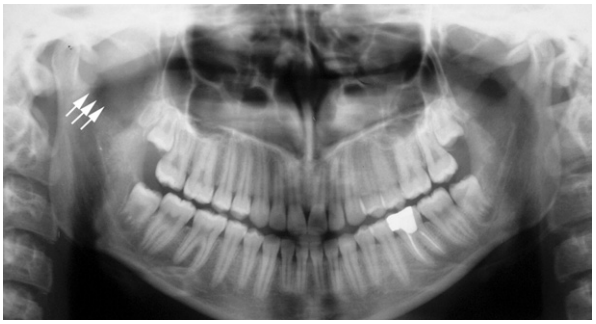

Fig. 2. Panoramic radiography shows irregularity and radiolucent erosive area in right ramus mandible (*arrows*).

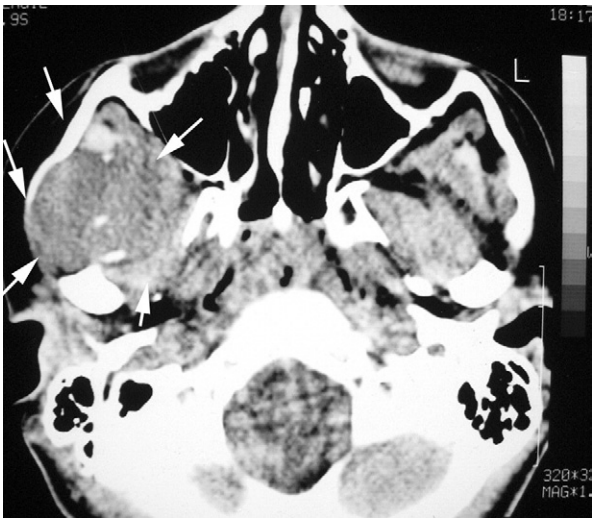

Fig. 3. Computed tomographic image shows a large homogeneous mass (*arrows*).

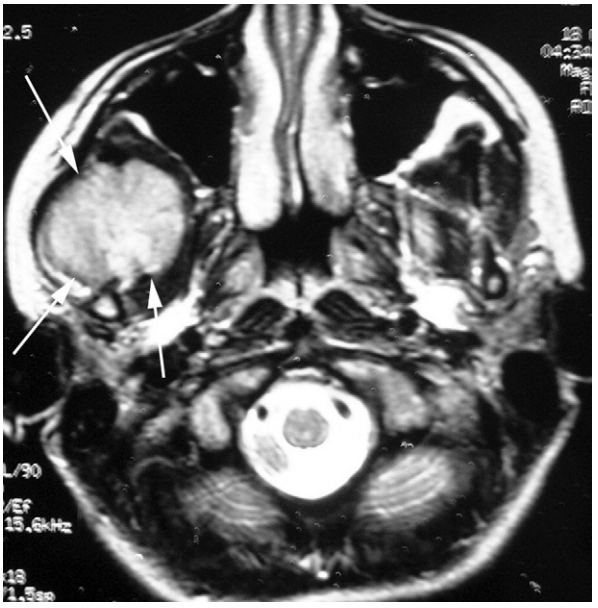

Fig. 4. Fat-saturated T2-weighted axial images demonstrate an inhomogeneous hyperintense lesion that was located on the ramus mandible, extending through the infratemporale fossa, orbita, and soft tissue (*arrows*).

right submandibular region and in both the preauricular and the retroauricular regions. The patient could open her mouth for approximately 17 mm.

There was no evidence of any disease on conventional posterior-anterior radiographs because of superimpositions of overlapping structures on the TMJ region (Fig. 1). However, the panoramic radiography (Planmeca 2002 CC Proline, Helsinki, Finland) revealed a suspicious irregularity and radiolucent erosive change on the right ramus between the condyle and coronoid processes (Fig. 2). Therefore, the area was examined with computed tomography (CT) to assess any bone involvement. The CT revealed a large homogeneous mass including the TMJ region and the zygomatic bone (Fig. 3). Magnetic resonance imaging (MRI) can contribute significantly to the diagnosis of these masses and can also supply valuable information about the extent of the lesion. T1- and T2-weighted MRI showed a mass with heterogeneous low-signal intensity, which spread from the ramus mandible to the infratemporal fossa and orbita (Fig. 4). The patient underwent surgical tumor resection. At microscopic examination, the final diagnosis was high-grade fibrosarcoma. She also received postsurgical chemotherapy and radiotherapy with a total dose of 60 Gy. At the present time, she is followed up at an interval of every 3 months.

DISCUSSION

Fibrosarcoma is a primary malignant mesenchymal tumor of fibroblastic origin.[1-7] Kulander and Bolen[8] defined it as "a neoplasm of fibroblasts capable of producing metastatic spread." Fibrosarcomas can arise in soft tissue or within bone. Intraosseous fibrosarcomas may develop endosteally or possibly periosteal, the latter affecting bone by spread from adjacent soft tissue.[5,6,9-11] It can occur in any location, but the bone extremities are the main affected site; occurrence in the maxilla is rare with an incidence ranging from 0% to 6.1% of all primary fibrosarcoma of the bone.[6] The mandible is the most common site.[1,6,7] Initially, primary intraosseous fibrosarcoma is symptom free. When it reaches a considerable size, symptoms begin, but these symptoms are usually nonspecific.[5,7,10-12] As a result, it can be overlooked for a prolonged time.[4] Soares et al.[6] reported a case of a painless swelling in the right mandibular canine in a 16-year-old girl. The patient had used orthodontic appliance for 1 year because of posterior crossbite. The general dentist preferred to follow-up the lesion because he supposed that the bone resorption was caused by the orthodontic appliance. After 1 month, an incisional biopsy was performed, and the final diagnosis was a fibrosarcoma.

A patient with nasopharyngeal carcinoma, complaining of headache, facial pain, and trismus, was misdiagnosed as TMD.[13] Cimino et al.[14] reported a case of mandibular condyle osteochondroma that was treated as TMJ internal derangement for half a year.

Symptoms of a tumor may be pain as headache, earache, or as swelling, paraesthesia, and limited mouth opening. All these symptoms can be present in patients with TMDs. Therefore, the same symptoms caused by a tumor in the head and neck region may be detected by a concurrent temporomandibular disorder.[4,13-16]

In this article, we have presented a patient with a fibrosarcoma in the ramus mandible, which had been incorrectly diagnosed and treated as TMD.

The physicians who perform the initial examination may be less likely to identify symptoms associated with TMD than a dental professional who may have specific interest in TMD. In some instances, a narrow TMD-oriented approach may have fatal consequences. There are 2 potential dangers: (1) a person who does not need treatment may be overdiagnosed (and treated), and (2) a patient who is in need of treatment but does not have a TMD may be misdiagnosed as having a TMD and subsequently treated due to misdiagnose.

Gobetti and Türp[4] emphasized the need to avoid such a diagnostic bias in patients with complaints of orofacial pain in which pain and limited range of mandibular motion are rather nonspecific and may be the result of a number of medical and dental problems. They thought that several features which might help the clinician to differentiate a TMD condition from a neoplasm in a patient who complains of facial pain could be identified. The most prominent of these features are as follows:

- neurologic signs, such as numbness in the distribution of the trigeminal nerve, or auditory complaints, such as a decrease in hearing
- constant pain that is not influenced by mandibular movements
- an increase in severity of symptoms over time, or an absence of change in symptoms, despite several treatment attempts by previous providers
- the presence of swelling, including lymphadenopathy
- ear, nose, and throat signs and symptoms, such as nosebleed, nasal stuffiness, hemoptysis, drainage from the ear, and dysphagia
- unexplained weight loss

Patient history and detailed clinical and radiographic examination are very important to evaluate the malignant condition. Radiographically, the fibrosarcoma appears in most cases as a purely lytic lesion with a destructive pattern. The cortex is thinned or disrupted, and soft tissue invasion is detected in up to 86% of cases.[4-6,7,10,11] The findings of CT and MRI led to show suspicion of the process.[4,17,18]

In the present study, panoramic radiography showed a radiolucent area with an irregular border and a destructive pattern between the condyle and coronoid processes in the ramus mandible; however, posterior-anterior projection was normal. In addition, CT and MRI showed that the lesion had both bone and soft tissue involvement.

The treatment of choice is surgical resection with a wide margin. The need for adjuvant radiotherapy and/or chemotherapy is still unclear and is normally indicated in high-grade tumors because these tumors may present subclinical or microscopic metastases at the time of diagnosis.[6,7,13,19-22] In addition, prophylactic neck dissection is controversial.[7] Our case was treated with a combination of surgical resection, chemotherapy, and radiotherapy. The overall survival rate at 10 years may vary from 21.8% to 83%, and clinical stage, histological grade of malignancy, and local recurrences are the most important prognostic factors.[7]

Although fibrosarcomas are rare pathologies, dentists have to be aware of these lesions and should keep in mind that patients who are admitting for TMD symptoms and malocclusion can also possibly be affected from neoplasms. Hence, those patients have to be examined meticulously to avoid misdiagnosis and mistreatment.

CONCLUSION

Patient history and thorough clinical and radiological evaluations are essential for differential diagnosis before initiating therapy. If the patient does not respond to conventional treatment, the diagnosis should be reassessed and the patient should be reevaluated. This case should remind clinicians that patients with persistent facial pain and limited mouth opening may indicate malignancy instead of TMD.

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