Fibrosarcoma of the temporomandibular joint area: benefits of magnetic resonance imaging and computed tomography

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Fibrosarcoma is a malignant neoplasm composed of fibroblasts that produce collagen and elastin. It may arise either in the soft tissue or in bone, with the latter comprising 20% of all cases. The osseous origin of fibrosarcomas has been debated since 1940, when Ewing named the entity “fibroblastic sarcoma,” but the concept has been generally accepted. Fibrosarcomas in the masticator space can be diagnostically challenging. A total of 76 cases of fibrosarcoma in the jaw exist in the English-language literature from before 2007. Here, we report a case of primary fibrosarcoma in the mandible of a 33-year-old woman with symptoms and radiologic signs mimicking temporomandibular disorder. We also present a literature review of fibrosarcomas in the jaw.

CASE REPORT

A 33-year-old woman presented with a 1-year history of trismus associated with a clicking noise in the left temporomandibular joint (TMJ). Her chin shifted to the right side when she opened her mouth (Figure 1). Panoramic radiographs (Figure 2, A, B) and magnetic resonance imaging (MRI) scans (Figure 3, A-H) of the TMJ regions showed signs of temporomandibular disorder (TMD), so conventional treatment for TMD was prescribed, including occlusal splints, mouth-opening exercises, and a muscle relaxant for 6 months. When the MRI scout images were analyzed retrospectively, the findings of 2 of 5 scout images were suggestive of tumor (see Figure 3, A, E).

The patient was referred again 6 months after the initial visit for investigation of insomnia caused by spontaneous pain in the left TMJ area. She also had paralysis of the lower lip and left chin. She had herpes labialis and gastroenteritis caused by stress. Given the suspicion of malignancy, both MRI and computed tomography (CT) were performed. MRI showed a round, inhomogeneous mass in the pterygomandibular portion of the masticator space, which caused minimal medial narrowing of the left parapharyngeal space (Figure 4, A, B). CT found significant bony erosion of the inner cortex of the left ramus of the mandible (Figure 5). The patient was referred to another hospital for further management because the tumor grew toward the pharyngeal arches. An incisional biopsy was subsequently performed, and the initial diagnosis was a sarcoma based on the histopathologic examination of the biopsy. A definitive histologic diagnosis was reached by exclusion. The tumor was diagnosed as a grade 3 (poorly differentiated) fibrosarcoma using the FNCLCC (Fédération Nationale des Centres de Lutte Contre le Cancer) grading system. Histopathologic examination found that the tumor contained spindle cells arranged in compact fascicles that were intersected by various amounts of delicate thin to dense keloid-like collagen. Cell bundles were arranged at acute angles to each other, whereas the presence of fascicles was subtler in other areas. A prominent storiform pattern was not seen (Figure 6). The fibrosarcoma was resected followed by postoperative radiotherapy and chemotherapy. The tumor was removed, along with the left ramus of the mandible, a portion of the left maxilla, a deep part of the parotid gland, and lymph nodes located superior to the omohyoid muscle. The cheek mucosa was reconstructed using a free anterolateral thigh flap (Figure 7). The patient recovered and got married afterward; however, lung metastasis with

Statement of Clinical Relevance

There are significant pitfalls of relying on a panoramic radiograph when patients have symptoms of temporomandibular disorders.
chest wall invasion was detected 3 years after the surgical procedure. After further surgery and chemotherapy for the lung metastasis, the patient remains well with no signs of recurrence.

**DISCUSSION**

Fibrosarcoma is a rare malignant tumor of fibroblastic origin that particularly affects the long bones and accounts for about 5% to 6% of all adult soft tissue sarcomas.\(^5\)\(^-\)\(^8\) According to Leitner et al., fibrosarcomas constitute approximately 10% of sarcomas in the head and neck region, with the mandible being the most common site of occurrence. Fibrosarcomas may arise in any part of the jaw and are classified as either peripheral or central type.\(^10\)\(^-\)\(^11\) Fibrosarcoma of the bone may be associated with conditions such as fibrous dysplasia\(^12\)
and Paget disease and may arise after radiotherapy. Onset is usually before the age of 50, with a peak incidence between 20 and 40 years, but it may occur at any age. They usually present as an asymptomatic, firm mass in the soft tissue of the face, neck, scalp, or paranasal sinuses and may grow to a considerable size before causing symptoms. Symptoms include pain, swelling, loosening of the teeth, paresthesia, and occasionally ulceration of the overlying mucosa.

Involvement of the TMJ or paramandibular musculature is often accompanied by trismus. Most fibrosarcomas appear radiographically as lytic lesions with a destructive pattern. The cortex is thinned or disrupted, and soft tissue invasion is detected in up to 86% of cases. However, fibrosarcoma of the jaw cannot be distinguished from other conditions that cause destructive lesions in the bone.

Histologically, fibrosarcomas are composed of a uniform population of spindle-shaped cells arranged in a fascicular or herringbone growth pattern with a variable amount of collagen production. Given that these “fibrosarcomatous” features can be seen in a wide variety of tumors, the majority of tumors previously considered to reside in this category are probably better placed in other categories. Thus, fibrosarcoma is a diagnosis of exclusion that cannot be made using a limited amount of tissue. In the present case, we could not reach a definite histologic diagnosis without an excisional biopsy. Other malignant tumors (such as metastatic carcinoma, multiple myeloma, monophasic fibrous synovial sarcoma, malignant fibrous histiocytoma, malignant nerve sheath tumor, and liposarcoma) must be considered in the differential diagnosis, as well as benign tumors such as benign fibrous histiocytoma, fibroma, and fibromatosis. If a suspected fibrosarcoma enlarges the affected jaw, the practitioner must rule out chondrosarcoma and osteosarcoma, both of which have a defined internal structure.
Wide surgical excision is the mainstay treatment for fibrosarcoma, with overall survival rates of 45% to 82%. Despite minimal benefit, radiochemotherapy is recommended in cases of incomplete resection with positive margins or high-grade tumors with locoregional spread. Complete resection with clear surgical margins is associated with a favorable prognosis.

Soames and Southam stated that fibrosarcoma of the oral soft tissues is rare but appears to have a good prognosis, with reported 5-year survival rates of about 70%. Distant metastasis is rare, with the lungs being the most common site of metastasis, as was seen in the present case. Fibrosarcoma of the jaw is rare; few cases have been reported. Orhan et al. reported a case of primary fibrosarcoma affecting the mandible and its surrounding structures in a 14-year-old girl presenting with signs and symptoms similar to those of TMD. Herrera et al. described a 72-year-old woman with a left parotid, low-grade, mucoepidermoid carcinoma and a myxoid fibrosarcoma of the condylar head of the left ramus of the mandible who presented with symptoms similar to those found in TMD. Angiero et al. described 2 men, aged 53 and 71 years, with primary fibrosarcoma of the jaw with intraosseous growth. In the present case, the periosteal origin of the lesion made its detection on radiographs difficult, and it exerted pressure on the medial surface of the mandible.

Case reports of fibrosarcomas of the jaw are summarized in Table I. Among these reports, only the cases described by Herrera et al. and Orhan et al. had symptoms that mimicked those of TMD. The presence of a clicking noise is a frequent manifestation of TMD. In TMD, joint noises are accompanied by a restriction of mandibular mobility and joint pain. The characteristic feature in our case was the acute, spontaneous pain in the left TMJ, but reduced joint movement was found only in the right TMJ on radiography; these features did not fit those of TMD. Specifically, panoramic visualization of the TMJ found that restricted movement occurred only on the right side. Careful observation found that the right condyle rotated and rolled, whereas the left condyle merely rolled without rotating. These findings were unusual for TMD, assuming that “TMD” represents symptoms caused by abnormalities of the masticatory muscles or joint capsule, disk displacement, and condyle deformities. TMD is defined as “a collection of medical and dental conditions affecting the temporomandibular joint (TMJ) and/or the muscles of mastication, as well as contiguous tissue components.” We usually diagnose TMD if the patient experiences trismus, clicking or crepitation noise in the TMJ, or pain on mouth opening, mastication, or occlusion. Considering the history of trismus and clicking noise followed by jaw deviation, a provisional diagnosis of TMD because of nonreducible disk displacement was made in the present case, and initial radiologic examination was performed based on this assumption. The deviation of the jaw might suggest the existence of a space-occupying lesion. The jaw can also be shifted from the clicking side toward the contralateral side in cases of TMD. Notably, jaw deviation was present when the patient opened her mouth maximally, which is not possible in TMD. Enlargement of the

Table I. Case reports of fibrosarcomas of the jaw

<table>
<thead>
<tr>
<th>Authors</th>
<th>Year</th>
<th>Age (y)</th>
<th>Sex</th>
<th>Site</th>
<th>Maximal opening</th>
<th>Treatment</th>
<th>Recurrence</th>
<th>Follow-up</th>
<th>Status</th>
</tr>
</thead>
<tbody>
<tr>
<td>Herrera et al.</td>
<td>2007</td>
<td>72</td>
<td>F</td>
<td>Mandible</td>
<td>40 mm</td>
<td>Surgery</td>
<td>None</td>
<td>3 mo</td>
<td>Alive</td>
</tr>
<tr>
<td>Angiero et al.</td>
<td>2007</td>
<td>53</td>
<td>M</td>
<td>Mandible</td>
<td>NA</td>
<td>Surgery</td>
<td>Local + lung</td>
<td>3 y</td>
<td>Dead</td>
</tr>
<tr>
<td>Orhan et al.</td>
<td>2007</td>
<td>14</td>
<td>F</td>
<td>Mandible</td>
<td>17 mm</td>
<td>Surgery + RT + CT</td>
<td>None</td>
<td>3 mo</td>
<td>Alive</td>
</tr>
<tr>
<td>Pereira et al.</td>
<td>2005</td>
<td>41</td>
<td>F</td>
<td>Mandible</td>
<td>NA</td>
<td>Surgery + RT</td>
<td>None</td>
<td>3 y</td>
<td>Alive</td>
</tr>
</tbody>
</table>

NA, not available; RT, radiotherapy; CT, chemotherapy.
tumor during the 6-month period, as shown by the difference in tumor size between the initial and subsequent MRI (see Figures 3 and 4), most likely caused the spontaneous pain in the left TMJ area. As shown in Figure 5, the close proximity of the tumor to the mandibular canal might have caused paralysis of the lower lip and left chin.

Signs and symptoms of TMD could be caused by a variety of intra- and extra-articular conditions, thus making the diagnosis of pain and dysfunction of the masticatory system complex. Our case illustrated that lesions in the masticator space in the early stages can mimic TMD by manifesting as pain and trismus. Subsequent mandibular swelling and deviation prompted further investigation that found the underlying cause. Careful evaluation including radiography should be performed in all patients presenting with TMJ symptoms. Causes other than TMD should be considered in individuals with features of TMD who do not fit the typical demographics of patients with TMD.

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REFERENCES

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