Advancing Mandibular Swelling: A Diagnostic Dilemma

KF Basavaraj, AU Madihalli, Abdul Mujeeb, Samir Mansuri, Mohammed Abid Hussain

ABSTRACT
Asymmetrical swelling of the mandible in adolescence may pose a significant diagnostic dilemma. The differential diagnosis ranges from traumatic, infectious, and metabolic processes to benign and malignant tumors. Also may present with similar clinical and radiological features, making an accurate diagnosis quite difficult. This is an illustrative case involving a 30-year-old female who initially presented with complaint of pain and swelling in the lower left side of the face for 2 months. Multiple investigations and several biopsies were required to arrive at a diagnosis. This paper deals with a case report of a fibrosarcoma involving the mandible highlighting the importance of early diagnosis and treatment planning.

Keywords: Fibrosarcoma, Mandible, Diagnostic dilemma.


INTRODUCTION
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 for clinicians. Fibrosarcoma is considered to be one of the least common primary malignancies of bone in the head and neck areas. It represents only 0.15 and 0.11% of two large series of cases of fibrosarcoma reported. In both these series the mandible was more often involved than the maxilla. It is most commonly seen in the 3rd and 6th decades of life. This neoplasm produces only collagen fibers unlike an osteosarcoma or chondrosarcoma where a diagnosis is made based on the production of new bone or cartilage. This tumor can be located centrally, in which case it could arise from embryonic, neurogenic or odontogenic tissue, or peripheral in origin with secondary spread to the bone. If the tumor arises from normal bone it is termed as ‘Primary fibrosarcoma’, where as if found to be originating from a pre-existing lesion of the bone it is termed ‘Secondary fibrosarcoma’. Another variant termed ‘Periosteal fibrosarcoma’ has its origin from the soft tissues adjacent to the bone and is considered to have a better prognosis than the medullary type of fibrosarcoma.

CASE REPORT
A 30-year-old female patient was admitted to the Department of Oral, Maxillofacial and Reconstructive Surgery, Bapuji Dental College and Hospital, Davangere, for an incisional biopsy of a radiolucent lesion present at the left angle of the mandible. The apparently healthy patient had a chief complaint of pain and swelling in the lower left side of the face for 2 months. The pain was continuous, throbbing and localized to the above mentioned area and was associated with a gradual increasing swelling (Fig. 1).

Past dental history revealed that the patient had undergone extraction of the left posterior teeth of the mandible about 6 to 7 months back and was prescribed routine analgesics and antibiotics for the same. The healing was uneventful, but the patient was experiencing discomfort and pain at the extraction site since then.

On general physical examination the patient was found to be moderately built and nourished with all vital signs within normal limits. Extraoral examination revealed a diffuse swelling measuring about 5 × 3 cm on the left side of the ramus of the mandible. Anteriorly the swelling extended about 1 cm (Fig. 1). Beyond the anterior border of the masseter muscle and posteriorly beyond the posterior border of the ramus. Inferiorly the swelling extended below the lower border of the mandible with diffuse extension onto the cheek superiorly. There was no localized increase in temperature. The swelling was firm to hard in consistency and showed mild tenderness.

Mouth opening was normal and upon intraoral examination a swelling was noticed in the lower molar region with obliteration of the buccal vestibule. The lower
left second and third molars were mobile and the swelling was bony hard on palpation with no obvious expansion of the lingual cortical plate. Pain and discomfort were felt in the TMJ region which was otherwise functionally normal.

**Radiographic Features**

The preoperative radiograph revealed an ill-defined radiolucency at the left angle of the mandible which extended anteriorly till the distal root of the second molar. Posteriorly and inferiorly it extended till the lower and posterior borders of the ramus respectively. Loss of lamina dura was observed around the distal root of the third molar. The adjacent teeth were normal.

**CT Scan Report**

Serial transaxial sections (Plain and contrast) from the hard palate to the symphysis of a mandible employing 5 mm slice of coronal section were performed. 5 ml of angiograffin 65% was used for contrast enhancement.

*Observations:* An expensive soft tissue density mass involving the body and ramus of the left mandible was seen measuring 4.4 × 4.0 cm enhancing from 68 to 143 HU. The lateral expansion of the mass was observed more than the medial expansion (Fig. 3). The mandible on the right side and the symphysis region appeared normal. Left maxillary and left posterior ethmoidal sinusitis was evident. There was no evidence of fluid or mucosal thickening (Figs 1 and 2).

*Impression:* CT features were suggestive of soft tissue density mass involving the ramus and body of the left mandible. The mass appeared diffuse and was suspected to be malignant.

**Hematological Investigations**

Hb—12%, TC—6,800 cells/mm³, ESR—90 mm/hr, PCV—38%, BT—1 min, CT—3 mins.
- DLC-Neutrophil—62%.
- Lymphocytes—34%.
- Eosinophils—2%.
- Monocytes—2%.
- Basophils—0%.
- HIV/HBS Ag—Negative.
- RBS—100 mg%.
- Blood group: A, Rh Type—Negative

**Biochemical Investigations**

- Alkaline phosphate—83.2 IU/L
- Calcium—9.0 mg%

**Histopathology**

Histopathological examination of the lesion revealed a characteristic picture of an infiltrating neoplasm. The tumor cells were relatively uniform and mitotic figures were not pronounced. Pleomorphic fibroblast like spindle cells were seen predominantly and they were interspersed between interlacing bundles of collagen fibers that were arranged perpendicular to each other. This gives the characteristic ‘Herringbone’ pattern (Fig. 4).

The pleomorphic cells also showed large irregularly outlined vesicular nuclei with scanty cytoplasm. They were closely packed with very little intercellular substances. In certain areas dysplastic cells were separated by thick bundles of collagen fibers that appeared to be hellenized (Fig. 5). The features observed were suggestive of fibrosarcoma.

**TREATMENT**

Tumor resection which included a left disarticulating-hemimandibulectomy via a modified submandibular approach. The lesion was submitted for histopathological examination.

*Fig. 1:* Extraoral examination revealed a diffuse swelling measuring about 5 × 3 cm on the left side of the ramus of the mandible (preoperative)

*Fig. 2:* CT-transaxial sections: soft tissue density mass involving the ramus and body of the left mandible
Incision with a lip-split extension was performed under general anesthesia (Figs 6 and 7). Recovery was uneventful and healing was satisfactory.

At the 5th month periodic review recurrence was observed in the cheek (Fig. 8). Tumor excision was followed by reconstruction of the defect with a temporalis myofascial flap.
flap as a second stage surgery. Subsequently the patient
was started on chemotherapy (Inj. Doxycrubicin 60 mg, Inj
Cyclophosphamide 700 mg and Inj Vincristine) which was
administered in 5 cycles at 3 weeks intervals.

**DISCUSSION**

Fibrosarcoma is a malignant mesenchymal tumor of
fibroblast. Although it can occur in any location, the bone
extremities are the most commonly affected sites. Primary
fibrosarcomas are rare in mandible which is a common sight
in the jaws10 (Table 1). Intraosseous fibrosarcomas may
develop industrially or possibly periosteally, the latter
affecting bone by spread from adjacent soft tissue to present
a clinical and radiographic appearance of primary bone
lesion.11 A fibrosarcoma arising in the region of a dental
extraction has been reported12 in the past. Our case report
also coincides with the history of extraction. However,
others accept that the fibrosarcoma of bone as a distinctive
lesion can arise in pre-existing benign lesions such as
ameloblastic fibroma, chronic osteomyelitis, Paget’s disease,
fibrous dysplasia, and giant-cell tumor of bone.13 Typically,
the tumor presents with swelling, associated with pain and
paresthesia.10,14-16 Radiographically, fibrosarcoma often
appears as a purely osteolytic lesion without calcification
and with poorly defined, irregular margins if it has arouse
intraosseously. There is usually destruction of the cortical
plates without expansion,17 and the lesion may be
misdiagnosed as an odontogenic abscess or cyst. The roots
of the adjacent teeth may or may not show resorption.17

The overall 5 years survival rate for fibrosarcoma of
bone has been reported to be between 28.7 and 34%.3,4
Periosteal fibrosarcoma has a better prognosis than that of
medullary origin, with 5 years survival rates of 52 and 27%
respectively.3 Similar data exist for fibrosarcoma of the bone
of the head and neck. Reported 5 years survival rates are
38% for periosteal fibrosarcoma and 27% for medullary
fibrosarcoma. Adequate resection appears to be the most
important factor in treatment.3,4,18

The exact cause of fibrosarcoma is not entirely
understood;19 however, studies have indicated that genetic
alterations may play a role. A chromosomal rearrangement
has been found in some fibrosarcomas.20,21 Radiation
therapy is considered in cases where resection is impossible
and chemotherapy is used as a palliative measure where
the chance for survival is obscure.21,22

This case report highlights the importance of early
investigation of symptoms that are chronic in nature. The
dental surgeon must be suspicious of an underlying disease
process when there is no relief of symptoms even after the
institution of basic treatment. Any complaints of pain,
loosening of teeth, paresthesia, and sudden denture
instability should be scrutinized carefully. This case
classically demonstrates the importance of postoperative
checkup following extraction of teeth in cases where the
loosening of teeth occurs in a young adult without a
demonstrable etiology.

Histopathologically the tumor may not show
characteristic features of fibrosarcoma in all the cases. The

<table>
<thead>
<tr>
<th>Years</th>
<th>Authors</th>
<th>Number of case</th>
<th>Site</th>
<th>Treatment</th>
<th>Recurrence</th>
<th>Follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>2008</td>
<td>Gosau et al</td>
<td>01</td>
<td>Mandible</td>
<td>Surgery</td>
<td>No</td>
<td>3 yrs</td>
</tr>
<tr>
<td>2007</td>
<td>Orhan et al</td>
<td>01</td>
<td>Mandible</td>
<td>Surgery + RT + CT</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>2006</td>
<td>Borges S et al</td>
<td>01</td>
<td>Mandible</td>
<td>Radical surgery</td>
<td>No</td>
<td>1yr 9 months</td>
</tr>
<tr>
<td>2005</td>
<td>Pereira et al</td>
<td>01</td>
<td>Mandible</td>
<td>Radical surgery</td>
<td>No</td>
<td>36 months</td>
</tr>
<tr>
<td>2003</td>
<td>Yamaguc HI et al</td>
<td>03</td>
<td>Mandible</td>
<td>Surgery</td>
<td>No</td>
<td>9 yrs</td>
</tr>
<tr>
<td>1998</td>
<td>L. Lo Muzio et al</td>
<td>01</td>
<td>Mandible</td>
<td>Radical surgery</td>
<td>No</td>
<td>4 yrs</td>
</tr>
<tr>
<td>1997</td>
<td>Lillenget et al</td>
<td>01</td>
<td>Mandible</td>
<td>Surgery + RT</td>
<td>Local + lung</td>
<td>21 yrs</td>
</tr>
<tr>
<td>1990</td>
<td>Sadoff et al</td>
<td>01</td>
<td>Mandible</td>
<td>Surgery</td>
<td>Local</td>
<td>NA</td>
</tr>
<tr>
<td>1989</td>
<td>Moloy et al</td>
<td>01</td>
<td>Mandible</td>
<td>Surgery</td>
<td>Local + regional</td>
<td>6 months</td>
</tr>
<tr>
<td>1986</td>
<td>Taconis et al</td>
<td>14</td>
<td>Mandible</td>
<td>Surgery + RT</td>
<td>Local + lung</td>
<td>NA</td>
</tr>
<tr>
<td>1985</td>
<td>Handlers et al</td>
<td>01</td>
<td>Mandible</td>
<td>Surgery + RT + CT</td>
<td>Local</td>
<td>15 months</td>
</tr>
<tr>
<td>1985</td>
<td>Zachariades et al</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>1984</td>
<td>Slootweg et al</td>
<td>07</td>
<td>Mandible</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>1979</td>
<td>Lam et al</td>
<td>03</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>1979</td>
<td>Ferullo et al</td>
<td>01</td>
<td>Mandible</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>1976</td>
<td>Looser et al</td>
<td>04</td>
<td>Mandible</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>1976</td>
<td>Jeffree et al</td>
<td>07</td>
<td>Mandible</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>1975</td>
<td>Huvos et al</td>
<td>12</td>
<td>Mandible</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>1975</td>
<td>Haidar et al</td>
<td>01</td>
<td>Mandible</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>1971</td>
<td>Van Biarcot et al</td>
<td>13</td>
<td>Mandible</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>1971</td>
<td>Jochimsen et al</td>
<td>01</td>
<td>Mandible</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>2011</td>
<td>Monaly Uwanati et</td>
<td>01</td>
<td>Mandible</td>
<td>Surgery + CT</td>
<td>Local</td>
<td>NA</td>
</tr>
<tr>
<td>2012</td>
<td>Basavaraj KF et al</td>
<td>01</td>
<td>Mandible</td>
<td>Surgery + CT</td>
<td>Local</td>
<td>NA</td>
</tr>
</tbody>
</table>
appearance varies with the level of differentiation, although well differentiated tumors seem to be more common. In the well differentiated tumor the fibroblasts are regular in shape with evenly staining nuclei while the anaplastic variants are difficult to diagnose as in the case of other malignant connective tissue tumors. Advanced techniques like immunohistochemistry is advisable in such cases.

SUMMARY
A case of a fibrosarcoma of the mandible is presented. The patient had a 6 months history of chronic symptoms in the area of the tumor before a formal diagnosis of fibrosarcoma was made and subsequent surgical ablation was carried out.

The prognosis for fibrosarcoma is poor even with early diagnosis and radical surgery.

REFERENCES

ABOUT THE AUTHORS
KF Basavaraj (Corresponding Author)
Lecturer, Department of Oral Pathology and Oral Biology, Faculty of Dentistry, Al-Jabal Al-Gharbi University, Gharian, Libya, e-mail: dr_basu78@yahoo.com

AU Madihalli
Lecturer, Department of Maxillofacial Prosthodontics, Faculty of Dentistry Al-Jabal Al-gharbi University, Gharian, Libya

Abdul Mujeeb
Associate Professor, Department of Operative Dentistry, College of Dentistry, Taibha University, Al-Madinah Al-Munawarah, Saudi Arabia

Samir Mansuri
Assistant Professor, Department of Oral and Maxillofacial Surgery College of Dentistry, Taibha University, Al-Madinah Al-Munawarah Saudi Arabia

Mohammed Abid Hussain
Professor, Department of Maxillofacial Prosthodontics and Implantology, College of Dentistry, Taibha University, Al-Madinah Al-Munawarah, Saudi Arabia