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CASE REPORT

Advancing Mandibular Swelling: A Diagnostic Dilemma

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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.

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INTRODUCTION

The jawbones can be sites of various neoplastic conditions.[1] Given the variety of processes affecting this particular anatomical area, formulation of a precise diagnosis can be challenging for clinicians.[2] 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.[3-5] 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.[6] 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.[7] 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.[8] 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.[9]

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/mm^3, 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

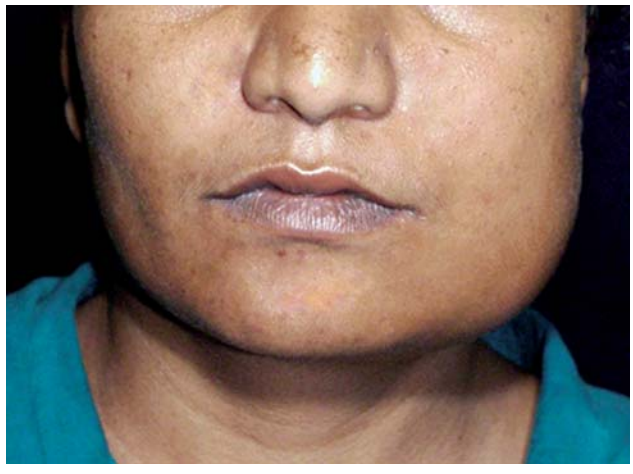

Fig. 1: Extraoral examination revealed a diffuse swelling measuring about 5 × 3 cm on the left side of the ramus of the mandible (preoperative)

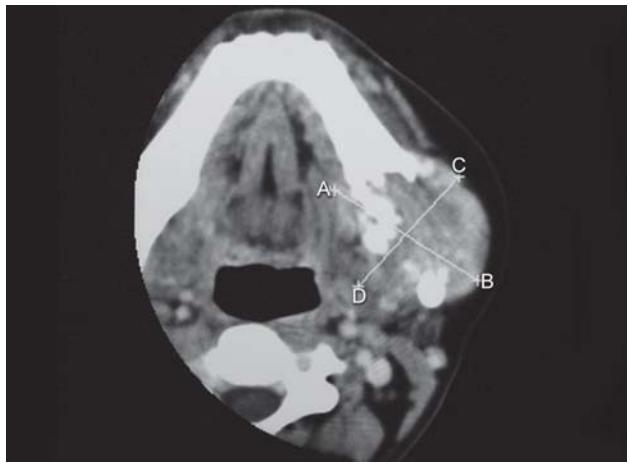

Fig. 2: CT-transaxial sections: soft tissue density mass involving the ramus and body of the left mandible

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

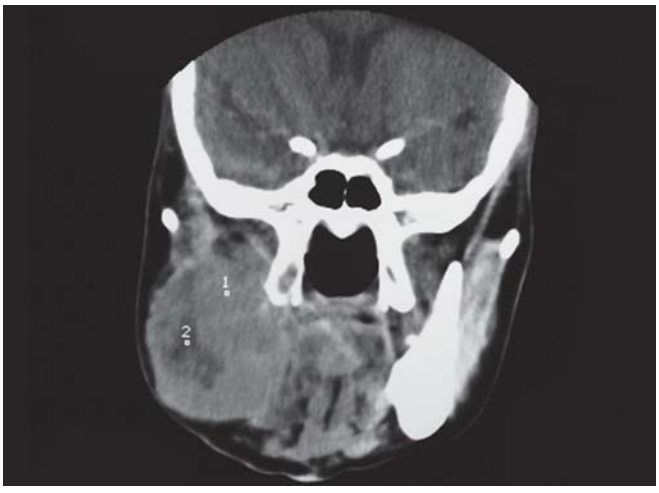

Fig. 3: CT—The lateral expansion of the mass was observed more than the medial expansion

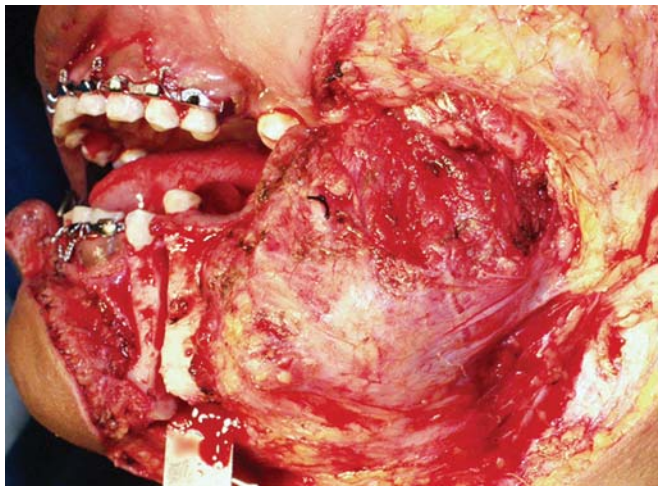

Fig. 6: Modified submandibular incision with a lip-split extension

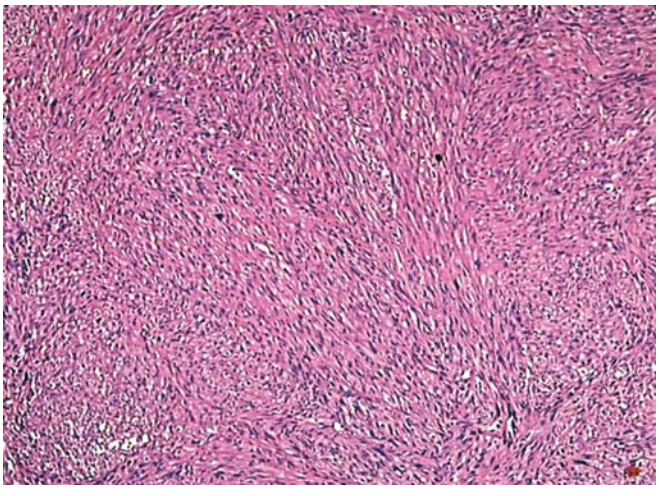

Fig. 4: Homogeneous spindle shaped cells arranged in a herringbone pattern

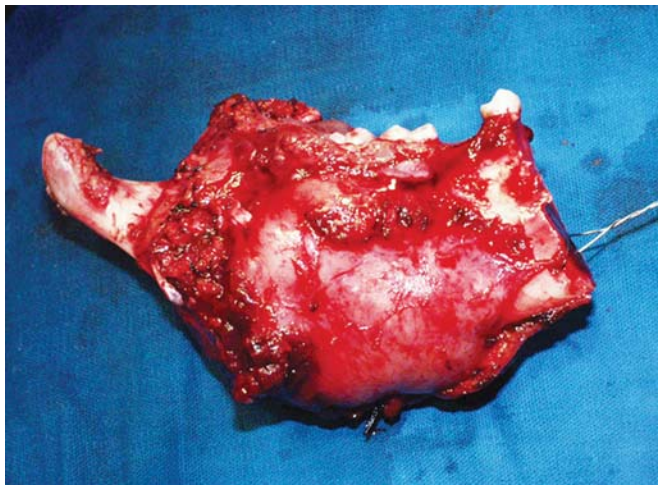

Fig. 7: A left disarticulating hemimandibulectomy

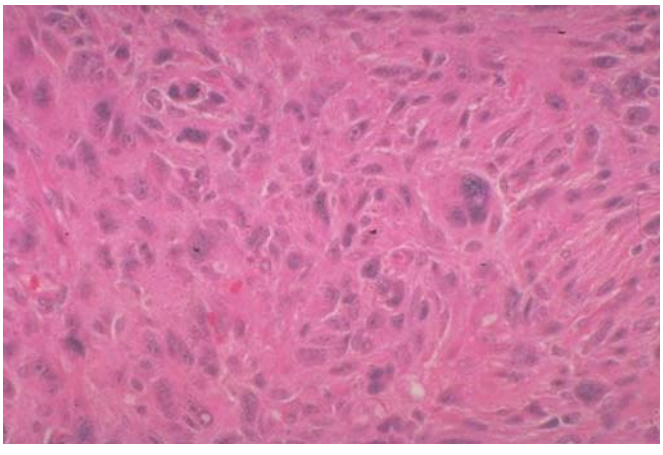

Fig. 5: Tumor cells showed large irregularly outlined vesicular nuclei with scanty cytoplasm, closely packed with very little intercellular substances

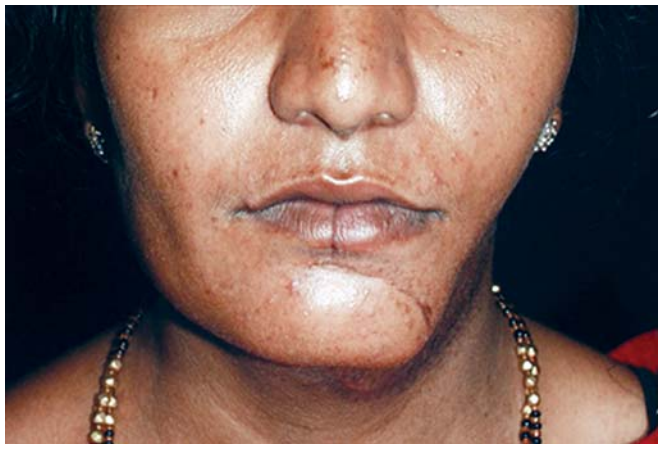

Fig. 8: Postoperative reconstruction of the defect with a temporalis myofascial flap

flap as a second stage surgery. Subsequently the patient was started on chemotherapy (Inj. Doxyrubicin 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 jaws[10] (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 reported[12] 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 1: Cases of primary fibrosarcoma of mandible reported in the literature

Years	*Authors*	*Number of case*	*Site*	*Treatment*	*Recurrence*	*Follow-up*
2008	Gosau et al	01	Mandible (1)	Surgery	No	3 yrs
2007	Orhan et al	01	Mandible (1)	Surgery + RT + CT	NA	NA
2006	Borges S et al	01	Mandible (1)	Radical surgery	No	1yr 9 months
2005	Pereira et al	01	Mandible	Radical surgery	No	36 months
2003	Yamaguc HI et al	03	Mandible	Surgery	No	9 yrs
1998	L.Lo Muzio et al	01	Mandible	Radical surgery	No	4 yrs
1997	Lillenget et al	01	Mandible	Surgery + RT	Local + lung	21 yrs
1990	Sadoff et al	01	Mandible	Surgery	Local	NA
1989	Moloy et al	01	Mandible	Surgery	Local + regional	6 months
1986	Taconis et al	14	Mandible (10)	Surgery + RT	Local + lung	NA
1985	Handlers et al	01	Mandible	Surgery + RT + CT	Local	15 months
1985	Zachariades et al	NA	NA	NA	NA	NA
1984	Slootweg et al	07	Mandible (5)	NA	NA	NA
1979	Lam et al	03	NA	NA	NA	NA
1979	Ferulito et al	01	Mandible	NA	NA	NA
1976	Looser et al	04	Mandible (4)	NA	NA	NA
1976	Jeffree et al	07	Mandible (4)	NA	NA	NA
1975	Huvos et al	12	Mandible (10)	NA	NA	NA
1975	Haidar et al	01	Mandible	NA	NA	NA
1971	Van Blarcom et al	13	Mandible (13)	NA	NA	NA
1971	Jochimsen et al	01	Mandible	NA	NA	NA
2011	Monaly Uwanati et al	01	Mandible	NA	NA	NA
2012	Basavaraj KF et al	01	Mandible	Surgery + CT	Local	NA

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.

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