CASE REPORT: Desmoplastic Fibroma of the Mandible in a child presenting with TMJ dysfunction

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Abstract

Background: Desmoplastic fibroma of bone is a rare intraosseous benign but locally aggressive tumor of connective tissue origin. The lesion may affect the metaphyses of the long bones but it may also involve the skull bones and more specifically the mandible, with most lesions appearing in the ramus-angle area. Treatment is surgical although additional chemotherapy or radiation has been applied. **Case report:** A 10-year-old boy initially presented with restriction and deviation of mouth opening. Clinical and radiological examination revealed a tumor-like lesion of the mandible extending into the soft tissues, which on biopsy proved to be a desmoplastic fibroma. **Treatment:** The surgical treatment included peripheral ostectomy of the mandible, via an intraoral approach, for the removal of the lesion and restoration of the bone defect with an iliac bone autograft. **Follow-up:** The high recurrence rate of this type of lesion, demands a strict follow-up schedule. In the case presented, 5 years post-operatively, there are no signs of local recurrence. **Conclusion:** Changes in mouth opening when not attributed to obvious reasons, such as trauma, should make a dentist suspicious and lead to further investigation. In the case presented, a central lesion was revealed in the radiograph and the patient was referred and treated early. Extended surgical removal of the tumor, with wide margins, proved to be the appropriate treatment.

Introduction

Desmoplastic fibroma of bone is a rare benign intraosseous tumor of connective tissue origin with locally aggressive behaviour, which may lead to infiltration of the surrounding soft tissues [Shafer et al., 1974; Templeton et al., 1997; Wippold et al., 2005]. The lesion may affect the metaphyses of the long bones but it may also involve the skull bones and more specifically the mandible, with most lesions appearing in the ramus-angle area [Hopkins et al., 1996; Said-A-Naief et al., 2006]. Desmoplastic fibroma was first described by Jaffe in 1958, while in 1965 it was first reported in the jaws of an 8-year old girl [Griffith and Irby, 1965].

Clinically, signs and symptoms of desmoplastic fibroma of the jaws are non-pathognomonic, although asymptomatic swelling, intermittent pain, limited mouth opening or some degree of functional impairment may be found [Batsakis, 1984; Said-Al-Naief et al., 2006]. Radiographically, the lesion may present as a well-defined expansible unilocular or complex multilocular radiolucent area with a trabeculated appearance. The appearance on Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) is homogeneous and cortical thinning or cortical breakthrough with a soft-tissue component is often seen [Wippold et al., 2005; Said-Al-Naief et al., 2006].

Histologically, fragments of soft tissue composed of fibroblasts and exhibiting abundant collagen production may be seen. The fibroblastic nuclei are thin and elongated without evidence of nuclear atypia or increased mitotic activity [Herford et al., 2001; Sapp et al., 2004].

Most clinicians seem to favour surgical resection of the lesion with a wide margin, in order to avoid recurrence [Hopkins et al., 1996; Iwai et al., 1996; Templeton et al., 1997; Said-Al-Naief et al., 2006]. A case of a desmoplastic fibroma of the mandible in a 10-year old boy that was misdiagnosed as TMJ dysfunction is presented here. The clinical evaluation, the surgical treatment and the follow-up modalities are described and discussed.

Case report

A 10-year-old boy was referred to the Dept.of Oral and Maxillofacial Surgery of the Children's Hospital "P. & A. Kyriakou", in Athens, Greece, in November 2002. He complained of progressive difficulty in opening his mouth which had occurred over the previous 2 months. The referral letter of his general dental practitioner mentioned a diagnosis of TMJ dysfunction. Initial clinical examination revealed no extraoral abnormality, the maximal inter-incisal mouth opening was limited to 19 mm and a slight deviation to the right side during opening was noticed (Fig 1). Intraorally no bone swelling could be seen nor palpated. There was no paraesthesia or anaesthesia of the inferior alveolar nerve and no palpable cervical lymphadenopathy. The young patient was healthy overall and there was no history of trauma, infection or past TMJ dysfunction.

Following an orthopantomogram, a poorly circumscribed radiolucent lesion in the left angle region of the mandible, distally to the first permanent molar and including the bud of the second molar could be seen (Fig 2a). Further radiological

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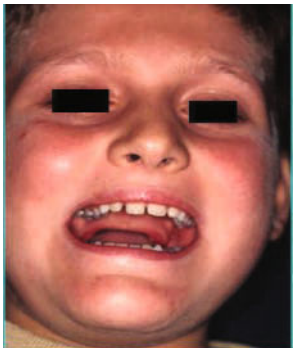
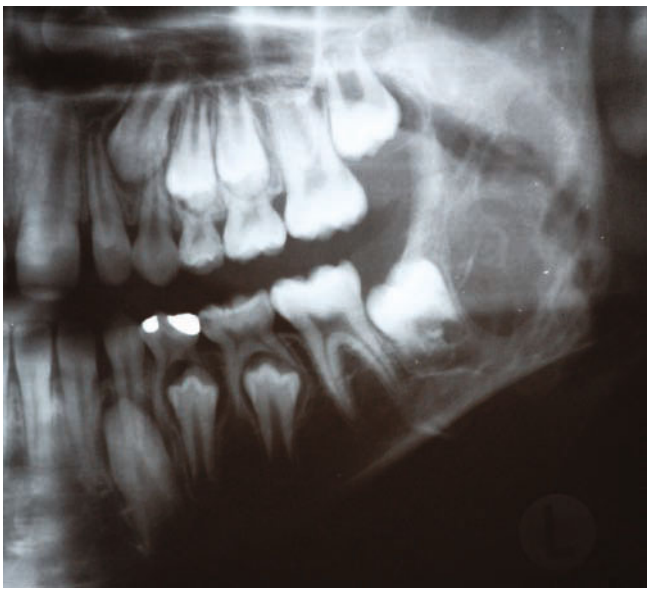


Figure 1. Extra-oral photograph showing limitation and deviation of the mouth opening.

Figure 2: a. Panoramic radiograph of the case showing poorly circumscribed radiolucent lesion of the left angle of the mandible; **b.** MRI showing the extent of the lesion, note the correlation with the median pterygoid muscle.



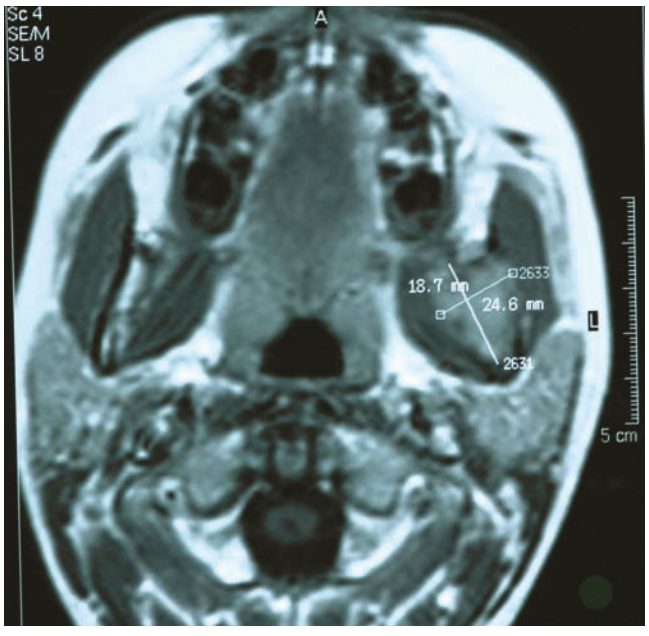


Figure 3. Photographs taken during the surgical procedure. **a.** Intra-oral view of the case; **b.** Immediate reconstruction of the defect with autogenous bone graft.

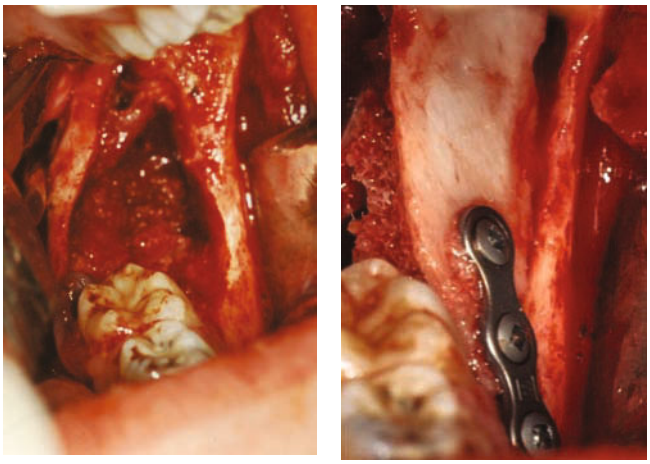
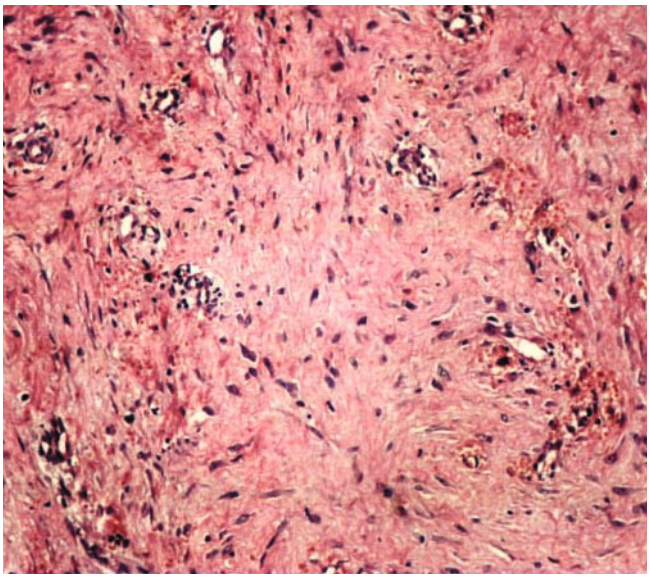


Figure 4. Histological section showing the lesion is composed of intersecting bundles of fibrous tissue without any atypia or mitosis. Scattered inflammatory cells are observed (H/E, original magnification x100)



examination included CT, where a lytic process with perforation of the lingual cortex in the angle region was obvious and subsequent MRI where a 18.7x24.5 mm bony lesion of the mandibular angle, eroding the cortex and infiltrating the associated medial pterygoid muscle, was revealed (Fig 2b).

Differential diagnosis of the findings included benign fibrous lesions of bone-like non-ossifying fibroma and odontogenic fibroma, myxoma and chondromyxoid fibroma or even fibrosarcoma. Histological findings of incisional biopsy from the left retromolar area were compatible with desmoplastic fibroma.

Treatment

Surgical removal of the lesion via an intraoral approach was performed under general anaesthesia. Large mucoperiosteal flaps were elevated both bucally and lingually, until the lesion was well exposed. At the lingual side of the mandible, where the lesion was mostly expanded, elevation of soft tissues went as deep as the mylohyoid muscle. The tumor was removed by performing a peripheral ostectomy of the mandible which included the osseous part of the lesion with wide margins. Ostectomy was more extensive on the lingual side, preserving a thin bone wall at the buccal side which included the bundle of the lower inferior nerve. The tumor was excised with wide margins in the soft tissues at the lingual side as well. The specimen, sized 4x3x2cm, included the bud of the second molar. The boney defect was restored at the same stage, with an autogenous iliac crest free bone graft fixed with a 4-hole titanium mini plate (Fig 3a and 3b). Careful suturing followed. Post-operative recovery was without any complications. Mouth opening ameliorated after surgery and was back to normal in 2 weeks.

Histological evaluation of the specimen confirmed the initial biopsy diagnosis. Intersecting bundles of uniform spindled or elongated fibroblasts, in a dense collagenous background were found. The fibroblastic nuclei were thin and elongated, without any atypia or increased mitotic activity. Scattered inflammatory cells were observed. There was no osteoid formation and the lesion infiltrated the adjacent tissues as there was no capsule. Immunohistochemically fibroblasts showed positive results for Vimentin and S-100 protein antibodies. No signs of EMA and Ker AE1/AE positive epithelial elements were found in the background. The combined histological and immunohistochemical findings were compatible with desmoplastic fibroma of the mandible (Fig 4).

Follow-up

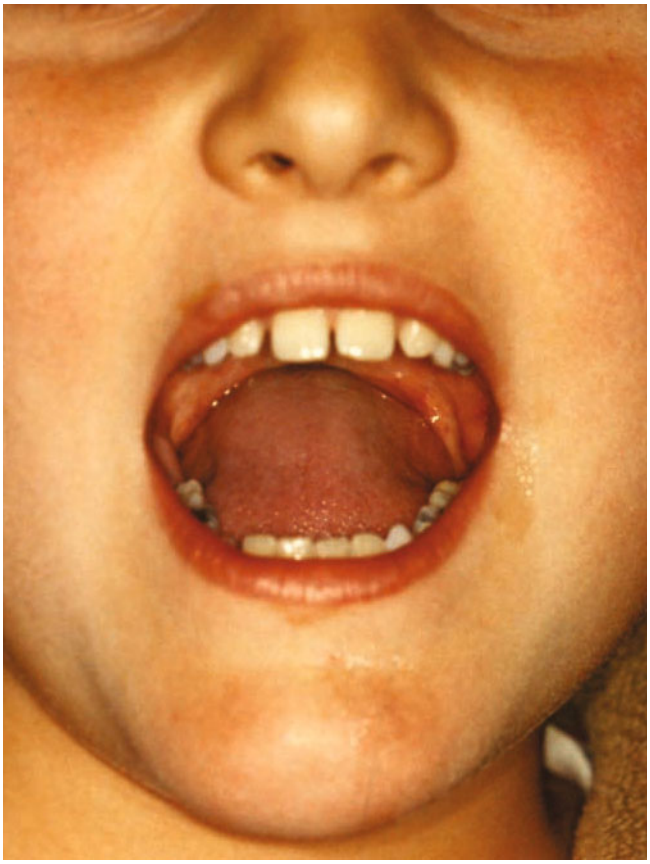
This was monthly for 4 months post-operatively, every 3 months for 2 years and then every 6 months onwards (Fig 5a and 5b). Healing of the mandible 5 years post-operatively, is complete without clinical or radiological signs of recurrence.

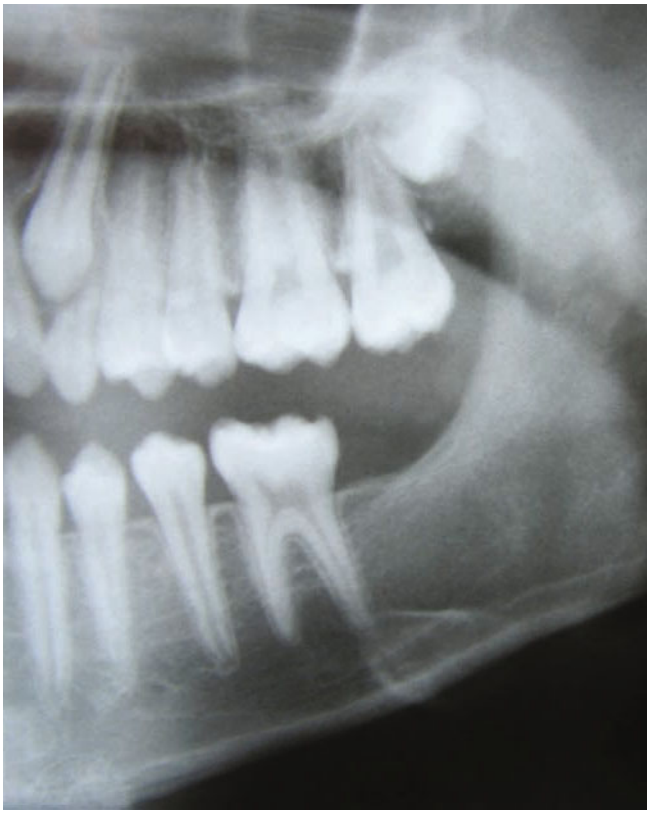
Discussion

The case presented was the only desmoplastic fibroma treated in our Department (referral age up to 14-years-old), over the past 7 years and it represented 0.9% of all bone tumors (1/110 tumors) and 1% of all benign bone tumors (1/99) treated. The clinical symptoms of the young patient, being impairment of mouth opening and deviation of the mandible only, without pain or palpable swelling, initially misled the first clinician who examined the patient, as they were compatible with condyle dysfunction.

Subsequent careful evaluation of the clinical and imaging findings established the diagnosis, size and type of the tumor, while treatment was scheduled only after biopsy which clarified exactly its nature. An interesting clinical finding

Figure 5. Post-operative. **a.** Full mouth opening at 1 year; **b.** Panoramic view at 3 years post-operatively.





was also the deviation of the mandible towards the healthy side, a fact that could be attributed to the implication of the mylohyoid muscle which takes part in the opening motion. Histological examination revealed absence of any atypia or increased mitotic activity excluded malignancy, while the absence of osteoid formation and the prominent collagenization, helped distinguish the lesion from non-ossifying fibroma or odontogenic fibroma. Intraosseous odontogenic or ameloblastic fibroma was also excluded. An absence of capsule and infiltration of the adjacent tissues are considered as landmarks in diagnosing desmoplastic fibroma, whilst immunohistochemistry is not always helpful [Herford at al., 2001, Said-Al-Naief et al., 2006].

Parameters of the present case, such as the age of the patient, the location and the clinical manifestations were similar to those previously reported [Griffith and Irby, 1965; Batsakis, 1984; Hopkins et al., 1996; Said-Al-Naief et al., 2006]. Regarding treatment, wide local excision of the tumor was performed, taking into account its locally aggressive nature, as has been suggested before [Hopkins et al., 1996; Iwai et al., 1996; Said-Al-Naief et al., 2006]. Treatment with local curettage only has been associated with high recurrence rates up to 70%, while the recurrence rate after excision or enucleation ranges from 20-40% [Hopkins et al., 1996; Said-Al-Naief et al., 2006]. Identification of the macroscopic margins during surgery has been reported to be sometimes difficult because of the absence of a capsule [Iwai et al., 1996]. Where there are signs of tumor spreading through bone with invasion of the surrounding soft tissue exist, even wider surgical margins of 2 to 3 cm have been recommended [Kwon et al., 1989; Hopkins et al., 1996].

Radiation therapy has been suggested to slow the growth rate of lesions that have been sub-totally excised, while chemotherapy has been used as an adjunct to total resection [Hopkins et al., 1996]. None of those therapeutic approaches was selected in the present case, as they were considered quite severe for such a benign tumor. Repair of the large boney defect with free bone graft from the iliac crest was decided, as the remaining mandibular bone was thin and prone to fracture [Keller and Triplett, 1987; Boustred et al., 1997]. Restoring the defect at the same operation gave the opportunity for the young patient to heal uneventfully without major changes in appearance and possible psychological implications.

Our patient has shown no clinical or radiographic evidence of recurrent tumor during more than 5 years of follow-up. Recent literature considers adequate follow-up period to be no less than 3 years, given the tendency for recurrence [Said-Al-Naief et al. 2006].

Conclusion

Desmoplastic fibroma of the mandible, although benign, may present with clinical symptoms related with its aggressive and invasive behavior. Dentists and Paediatric Dentists should have in mind that among other entities, desmoplastic fibroma of the mandible could present with restriction and deviation in mouth opening, leading to a misdiagnosis of TMJ dysfunction. Correlation of clinical, radiographic and histological findings is imperative for determining a definitive diagnosis and treatment plan. Whenever needed, immediate rehabilitation of the bone defect is to be recommended. Careful and long lasting follow-up is essential, due to potential recurrence.

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