Rare case of desmoid-type fibromatosis of the mandibular region in a child: diagnosis and surgical management

ABSTRACT

**Background** Desmoid-type fibromatosis is a broad group of rare disorders that originate from musculoaponeurotic structures. They represent less than 0.1% of all tumors and the annual incidence is 2-4 cases per million, with two peaks between 6 and 15 years of age and between puberty and age 40. They are rare in the oral and maxillofacial regions. Its pathogenesis is multifactorial. The treatment is still mainly represented, both in children and adults, by surgical excision.

**Case report** A case is reported of a 8-year-old girl with desmoid fibroma in the mandible who presented at the Department of Pediatric Dentistry with a swelling measuring 4x4 cm on the lower edge of the right mandible which had appeared a few months earlier and slowly developed. The family dentist had initially diagnosed it as an odontogenic abscess from the lower right deciduous molars, but the antibiotic therapy was unsuccessful. After x-ray examination, which showed a large osteolytic lesion, mandibular CT revealed a solid expanding mass. The child was referred to the Department of Paediatric Maxillofacial Surgery where the whole mass was surgically removed. At the 2-year follow-up no relapse was noted.

**Conclusion** This case stresses the importance, especially for paediatric dentists, of further diagnostic steps if suspect lesions do not heal after conventional treatment.

**Keywords** Children; Desmoid-type fibromatosis; Maxillofacial surgery.

Introduction

Desmoid tumours, currently named desmoid-type fibromatosis according to the latest WHO classification, is a broad group of rare disorders that originate from musculoaponeurotic structures. They represent less than 0.1% of all tumors and the annual incidence is 2-4 cases per million, with two peaks between 6 and 15 years of age and, especially in women, between puberty and age of 40; in children, the sex distribution similar. According to the site of onset, they are divided into abdominal, intra-abdominal and extra-abdominal desmoid, including facial desmoid fibromatosis. Primary desmoid-type fibromatosis in the oral and maxillofacial regions are rare, representing less than 3% of all the cases [Min et al., 2011; Abdelkader et al., 2011].

Although desmoid-type fibromatosis is considered a benign tumour, it has a strong tendency for local infiltrative growth and recurrence, but it does not metastatise to other tissues and, from a histology standpoint, it does not show malignant features [Min et al., 2011; Meazza et al., 2010]. In fact, the tissue is poorly cellular, with ovoid or elongated nuclei; the cellularity, pleomorphism and mitotic activity, which are peculiar features of fibrosarcoma, are not present.

The pathogenesis of desmoids fibroma is multifactorial. Trauma and endocrine factors are indicated as causes of an incorrect regulation of the connective tissue, as well as the genetic predisposition. Even if this kind of tumour does not seem to have a familiar tendency, its incidence is higher in case of Gardner syndrome, familial desmoid tumour and familial adenomatous polyposis [Sharma et al., 2008; Wilks et al., 2001].

The treatment is still mainly represented, both in children and adults, by surgical excision, even if local recurrence rates after surgical resection range from 10% to 80%, depending on the negativity or positivity of margins. In children, associated radiation therapy is not recommended because of the functional morbidity and the risk of developing second malignancies [Meazza et al., 2010], while chemotherapy and non-cytotoxic agents are sometimes considered as alternative strategies.

We report a case of a 8-year-old girl with desmoids type-fibromatosis in the mandible region, discussing the therapeutic implications.

Case report

On October 2010 a 8-year-old girl, in good health, presented at the Department of Paediatric Dentistry exhibiting a swelling of 4x4 cm on the lower edge of the right mandible (Fig. 1).

The growth, which was altering the girl’s features and hard in consistency, was painful, deeply rooted and covered by normotrophic skin. It had appeared...
a few months before, and had a slow development. The family dentist had initially diagnosed it as an odontogenic abscess from the deciduous lower right molars, but the antibiotic therapy proved unsuccessful. A panoramic X-ray was taken (Fig. 2), followed by a mandibular CT scan (Fig. 3), which revealed a solid expanding lesion probably originating from the mandible, grown by extensive erosion of the mandible. At that point, the diagnostic hypothesis was a neuroectodermal tumour or a sarcoma. A biopsy was requested. The child was referred to the Department of Paediatric Maxillofacial Surgery where a biopsy of the lesion was performed under general anaesthesia, with the following histological result: “desmoid-type fibromatosis tumour with a scarcely cellular proliferation of fused elements immersed in a copious collagen meshwork rich in blood vessels. The proliferation contains fatty tissue and clusters of skeletal muscle cells”.

The whole mass was isolated from the mandible through submandibular surgical access and then removed; to achieve complete avulsion, resection was performed of the surrounding tissues of the mandible’s external cortex and bone contour from the presynphysial area to the ascending branch, including the submandibular cavity, with the sacrifice of a few dental germs. Given the extent of the surgery and the patient’s young age, no further action was taken except a strict follow-up. At the two-year follow-up, no relapse was noted.

Conclusion

This case underlines the importance, especially for the paediatric dentist, of further diagnostic steps if suspect lesions do not heal after conventional treatment. Moreover, it is important to paediatric dentists for the following reasons:

- It describes a case of a rare tumour of the mandible in a child.
- It underscores the importance of a thorough diagnosis of a suspect bone lesion referred to a paediatric dentist.
- It underlines the importance of the multidisciplinary approach (paediatric dentist, paediatric radiologist and paediatric maxillofacial surgeon).

References