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Infantile myofibromathosis of the maxilla. A case report

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Background. *Infantile myofibromatosis is a rare benign tumour in children. Its characteristic symptoms are firm masses in soft tissues, bones and visceral organs, and its common locations are head and neck. Three forms are well known: solitary, multicentric and visceral myofibromatosis. All have excellent prognosis, except the last one that may be lethal. Spontaneous regression can occur.*
Case report. *We present an unusual case of infantile myofibromatosis of the maxilla in an adolescent.*
Conclusions. *The infantile myofibromatosis should be managed with special caution because of the differential-diagnostic similarity with fibrosarcoma, leomyosarcoma, and histiocytosis.*

Key words: myofibromatosis; maxillary neoplasms; infant

Introduction

Infantile myofibromatosis (IM) is a rare benign tumor in children. It was first described as congenital fibrosarcoma. Later, sporadic cases were discussed, and in 1981, 61 cases were examined and named as IM. Three different forms were described: solitary, multicentric and visceral IM.[1]

Three quarters of soft tissue tumours in children and adolescents are benign, 95 % of them are fibromatosis, 80 % of fibromatosis are IM and 19 % aggressive desmoid fibromatosis.[2] The most frequent type is solitary IM. One third of these occur are in the head and neck.

Its aetiology is not known. First, it was believed that original cells are fibroblasts; but with the presence of desmins receptors in IM, this belief proved to be false. In fact, the original cells of IM growth are smooth muscular cells.[3] Genetic predisposition is likely since solitary form is twice as frequent in males than in females.

Clinically, IM is expressed by slowly growing painless firm solitary or multicentric nodes in the soft tissue, bones or visceral organs. Half of the cases develop in the dermis or subdermally.[4] Their size is few millimetres to few centimetres. Half of the cases are congenital, 90 % of all cases develop in first

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two years of life.[1] The symptoms are rare except when IM obstructs the visceral organs or nerves. Spontaneous ulcerations can also occur.

Pathohistological characteristics of IM are bundles of pinal cells – myofibroblasts, round and less differentiated cells may be found. In the centre, necrotic processes and calcinations are also seen. Mitotic activity is most diversified.

On the X-ray of the skeleton, the IM is seen as well differentiated osteolitic lesions with sclerotic margins. The IM may well be seen also by other diagnostic imaging techniques, such as CT and MR. Before final diagnosis, pathohistology of bioptic material should be performed. Differential diagnosis discovered some resemblances with fibrosarcoma, histiocytosis, and leiomyosarcoma; therefore, the misinterpretations are possible.

The visceral type is lethal in 75 % of cases in neonatal period because of an acute cardiopulmonary failure, haemorrhage or gastrointestinal obstruction,[1] whereas nonvisceral forms have excellent prognosis.

The conservative tumour excision is curative. Spontaneous regression may occur in the third of cases[4] 1 to 2 years;[5] therefore, the observation is a method of choice if IM is definitely pathohistologically conformed. The reported recurrence rates ranged from 7-10 % (1) to 31 % (4).[1,4]

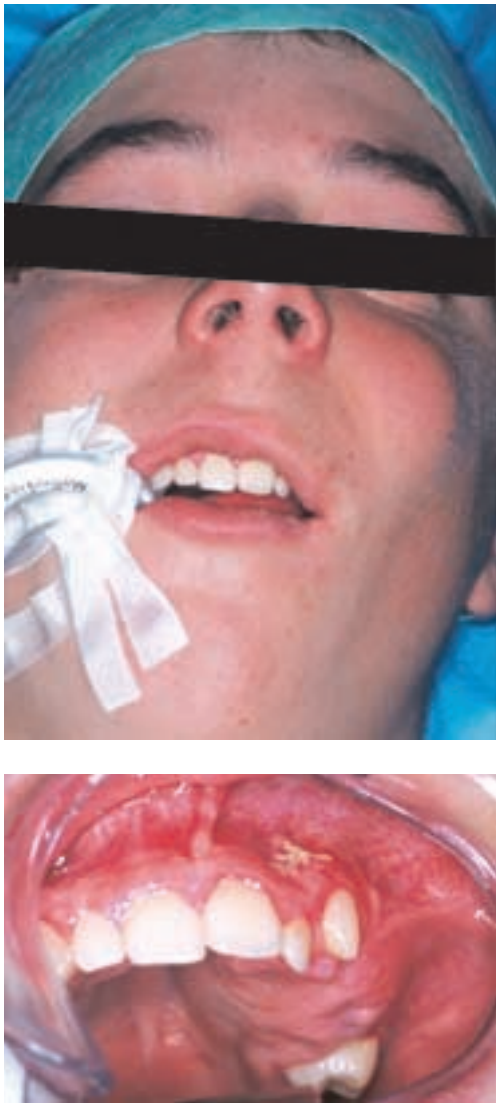


Figure 1a, 1b. The patient before surgery.

Case report

A fifteen-year-old boy came to our outpatients department because of a massive, firm oedema on the left cheek, which had been slowly progressing for 1 year. (Figures 1a, 1b) He as well as his parents connected it with the impacted upper canine tooth on the left that was operatively released by an oral surgeon one year ago. The X-ray of the teeth made at that period showed a tumour with a diameter of 2 cm in the alveoli of the maxilla. Medical history detected no genetic predisposition.

Clinical investigations confirmed a large tumour mass, which vaulted over the hard palate, alveolar and buccal tissues. The boy's face was strongly disfigured. The dermal and mucosal coating was intact. There were no other pathological changes in the oral cavity

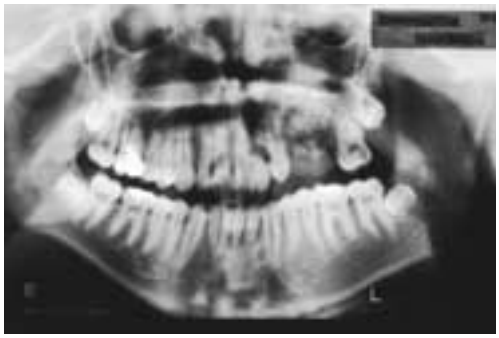
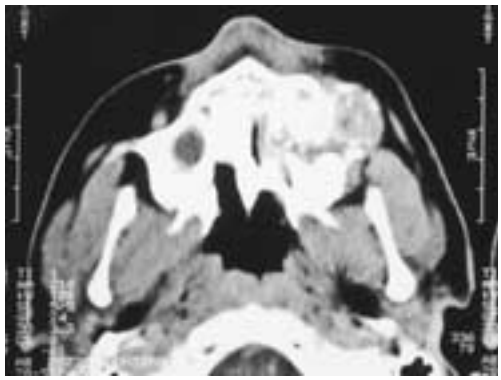


Figure 2. Panoramic X-ray.



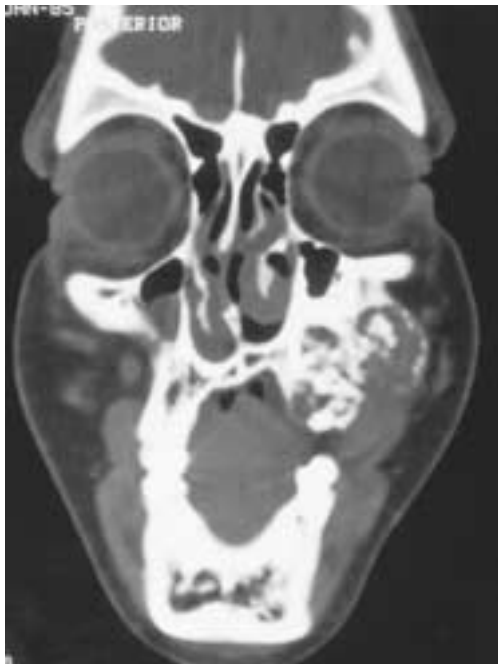


Figure 3a, 3b. CT scan of infantile myofibromatosis lesion in left maxilla before surgery.

and head, except the lacking permanent premolar and first permanent molar teeth in the left maxilla. From the medical history it is evident that they never existed.

X-ray showed significant changes in the bone of the maxilla, partly osteolytic partly calcinated. They spread over the alveoli from the canine tooth to maxillary tuber and invade the maxillary sinus too orbital floor (Figure 2). CT scan showed no invasion into the orbit, nasal cavity and pterigoids (Figures 3a, 3b). The serum alkaline phosphatase value was very high, the serum phosphate rate was also increased, whereas the values of the serum calcium and hormones were normal.

From the clinical viewpoint, it could be possible that the boy had a sarcoma or a rare odontogenic tumour because of the lacking teeth. We took the biotic material samples from three different parts. Pathohistologic results confirmed IM without mitotic activity and necrosis. The boy was operated on; the tumour was extirpated by conservative surgical approach. It size was minimally $7 \times 4 \times 4$ centimetres. After partial maxillectomy, the reconstruction was performed with local tissues. The final pathohistological findings were the same as those of previous biopsy. Moreover, the examination of IM tissue confirmed the presence of the ortotopic bone tissue.

No postoperative complications occurred. The disfigurement was restored, whereas the lacking the lacking alveolar bone and the teeth were replaced by obturator prosthesis.

A year and a half after surgery, the boy is without the local recurrence or recurrence elsewhere in the body (Figure 4a, 4b). The bone defect became smaller after the reparation of the marginal bone. After the boy's growth is completed, a reconstruction by autologous bone and dental implants is planned.

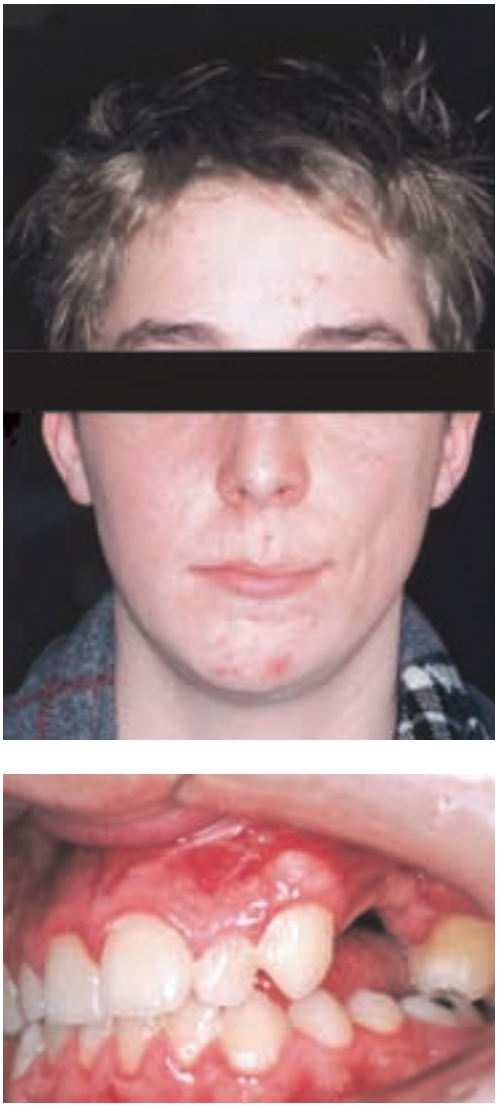


Figure 4a, 4b. The patient after surgery.

Discussion

IM is a rare, but important tumour in children and, because of its common localisation, it should be known to all the surgeons of the head and neck.

The spontaneous regression of IM is induced by programmed cell death – apoptosis.[6] This is probably the most significant example of this massively enhanced physiological mechanism. However, it does not occur in all cases, or it may be so slow that, in most cases, the conservative surgery is necessary. If its results are very mutilating, adjuvant chemotherapy may be applied.[4]

From the revised references, the maxilla, as in our case, is a rare localisation. The most frequent localisations are the dermis or subcutis.[4] Among the head bones, the calvarian bone is most frequently affected, whereas the temporal bone,[7] the orbita with the zygomatic bone8 and the nasal cavity with the inferior turbinate are less frequent localisations.[9]

What is also unusual in our case is the boy's age, which was relatively high for developing IM in spite of late diagnostic treatment. The teeth germs destruction is also most unclear. If the obstruction and necrosis had been the causes, it should have developed at the age of 4 to 6 years, well before the mineralization of the teeth germs occurred. In such circumstances it is unlikely that the tumour would grow so slow until the boy's age of 14 years and speeded-up so much in the last year. In our case, the differential diagnosis suspected of rare odontogenic tumours, which was exceptionality also in our case.

As with other fibrous tumours, IM is frequently misdiagnosed as malignancy, most commonly as fibrosarcoma.[10] It is also possible to mistake it for with leiomyosarcoma.[11] In case of the malignant fibrous tumour, surgery should be very radical and, if it was but a misdiagnosed IM, the morbidity unnecessary.

The recurrence and new growth of IM is possible also on localizations other than primary;[5] therefore, a long-term follow-up of these patients is recommended.

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