Oral and Maxillofacial Surgery Cases 2 (2016) 35–39

Contents lists available at ScienceDirect

Oral and Maxillofacial Surgery Cases

journal homepage: www.oralandmaxillofacialsurgerycases.com

Case report

Infantile myofibroma of the zygomatoco-maxillo-orbital complex: Case report with spontaneous regression

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ARTICLE INFO

Article history:
Received 12 January 2016
Accepted 5 November 2016
Available online 9 November 2016

Keywords:
Myofibroma
Infantile myofibroma
Myofibromatosis
Zygomatico-maxillo-orbital complex tumors
Head and neck

ABSTRACT

Introduction: Infantile myofibroma (IM) is a benign mesenchymal disorder. Although it is the most common tumor of infancy, it is a relatively rare pathological entity. IM occurs predominantly before the age of two. It is characterized by tumor formation in skin, muscle, viscera, bone and subcutaneous tissues. The tumor can occur in either solitary or multicentric forms. The solitary form without visceral involvement usually has a benign course. Male gender predominates in the solitary form.

Case presentation: We report a clinical case of an osseous infantile myofibroma of the zygomatico-maxillo-orbital complex in a 4 months old male. We also show the radiological spontaneous regression without surgical intervention. Clinical, radiological, and histopathological examinations established the diagnosis. Our treatment was conservative with clinical follow-up. No surgical intervention was carried out during the course of the disease. Significant spontaneous regression occurred after a year and was confirmed by CT scan.

Conclusion: Radiologically aggressive infantile myofibroma has been previously treated by surgical intervention. In this case report there was a significant spontaneous regression. Conservative treatment and follow-up may be an appropriate alternative.

1. Introduction

Infantile myofibroma is a benign mesenchymal tumor of infancy. It is characterized by tumor formation in skin, muscle, viscera, bone and subcutaneous tissue [11]. This tumor affects exclusively infants. Eighty-eight percent of the cases occurred before the age of two. Solitary forms have a male predominance [3]. One third of the cases involve the head and neck region.

The clinical heterogeneity and misleading histopathological appearance make the diagnosis difficult [8]. The oldest reported patient that presented with “infantile” myofibroma was 49 year-old female reported by Wolfe [13]. Foss reported 79 cases of myofibroma with different patterns including infantile or adult, and multicentric or solitary. The patients' ages at diagnosis ranged from birth to 84 years [5]. In his series the oldest patient was not clearly identified as an “infantile” myofibroma. All tumors are positive for vementin and alpha-smooth muscle actin [7]. We share our experience with a unique case

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http://dx.doi.org/10.1016/j.omsc.2016.11.001

of infantile myofibroma of the zygomatico-maxillo-orbital complex that spontaneously regressed without any surgical intervention, sparing the child from tumor extirpation as well as reconstructive procedures despite the large size of the tumor.

2. Case report

We describe a case of a male child born with uneventful vaginal delivery with normal Apgar score and no complications at birth. This boy was referred to the pediatrician at the age of 4 months because of progressive swelling of the right cheek with ectropion of the lower eyelid. The differential diagnoses of this swelling included: hemangioma, vascular malformation, leiomyoma, rhabdomyosarcoma and osseous benign or malignant tumors. An ultrasound detected a mass of 28 × 30 × 20 mm at the level of zygomatic bone.

An MRI and a CT scan were performed and showed a 27 × 28 × 22 mm mass in the right zygomatic bone with erosions and an expanding osseous tumor beyond the normal borders of the zygoma involving the zygomatico-maxillo-orbital complex (Figs. 1 and 2).

A needle biopsy was performed and showed benign myofibroblastic proliferation with osseous involvement and confirmed the diagnosis of infantile myofibroma (Fig. 3).

At the age of 8 months our craniofacial team received a referral from a pediatric ENT surgeon. On physical examination, the mass was slightly tender but no further increase in swelling was noted. There was no involvement of facial musculature. Extra-ocular muscles and ophthalmological evaluation were normal. No intra-oral lesions were present. The occlusion was normal and airway was patent. No other lesions identified. At that time surgery was preliminary scheduled but a CT scan was ordered for further evaluation and treatment planning. The new CT scan demonstrated a significant reduction of tumor size to 20 × 20 × 17 mm with increased mineralization of the tumor borders. At that stage surgical intervention was abandoned and the patient was followed up in the clinic (Fig. 4).

Clinical evaluation 6 months later showed further reduction of the mass. CT scan at 2 years and 5 month of age showed further reduction of the size of the tumor (13 × 10 × 10 mm) with additional mineralization and remodeling of the zygomatico-maxillo-orbital complex towards the normal anatomy (Fig. 5).

3. Discussion

Bone masses and osseous lesions in infancy can be challenging to diagnose. Differential diagnoses can include benign or malignant tumors, which need to be treated differently. The correlation between clinical and histological examination is essential for accurate diagnosis and treatment.

Stout initially described infantile myofibroma in 1954. He termed the disorder “congenital generalized fibromatosis” [15].

Infantile myofibroma (IM) is an extremely rare tumor although considered the most common tumor of infancy. Chung and Enzinger evaluated infantile myofibroma and concluded that 60% of the cases were noted at birth or shortly thereafter.

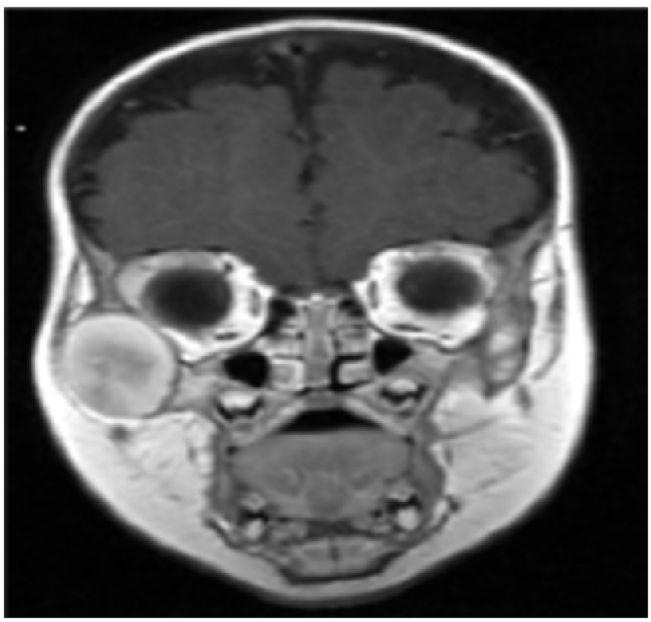

Fig. 1. MRI at 10 months of age showing the tumor, 27 × 28 × 22 mm in size.