

Case Report

Infraorbital Myofibroma of Infra-orbital Region: A Rare Case Report

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Abstract

Introduction

Infantile myofibroma is a rare tumor of myofibroblastic origin that can affect soft tissue, bones, or internal organs. The aim of the current study is to report a rare case of a solitary infra-orbital infantile myofibroma.

Case presentation

An 11-month-girl presented with progressive swelling of the left infra-ocular region for two weeks. On examination, there was a well-defined swelling with a localized border that was firm, non-mobile, tender, fixed to the underlying bone, and non-adherent to the superficial skin. Under general anesthesia, the right infraorbital mass was excised. The wound was closed by layers, and the histopathological examination revealed a 3 cm myofibroblastic mass that invaded the margins at multiple spots.

Conclusion

Infraorbital myofibroma is an extremely rare type of myofibroma. Excision with primary closure is the definitive management therapy.

1. Introduction

Myofibroma is a rare tumor of myofibroblastic origin that can affect soft tissue, bones, or internal organs as a single or multiple nodules. Although it is most common in children under the age of two years (1 in 140,000 newborns), with two-thirds of them present at birth, it can affect people of all ages. It is twice as common in females as in males [1]. Infantile myofibroma (IM)

is a nonencapsulated, non-metastasizing, locally infiltrative lesion considered to be part of the fibrous proliferations of infancy. IM can present as a solitary myofibroma, a multicentric myofibroma in soft tissues, or a multicentric myofibroma with visceral involvement. Despite the fact that it is a relatively rare lesion, it can be found in the scalp, forehead, parotid region, and oral cavity [2].

The etiology of IM is not well understood, as most cases of infantile myofibromatosis occur randomly. Rare familial cases have been identified, and genetic changes (mutations) in two different genes (PDGFRB and NOTCH3) have been found to be the source of the tumor in the vast majority of cases. While IM within the head and neck is extremely rare, it appears to have a tendency to develop around the orbit in the few cases reported in the genuine literature [3,4].

The purpose of the current study is to report a rare case of solitary infra-orbital IM in an eleven-month-old infant.

2. Case Presentation

2.1. Patient information

An 11-month female baby, born via cesarean section (CS) from a G3P3 mother, was brought to the clinic by her parents, weighing 3.5 kg. She was referred by a pediatrician for a checkup and presented with no symptoms. During routine fetal sonographic screening, she was found to have intrauterine hydronephrosis and enlargement above the left kidney. Apart from these findings, the baby appeared well and had no other abnormalities.

2.2. Clinical findings

On general examination, no abnormality was found apart from a small reddish discoloration of the overlying skin. The patient's pupillary reflex was normal in both eyes, with no limitations in eye movement. A detailed systemic clinical examination was unremarkable. On palpation, there was a well-defined swelling with a localized border, firm, non-mobile, mild discomfort, fixed to the underlying bone, and non-adherent to the superficial skin. There was no evidence of cervical lymphadenopathy.

2.2. Diagnostic assessment

The ultrasound showed a well-defined, lobulated surface, solid hypoechoic lesion of 23 *12*10 mm subcutaneously and inferiolateral to the right orbital cavity, with normal surrounding tissue and normal underlying bone. There were multiple bilateral cervical lymphadenopathy of variable size and shape, all of them are well defined, with cortical thickening of less 3 mm and suggested to be inflammatory lymph nodes. An incisional biopsy was obtained from the lesion in the infraorbital region on the right side, confirming the diagnosis of Myofibroma.

2.3. Therapeutic intervention

Under general anesthesia, through transverse incision, the right infraorbital mass was excised. Non-absorbable sutures were used to close the wound. The histopathological examination revealed a 3 cm myofibroblastic mass that involved the margins at multiple spots without 12a1' necrosis and aberrant mitosis (figure 1).

2.4. Follow-up

Postoperative recovery was uneventful, the one-year follow-up period showed no signs of recurrence.

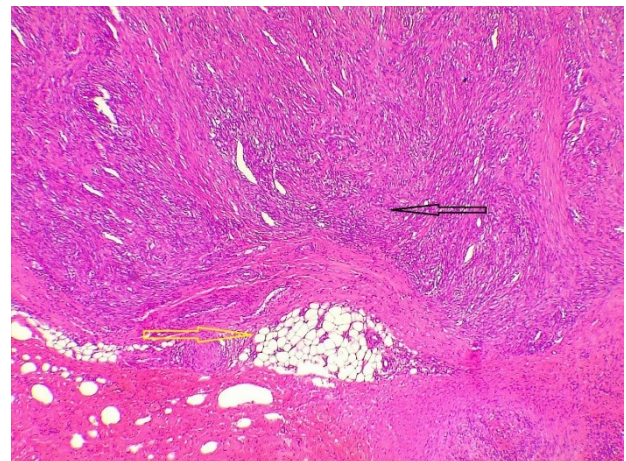


Figure 2. bland looking cellular spindle cells (dark arrow) infiltrating into adjacent fibrofatty tissue (yellow arrow), Hematoxylin and Eosin stain. Microscopic power 4x10.

4. Discussion

Infantile myofibroma is a proliferative disorder of mesenchymal origin that is characterized by nodular neoplasm formation in soft tissues, including cutaneous, subcutaneous, muscle, bone, and visceral structures. It typically occurs in infancy, predominantly in the head and neck. However, a solitary cutaneous IM has been reported in a 49-year-old lady [5].

Myofibroma was initially described in 1951 as congenital fibrosarcoma and subsequently as congenital generalized fibromatosis. Congenital fibromatosis was classified into two types: multiple forms involving lesions of the skin, subcutaneous tissue, skeletal muscle, and bone with a good prognosis; and a generalized form involving visceral lesions with a poor prognosis. Infantile hemangiopericytoma is now recognized as part of the spectrum of IM [3].

The etiopathogenesis of myofibroma is obscure. Some have reported its inheritance as an autosomal dominant pattern, while others have suggested an autosomal recessive pattern. Intrauterine estrogen hormone has a role in its genesis. Experiments on the oncogenic ability of estrogen in lab animals have resulted in the proliferation of lesions with similar histological features as IM [3].

The solitary nodules are most commonly seen in the head and neck region, which includes the scalp, forehead, orbit, parotid region, and oral cavity. Although considered the most common tumor in infancy, the reported incidence of solitary osseous myofibroma is rare. Apart from the soft tissues and the skeleton, rare involvement of organs like the lung, heart, gastrointestinal tract, and pancreas has been reported [6]. Myofibroma appears as a single swelling or mass, most commonly in the dermis and subcutis, which may be freely movable at times. The overlying skin is usually normal, sometimes it may resemble a purplish macule, and infrequently it may ulcerate. On physical examination of the current case, there was a slight reddish discoloration of the overlying skin; the swelling was seen as a

large mass with a localized border, firm, non-mobile, tender, fixed to the underlying bone, and non-adherent to the superficial skin. There were no palpable cervical lymph nodes.

Radiographically, myofibromas appear as a well-defined unilocular radiolucency in most cases, with a few exhibiting multilocularity. MRI is the most favorable and useful when dealing with soft tissue tumors, especially in children. The ultrasonography of the current case showed a well-defined lobulated surface, a solid hypoechoic vascular nodule with normal surrounding tissue, and normal underline bone without any features of invasion to the surrounding tissues [7].

On histopathological examination, the lesion presented with a biphasic cellular pattern, which is usually seen in myofibromas. The tumor cells showed diffuse immunopositivity for smooth muscle actin (SMA) (contractile protein actin) and vimentine (a mesenchymal cell intermediate filament) and were negative for desmin (smooth muscle antigen) and S-100 (nervous tissue antigen), which have been consistently used to spot a myofibroblastic lineage [8]. Histopathologically, the definitive diagnosis of myofibroma was challenging in light of the various differential diagnoses like nodular fasciitis, fibrosarcoma, leiomyoma, and neurofibroma. Nodular fasciitis was excluded as it is rarely seen in infants and has a prominent myxoid matrix in the absence of a hemangiopericytoma-like pattern of myofibroma. Leiomyoma and leiomyosarcoma were excluded based on the negative response of tumor cells to desmin. Another closely resembling tumor that is considered is fibrous histiocytoma, which is typically composed of polymorphous cells arranged in a storiform pattern and exhibits only focal staining with SMA. Neurofibroma and neurofibromatosis were excluded based on the immunonegativity of myofibroblasts to S-100 [9]. Fibromatosis usually has a monophasic growth pattern consisting of long fascicles of spindle cells among abundant wavy collagen fibers, which was not seen in the present case.

Surgical treatment is the most effective treatment for patients without invasion. Some cases show spontaneous regression and thus require no treatment. The prognosis is usually excellent, with the rate of recurrence being less than 10%. The patient should be monitored for at least five years to assess recurrences and exclude the manifestation of further nodules characterizing myofibromatosis [9]. When it invades the underlying tissues, it needs wide surgical resection.

5. Conclusion

Although IM is a very rare tumor, it can appear in the first two years of life, and the majority of them are in the head and neck region. It may regress spontaneously when they don't invade the surrounding tissues, but they need surgical resection when they invade other tissues.

Declarations

Conflicts of interest: The author(s) have no conflicts of interest to disclose.

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