ACTA SCIENTIFIC MEDICAL SCIENCES (ISSN: 2582-0931)

Volume 4 Issue 2 February 2020

Case Report

My fibroma Infantile Solitary: About A Case

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Received: January 02, 2020

Published: February 08, 2020

Abstract

Infantile myofibromatosis (ILM) described by Enzinger in 1981 is the most common childhood fibromatosis. It consists of a benign proliferation of fibroblasts and myofibroblasts. There are three forms. Solitary MFI, the most common type, characterized by isolated skin, bone or soft tissue injury; multiple MFI represented by several locations and generalized MFI with visceral involvement. The morphological characters are identical whatever the location. The histological diagnosis is based on the identification of a double component, myofibroblast fasciculate at the periphery and hemangiopericytomas at the centre, which expresses smooth muscle actin. There is usually no atypia or mitosis, but there can be observed images of intravascular extensions in the centre. In the solitary form, the prognosis of MFI is good but it is often fatal in cases of visceral involvement. The reported case concerns a 4-year-old child who has had a left inguinal nodule since the age of 1, whose histological study after surgical excision has led to the diagnosis of myofibroma.

Keywords: Myofibroma; Myofibromatosis; Localized; Multiple; Generalised

Introduction

The myofibromatose child is the most common of fibromatosis of the child. It consists of a benign proliferation of fibroblasts and myofibroblasts. There are three forms: the solitary form, the multiple form and the generalized form. The prognosis for MFI is generally good outside the generalized form with visceral involvement.

Observation

Child of 4 years presents since the age of 1 year a left inguinal nodule gradually increasing asymptomatic volume. The clinical examination found a 1 cm nodule with a purplish surface sitting on the left inguinal level (Figure 1) whose histological study after excision was in favour of a myofibroma.

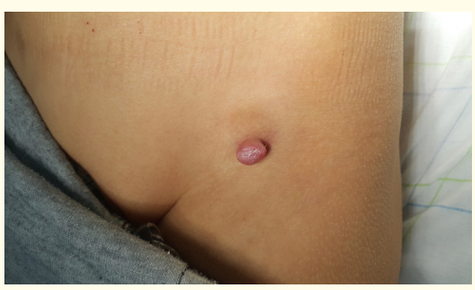

Figure 1: Purplish inguinal nodule on the left.

Discussion

There myofibromes are soft tissue tumours that vary depending on the age at which they are discovered. The myofibromatosis was first reported times in children by Stout in 1954 and called " generalized congenital fibromatosis ". Various synonyms exist in the literature (hamartoma widespread hamartomas mesenchymal multiple congenital leiomyomas multiple vascular of the new born, congenital fibromatosis multiple) [1]. The term " infantile myofibromatosis " has been proposed in 1981 by Chung and Enzinger [2]. There are 2 types, a solitary form and a disseminated form with multiple lesions. This second form, less frequent, can be accompanied by visceral involvement, the outcome of which is sometimes fatal [3]. In children, Lesions are most often present at birth or appear during the first 2 years of life. The solitary form is a nodule, renitent, well circumscribed and sometimes polylobed, of variable size. It can be red or purplish, sometimes covered with telangiectasia [4]. The seat is ubiquitous but the myofibroma predominates on the head and the neck including the oral cavity [3,4]. The sex ratio of 2/1 shows a male predominance [1,3]. For these solitary forms of the child, there is a natural tendency to spontaneous regression [4]. In disseminated myofibromatosis, myofibromas are identical to those of solitary forms and their number varies from a few to several hundred. This rarer form is characterized by more frequent muscle, bone and visceral involvement. The skull and bones long would be more affected than the spine. The forms limited to the skin, the soft tissue and bone have a good prognosis with spontaneous regression in most cases [2]. The invasion visceral would exist in 35 p. 100 of the forms disseminated. Death occurs at birth or soon after, from cardiac, pulmonary or gastrointestinal complications. Mortality is in the order of 15% 100 [1]. This potential gravity of the disseminated form justifies carrying out additional examinations.

A skeleton x-ray, a full body scanner, a ECG and an echocardiogram are systematically requested. MRI would be more efficient, able to detect an involution of the disease [3]. Autosomal dominant transmission with variable penetrance has been proposed due to family cases [1,3,4]. The morphological and immunohistochemical examination shows a well limited nodule, deep dermal and hypodermic, constituted by a cellular proliferation of biphasic aspect with a phenomenon of zone. At the periphery, there is a proliferation of spindle cells with abundant eosinophilic cytoplasm arranged in bundles, of myoid differentiation. Sometimes these cells form buds in the vascular lumens from which they remain separated by the endothelial lining. In the centre, the spindle cells of smaller size are arranged in bundles of concentric arrangement around branched vessels achieving a hemangiopericytarian aspect. The two cellular components also express alpha-AML and vimentin and are negative for desmin and the protein S100. This central area can be necrotic, haemorrhagic or calcified [5,6]. The therapeutic choice depends on the solitary or multifocal character of the myofibromas. The small solitary my fibroma is mostly excised for diagnostic purposes. This excision is often curator. A recurrence occurs in 10%. About 100 of the cases [2] and re- cutting is recommended [3,7]. For the other forms of myofibromatosis described in children, the possibility of spontaneous regression must always be taken into account, especially in the neonatal period. The diagnosis will be confirmed by a biopsy.

When the lesion is unique but locally aggressive or extensive, destruction gradual neighbouring tissue by the fibrosis is likely to cause functional impairment. Some authors then recommend partial resection in order to preserve vital functions. Complete excision may be delayed, assuming spontaneous involution of the lesion [8,9].

Certain asymptomatic myofibromas for which a surgical gesture would cause disastrous esthetic or functional sequelae, do not justify any therapeutic gesture. A monitoring and clinical examinations image of regular series are available [10]. In disseminated childhood myofibromatosis, therapeutic abstention is required. Spontaneous regression is observed before the age of 2 years in the vast majority of congenital myofibromatosis limited to the skin, soft tissue and bone. The poor prognosis for visceral forms is linked to the local mass effect of the nodules, responsible for obstruction and tissue destruction [9]. Radiotherapy and chemotherapy have been used for these unresectable forms. Products used are vincristine, actinomycin D and cyclophosphamide, but the efficacy is uncertain [9,11]. However, in the absence of repercussions on vital functions and despite an impairment diffuse visceral, spontaneous regression is possible after a few months [1].

Conclusion

MFI, although the most frequent of fibromatosis in infants and children, especially in its solitary and cutaneous form, is a rare form of benign mesenchymal lesion in children. Its histological diagnosis, in typical forms as in our observation or in less differentiated forms, is based on the evidence, in immunohistochemistry, a differentiation myofibroblastic. The treatment of choice consists, in localized forms, in a surgical excision, this condition being of excellent prognosis with, in certain cases, a tendency to spontaneous regression.

Bibliography

1. C Giannakopoulou. "Infantile myofibromatosis: a case study and review of literature". *The Journal of Dermatology* 26.9 (1999): 595-598.
2. EB Chung and FM Enzinger. "Infanti the myofibromatosis". *Cancer* 48.8 (1981): 1807-1818.
3. SW Weiss and JR Goldblum. "Enzinger and Weiss's Soft tissue tumors". 4th ed. St. Louis: Mosby (2001): 357-360.
4. Stanford D and Rogers M. "Dermatological presentations of infantile myofibromatosis: a review of 27 cases". *Australasian Journal of Dermatology* 41.3 (2000): 156-161.
5. T Hasegawa., *et al*. "Solitary infantile myofibromatosis of bone". *The American Journal of Surgical Pathology* 17.3 (1993): 308-313.
6. S Ijima., *et al*. "Solitary form of infantile myofibromatosis: a histologic, immunohistochemical, and electronmicroscopic study of a regressing tumor over a 20-month period". *The American Journal of Dermatopathology* 21.4 (1999): 375-380.
7. SF Hogan and JR Salassa. "Recurrent adult myofibromatosis. A case report". *The American Journal of Dermatopathology* 97.6 (1992): 810-814.
8. DT Netsche. "Infantile myofibromatosis: case report of a solitary hand lesion with emphasis on differential diagnosis and management". *Annals of Plastic Surgery* 46.1 (2001): 62-67.
9. NS Goldberg., *et al*. "Infantile myofibromatosis: a review of clinicopathology with perspectives on new treatment choices". *Pediatric Dermatology* 5.1 (1988): 37-46.
10. N Loundon. "Head and neck infantile myofibromatosis: a report of three cases". *International Journal of Pediatric Otorhinolaryngology* 51.3 (1999): 181-186.
11. B Raney., *et al*. "Nonsurgical management of children with recurrent or unresectable fibromatosis". *Pediatrics* 79.3 (1987): 394-398.