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Congenital Desmoid Fibroma of the Soft Tissues in a 6-Month-Old Girl

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

A desmoid tumor (DT), also known as aggressive fibromatosis, is a benign neoplasia of the soft tissues. It arises from conjonctive tissues, fascial sheaths, and muscular aponeurotic structures of muscles. According to biological characteristics, they are classified into three groups: -sporadic, -associated with familial adenomatous polyposis, -and familial or multicentric form. The pediatric form is ranged as a fourth group. To our knowledge, we report the first case in the world of congenital desmoid fibroma in the soft tissues of the ankle in a 6-month-old girl who is treated by surgical resection, with a good evolution over a 6-year follow-up. Through this observation, the authors suggest a review of the literature.

Keywords: Desmoid fibroma; newborn; female.

1. INTRODUCTION

Desmoid fibroma is a rare benign fibrous tumor. It is characterized by local malignity with recurrence tendency, arising from conjunctive tissues, the fascial sheaths, and musculoaponeurotic structures of muscles.

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In the limbs, the involvement is mainly at the root; feet and hands are rarely affected, with variable clinical appearances.

The clinical and radiological signs are not specific, and a histologic study confirmed the diagnosis, but this one can be difficult. More than 75% of cases occur during the first three decades [1]. The neonatal or congenital form has never been described in the literature.

We report the first worldwide case of congenital desmoid fibroma in the soft tissues of the ankle in a 6-month-old girl. Through this observation, we suggest reviewing the clinical, therapeutic, and evolutionary aspects of desmoid fibroma.

2. CASE PRESENTRATION

A 6-month-old girl, has been born from a well-monitored pregnancy. APGAR score was 10/10. Her father died from an unlabeled digestive neoplasia pathology.

There was a swelling of the right ankle, which has a hard consistency and regular contours, measuring 5 cm in diameter, with some bleeding ulcerations during dressing change (Fig. 1) and a gradual increase of its size.

She was received first at the age of one month.

The x-ray of the foot was normal (Fig. 2), and MRI found signs of hemangioma (Fig. 3), then treatment with beta-blockers was conducted for three months without improvement.

The resection of the tumor was made given to the non-improvement and the anemic syndrome due to the bleeding.

An incision was made around the tumor, taking 1 cm of healthy skin. The exploration revealed a whitish tumor measuring 6 cm, with a clear cleavage plane. But the tumor infiltrated the sub-aponeurotic plane. Extensor tendons were intact as well as the pedal pedicle. The resection also involved the fascia. The closure was made in the transverse plane without the need for a skin graft or a flap.

The histologic study revealed a desmoid fibroma. Genetic complement was performed showing a normal molecular analysis. She was followed up every six months, and actually each year. Good evolution over 6 years (Fig. 4: 3-year follow-up).

3. DISCUSSION

Desmoid tumors are an uncommon part between fibromatosis and fibrosarcomas. It appears to be a rare tumor representing only 0.3% of benign bone tumors, according to Taconis et al. [1] and Vaz et al. [2].

The incidence of desmoid tumors in children is 2–4/ million a year with local malignancy (infiltrating tumor) and no metastasis or malignant transformation [3-4]. They are known as progressive fibromatosis.

Desmoplastic fibroma occurs twice in women. Hormonal factors, especially estrogen, promote the occurrence of TD. The arguments in favor of this hypothesis are the more frequent occurrence of TD in women during periods of high hormonal impregnation such as pregnancy and until the year following childbirth, their regressions possibly spontaneous at menopause, and, finally, the effectiveness of some anti-hormonal therapies, especially anti-estrogens [5-6].

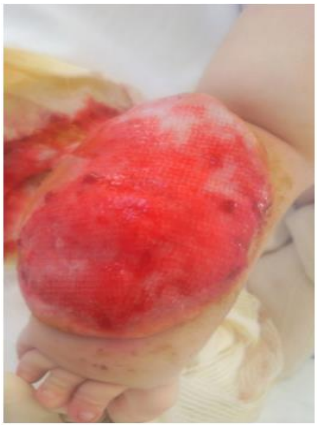

Fig. 1. Clinical aspect of the mass with an ulcerated surface

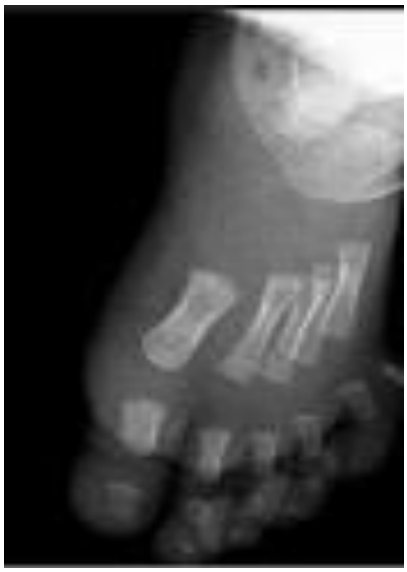

Fig. 2. X ray

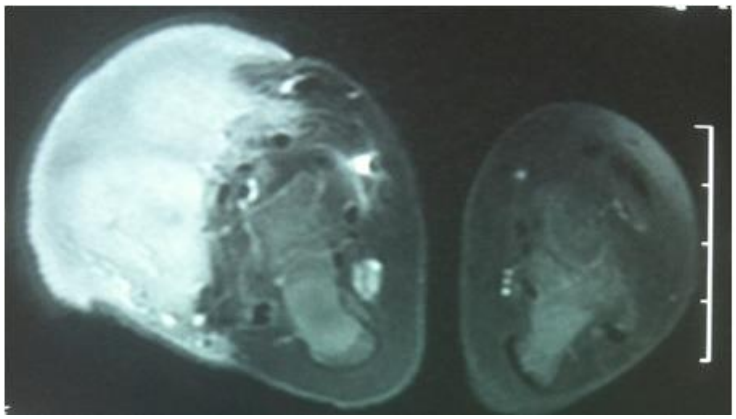

Fig. 3. MRI aspect of the mass which sends partitions in under aponeurotic

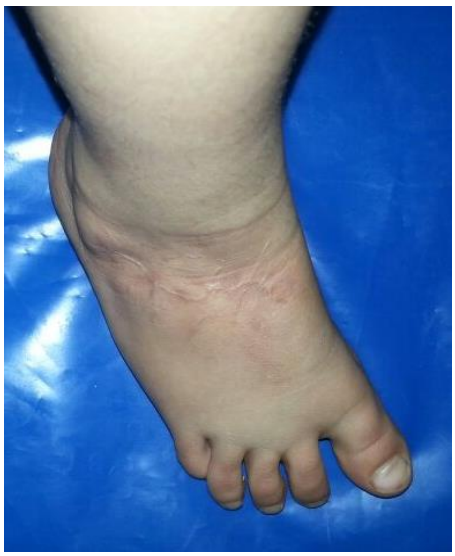

Fig. 4. Current appearance of the foot and the scar (3-year follow-up)

This theory can be applied to our patient, being first of all female, and its occurrence intrauterine.

More than 75% of cases appear in the first three decades. The incidence is high between 20 and 40 years old. Contrary to our patient which presented a neonatal mass, and therefore congenital [2-4,7], that led to confusion with hemangioma and then a delay in the diagnosis.

The discovery mode is not specific, It's a very slow-growing tumor. The most frequent symptoms are pain and swelling [7].

The role of trauma caused by previous surgery has also been suggested [4]. Nevertheless, that theory seems to be questioned given the role of genetic factors [8]. Genetic factors have been well incriminated in association with PAF which is an autosomal dominant disease linked to the mutation of the adenomatous polyposis colic tumor suppressor gene (APC). The hypothesis retained in our patient was hormonal impregnation probably associated with genetic factors considering that the father died of non-specified digestive neoplasia.

The macroscopic aspect of desmoplastic fibromas has been described by Mazabraud et al. [9]. It's a whitewash or grayish homogeneous tissue with firm or elastic consistency, rubber-like. The sectional slice of the tumor appears glossy with fasciculations. In our case, it was a whitewash, elastic but ulcerated mass bleeding on touch, which led to confusion with hemangioma. The diagnosis was later rectified. Sometimes there is an impression of relatively clear cleavage between the tumor areas and the adjacent bone tissues.

In the Microscope, there is a large histological similarity between the bone desmoid fibroma and the soft tissue desmoid fibroma (abdominal or extra-abdominal desmoid tumors). It's a connective tissue very rich in collagen. These collagen fibers are arranged in large parallel bundles or, on the contrary, in fine wavy fibers without a clear fasciculate arrangement. There is a moderate cellular richness, even weak. The cells are spindle-shaped and small. The nuclei are small, regular, round, or oval, with fine chromatin, mitoses are rare. There is no atypical mitosis. The desmoplastic fibroma has moderate vascularization without bony or cartilaginous components.

Their benign histological structure and poor metastatic potential contrast with a high local aggressiveness, and a high rate of recurrence after surgical excision. This justify an extra-lesional removal with an intact margin of peri-tumoral bone tissue to limit the risk of local recurrence.

Analysis of the treatment of desmoplastic fibromas in the literature is difficult. Indeed, the therapeutic approach is based on the experience of each author.However, a high local recurrence rate isobserved [9-12]. The main risk factor for recurrence is incomplete surgical resection with positive surgical margins [12,13-14]. The other risk factors are young age, very large tumour and specific tumour sites. Head and neck tumours are associated with the poorest prognosis [9,10,12]. In the present series, positive surgical margins appeared to be a risk factor for recurrence, as also reported in other studies.

After surgery, it needs a long term of follow up, even if the average delay of recurrence reported is three years [7]. Recurrences have been described up to ten years after the initial treatment. Surveillance should be clinical and radiological.

The natural evolution of desmoid fibroma after treatment is marked by high risk of tumor recurrence.

The prognosis is good in terms of survival. It determines the modalities of treatment which must remain conservative in the majority of cases. Wide surgical excision, without mutilation, ensures the best chances of recovery without sequelae. When It's not possible, radiotherapy is an alternative and leads to high rate of complete regression [13-15].

4. CONCLUSION

Neonatal forms of desmoid fibroma of the extremities represent a new clinical entity of desmoid tumors. Their locally invasive character is not yet demonstrated. The definitive surgical treatment is difficult.

CONSENT

As per international standard or university standard, parental(s) written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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