



## Foot fibromatosis in children. A case report and literature review

### Fibromatose du pied. A propos d'un cas pédiatrique et revue de la littérature

<sup>1</sup>Hizem-Harzallah W., <sup>2</sup>Zrig M., <sup>1</sup>Salem R., <sup>2</sup>Mnif H., <sup>2</sup>Abid A., <sup>1</sup>Golli M.

<sup>1</sup>Department of Radiology – Fattouma Bourguiba's Hospital. Monastir – Tunisia.

<sup>2</sup>Department of Orthopaedic Surgery – Fattouma Bourguiba's Hospital. Monastir – Tunisia.

CORRESPONDENCE: **Dr. Wissem HIZEM-HARZALLAH**

Department of Radiology – Fattouma Bourguiba's Hospital. Avenue Farhat HACHED, 5000 Monastir – Tunisia.

E-mail : [wissemharzallah@hotmail.com](mailto:wissemharzallah@hotmail.com)

#### ABSTRACT

Plantar fibromatosis is a relatively uncommon benign, focally invasive fibrous neoplasm. It is most often found to invest the heel and medial portions of the plantar fascia. Imaging particularly MRI is often used in the evaluation of these tumors.

We report a rare case of fibromatosis involving the lateral portions of the plantar fascia, which was diagnosed in 18 months aged girl. She underwent tumor resection with a partial plantar fascia resection and without recurrence at 24 months follow-up.

**Keywords:** fibrous tumours, children, foot

#### RÉSUMÉ

La fibromatose plantaire est une tumeur bénigne relativement rare avec une agressivité locale, Elle se développe le plus souvent au niveau du talon et la partie médiale de l'aponévrose plantaire. L'IRM est souvent utilisée dans l'évaluation de ces tumeurs.

Nous rapportons un cas rare de fibromatose siégeant au niveau de la partie latérale de l'aponévrose plantaire et diagnostiqué chez une fille âgée de 18 mois. Une aponévrectomie plantaire partielle emportant la tumeur a été réalisée sans récurrence à 24 mois de suivi.

**Mots clés :** tumeur fibreuse, enfant, pied

#### I. INTRODUCTION

Tumours of fibroblastic-myofibroblastic origin, account for 12% of soft tissue neoplasms in children, being fourth in frequency [1]. Fibrous tumours of childhood include several disorders with variable biologic behaviour [2]. In children, fibromatosis may involve the abdominal wall or have extra-abdominal involvement [3].

Infantile (desmoid-type) fibromatosis represents the childhood counterpart of extra-abdominal desmoids in adults [3, 4]. Lesions most frequently occur in the head and neck; the limb girdles, trunk, and proximal extremities are also common sites [5]. The foot is rarely involved [2].

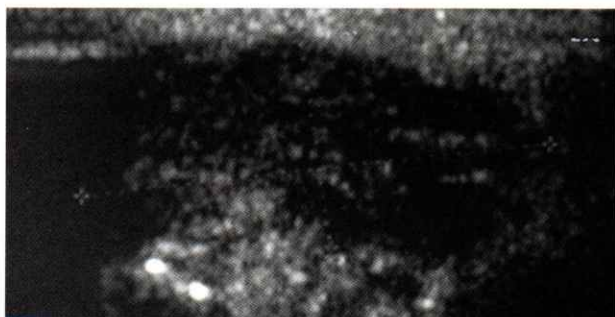
We report a rare case of foot fibromatosis in an infant. Value of MRI will be emphasized.

#### II. CASE REPORT

The 18 months aged girl presented with a swelling in the plantar aspect of the left forefoot. Physical examination revealed a non-tender soft tissue mass over the lateral aspect of the forefoot, which was quite firm. The overlying skin was normal. No flexion contracture of toes or neurovascular deficits was noted. The infant was otherwise quite well especially the examination of skin and subcutaneous tissues elsewhere in her body and the palms did not reveal any similar swelling.

Preoperative radiographs were normal.

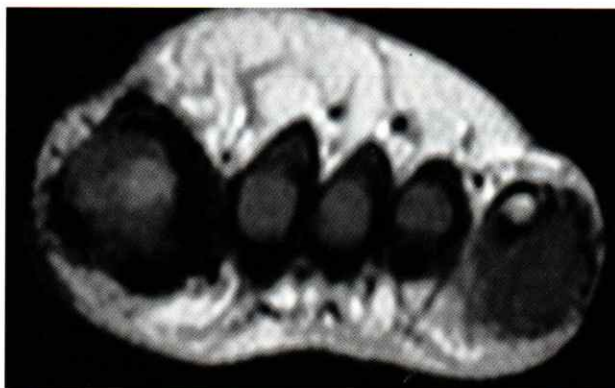
On ultrasonography (US), the solid mass appeared hypoechoic, mildly heterogeneous, and well-defined (Figure 1).



**Figure 1:** US image shows hypoechoic, mildly heterogeneous, and well-defined lesion

**Figure 1 :** L'échographie montre une masse hypoéchogène, un peu hétérogène et bien limitée

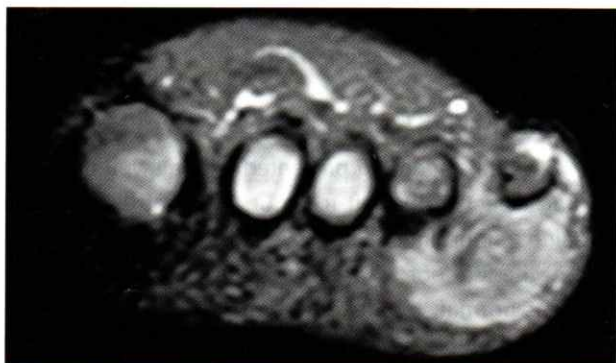
Magnetic Resonance Imaging (MRI) was performed. T1-weighted (TR 500, TE 22) pulse sequence revealed a well-circumscribed soft tissue mass measured 23x11mm lying on the plantar and lateral aspect of the left forefoot facing the heads of the 4th and 5th metatarsal bones and surrounding the head of the 5th metatarsal bone without osseous invasion (Figure 2).



**Figure 2:** Axial T1-Weighted image shows that tumor is hypointense and surrounding the 5th metatarsal bone without osseous invasion

**Figure 2 :** Tumeur hypoT1, entourant le 5ème métatarsien sans l'envahir

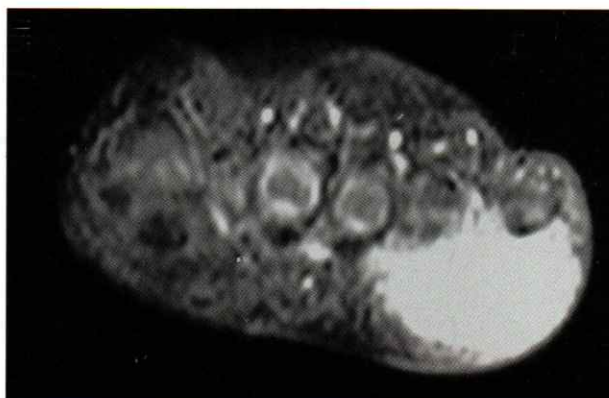
The tumor had low signal intensity on T1-weighted sequences and demonstrated heterogeneous high signal intensity on Fat Sat T2-Weighted sequences (TR 2550, TE 120) (Figure 3).



**Figure 3:** Tumor has heterogeneous high signal intensity on T2-weighted image (TR 2550, TE120) with Fat Saturation

**Figure 3 :** Tumeur hyperintense et hétérogène sur la séquence T2 Fat Sat

After injection of Gadolinium, the tumor showed a marked homogenous enhancement (Figure 4).

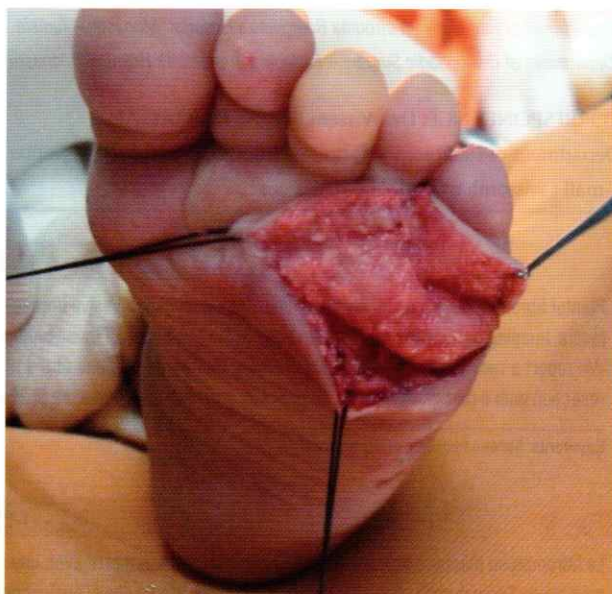


**Figure 4:** Gadolinium-enhanced axial T1-Weighted Spin-Echo MR image with fat saturation shows homogeneous marked tumor enhancement

**Figure 4 :** Prise de contraste homogène et intense de la tumeur sur les séquences T1 Fat Sat

A plantar fibroma was suspected and the infant was scheduled for surgical excision of this mass.

Horizontally plantar incision was performed from the head of the 3th to the 5th metatarsal bone (Figure 5).



**Figure 5:** Surgical findings demonstrate a subcutaneous fibrous soft tissue mass

**Figure 5 :** Tumeur fibreuse plantaire siégeant dans les parties molles sous cutanées

The tumor was confined to the plantar fascia without invasion of adjacent soft tissues particularly flexor tendons. Surgical resection with a wide margin was performed. A partial plantar fascia resection was done.

Histological exam revealed a dense fibrous tissue with cells that resembled normal fibroblasts in appearance and organized in fascicles. No atypical or mitotic activity was seen. It had concluded to fibromatosis.

The infant has had no recurrence twenty-six months following surgery.

### III. DISCUSSION

A tumour predominantly composed of myofibroblasts or fibroblasts is called a fibroma or fibromatosis if it appears benign on histological examination, or a fibrosarcoma if the pathological criteria for malignancy are fulfilled [1]. The term fibromatosis is used for fibroblastic-myofibro-

blastic proliferation with the following characteristics: a tendency to invade surrounding tissues, a tendency to recur after incomplete excision, and absence of metastasis.

In addition, some fibromatosis may show spontaneous regression and presentation with multifocal tumours.

In general, there is a high incidence of fibromatosis in neonates, infants and young children [1].

The most widely accepted classification scheme for fibrous tumours was proposed by ENZINGER and WEISS [4], who divided paediatric fibrous tumours into categories:

Tumours "corresponding in clinical setting, microscopic picture, and behaviour to similar lesions occurring in adults, including desmoid fibromatosis".

Tumours "that are peculiar to infancy and generally have no clinical and morphological counterpart in adult life". Desmoids fibromatosis includes a broad range of clinical disorders ranging from benign nodules to infiltrating masses with a tendency to recur. ENZINGER and WEISS [4] separate the entities into superficial (palmar, plantar, and penile fibromatosis) and deep [extra-abdominal (chest-wall, back, extremities), abdominal (abdominal wall) and intra-abdominal (pelvic, mesenteric) fibromatosis]. Extra-abdominal fibromatosis may involve the head and neck, shoulders or extremities. The feet are rarely involved [2].

The plantar fibromatosis is uncommon and literature on this topic is limited and poor and few series were reported [6-9]. PICKREN et al. [7] reviewed the literature and summarized the findings relative to 104 patients. There were 51 male and 53 female patients, whose ages at the onset of the disease ranged from twelve to sixty-three years old. Twenty-one patients had bilateral involvement. Fifty-five patients had isolated plantar disease and forty-nine had associated disease of the palm. Twenty-five patients were epileptic. Only one patient had penile contracture. PICKREN et al. [7] reported 16 patients and the youngest was 5 years old. The important pediatric series was reported by GODETTE et al. [10]. These authors reported fourteen children who had an asymptomatic lump on the anteromedial portion of the fat pad of the heel. Eleven were boys and the age at the onset of the disease ranged from nine months to sixteen years old. There was no association with lesions of the hands or penis, and the lesions did not behave in an aggressive fashion as may occur in patients in whom the lesions develop more anteriorly in the foot. Six were subjected to excision and histological studies showed irregular nodules of moderately cellular fibrous tissue attached to the plantar fascia.

We think that our case is different from this entity reported by GODETTE [10] and the very known entity of superficial plantar fibromatosis or Dupuytren's contraction; our case corresponds to an extra-abdominal desmoid fibromatosis. According to our literature research, we found no similar case to ours concerning the age, location and type of fibromatosis.

Proliferation in the plantar aponeurosis may be so aggressive that the overlying skin, the fascia and deep structures of the sole are affected. In contrast to palmar fibromatosis, which affects preferentially the 3rd to 5th rays of the hand

(lateral hand side), plantar fibromatosis mostly occurs at the 1st and 2nd rays of the foot (medial foot side).

Although the etiology of plantar fibromatosis remains a mystery, several etiological factors have been proposed, including trauma [11], neuropathy [12], biochemical and metabolic imbalance [12], faulty development [11], infection [6], and the patient's occupation [13, 14]. Hormonal etiology was suggested for desmoid fibromatosis. Radiographs may demonstrate osseous involvement with erosion, deformation and bowing of bone adjacent of the tumour [1, 4].

Ultra-sonographic appearances are not specific. The solid lesions frequently appears homogenous and hypoechoic, although some tumour may be heterogeneous with variable echogenicity [1, 5]. In our case, the tumor was hypoechoic and mildly heterogeneous.

CT scan usually shows a soft tissue density lesion, but high-density tumors have been reported in adults.

MRI shows intermediate or low signal intensity and high or low intensity on T2-Weighted images. Hypointensity on T2-Weighted images has been shown to be due to a greater collagenous component and reduced cellularity compared with lesions showing high signal intensity [15, 16].

In our case, the lesion exhibited low signal intensity in T1-Weighted images and heterogeneous high signal intensity on T2-Weighted images with fat saturation.

A nodular pattern is more frequently present in adults, while paediatric patients more often show an infiltrative pattern, which is associated with a higher risk of recurrence [1].

Although CT and MR findings are not specific, they can help define tumor margins and extent [2].

Pathological examination reveals a poorly circumscribed, firm infiltrative lesion consisting of elongated, slender, spindle-shaped cells. Cells and dense, often hyalinised, collagen fibers are arranged in bundles. Mitoses are variable [2].

These proliferative fibrous lesions are ill circumscribed and difficult to resect completely which probably accounts for their high recurrence rate.

The only reason to treat plantar fibromatosis is to relieve the associated symptoms that often result from local extension and invasion. An indolent lesion can invade the neurovascular structures, necessitating operative intervention. Non-operative treatment begins with the construction of a well-moulded, padded shoe and an orthosis. Transference of weight away from the prominent nodules is important; often, custom-made insoles are needed [17]. GODETTE et al. [10] suggest observation for asymptomatic tumors, which can disappear spontaneously.

The major indication for operative intervention is pain. When the nodules become large and painful enough to be disabling while the patient is standing or walking, several options are available. For GODETTE et al. [10], if a lesion expands rapidly, it should be subjected to excision biopsy. Resection may also be considered if a lesion has infiltrated the neurovascular structures evidenced by clinical examination or MRI.

ALLEN et al. [8] reported 69 patients and the lesions were excised in 28 of the sixty-nine patients, and they recurred

in 15. One patient had an amputation.

AVILES et al. [9] described 22 patients and 14 had a simple excision of the lesions and eight had a wide radical excision. Eight of the patients who had had a simple excision had a recurrence, compared with only one who had had a wide radical excision. No patient had an amputation.

Because recurrence after an incomplete excision is very common [7, 10], authors began to advocate a wider excision [7, 9, 18-21].

These benign tumors are locally aggressive, and wide local surgical excision is the treatment of choice owing to the high rate of recurrence in the lower extremities. Invasiveness into the surrounding soft-tissue structures often makes wide excision difficult without compromise of function particularly in the foot.

The importance of skin incision has been addressed by several authors [7, 22-24]. Care must be taken to place the incision away from the weight-bearing region. CURTIN [23] described a technique that utilizes a specific type of plantar incision.

Although effective in decreasing the recurrence rate, adjuvant radiotherapy should be used very selectively because of its serious side effects [25, 26].

The clinical presentation and MR characteristics may suggest the diagnosis of fibromatosis in a young patient. However other fibrous lesions, including malignant fibrous histiocytoma may have similar MR appearances [15, 16]. Thus histology is still needed for definitive diagnosis [15].

Fibromatosis refers to group of proliferative lesions of myofibroblasts. They are locally aggressive, and tend to recur locally after attempted surgical removal. However, they are not considered to metastasize [15, 27].

#### IV. REFERENCES

- Eich G.F., Hoeffel J.C., Tschappeler H., Gassner I., Willi U.V. Fibrous tumours in children: imaging features of a heterogenous group of disorders. *Pediatr Radiol* 1998; 28: 500-9.
- Patrick L.E., O'Shea P., Simoneaux S.F., Gay B.B., Atkinson G.O. Fibromatoses of childhood: the spectrum of radiographic findings. *Am J Radiol* 1996; 166: 163-9.
- Ahn J.M.O., Yoon H.K., Suh Y.L., Kim E.Y., Han B.K., Yoon J.H., Kim S.H., Cho J.M., Kim S.M., Kangs H.S. Infantile Fibromatosis in childhood: Findings on MR imaging and pathologic correlation. *Clin Radiol* 2000; 55: 19-24.
- Enzinger F.M., Weiss S.W. Fibrous tumors of infancy and childhood. In Enzinger FM, Weiss SW eds. *Soft tissue tumors*, 3rd ed. St Louis, Mosby; 1995: 231-68.
- Kingston C.A., Owens C.M., Jeanes A., Malone M. Imaging of desmoids fibromatosis in pediatric patients. *Am J Radiol* 2002; 178:191-9.
- Meyerding H.W., Shellito J.G. Dupuytren's contracture of the foot. *Internat Coil Surg* 1948; 11:595-603.
- Pickren J.W., Smith A.G., Stevenson T.W.J., Stout A.P. Fibromatosis of the plantar fascia. *Cancer* 1951; 4:846-56.
- Allen R.A., Woolner L.B., Ghormley R.K. Soft-tissue tumors of the sole. With special reference to plantar fibromatosis. *J Bone Joint Surg* 1955; 37A:14-26.
- Aviles E., Arlen M., Miller T. Plantar fibromatosis. *Surgery* 1971; 69:117-20.
- Godette G.A., O'Sullivan M., Menelaus M. Plantar fibromatosis of the heel in children: a report of 14 cases. *J Pediatr Orthop* 1997; 17:16-7.
- Skoog T. Dupuytren's contraction. With special reference to aetiology and improved surgical treatment. Its occurrence in epileptics. Note on knuckle-pads. *Acta Chir Scand* 1948; Suppl 139:47.
- Anderson W. Lectures on contractions of the fingers and toes; their varieties, pathology, and treatment. *Lancet* 1891; 2:1-5.
- Dupuytren C. Rétraction permanente des doigts: leçons orales de clinique chirurgicale, faites a l'Hôtel-Dieu de Paris. Vol. 1, pp. 2-24. Paris, Germer-Baillière, 1832.
- Dupuytren G. Leçons orales de clinique chirurgicale, faites a l'Hôtel-Dieu de Paris. Vol. 5, pp. 473-482. Paris, Germer-Baillière, 1832.
- Liu P., Thorner P. MRI of fibromatosis: with pathologic correlation. *Pediatr Radiol* 1992; 22:587-9.
- Sundaram M., McGuire M.H., Schwajowicz F. Soft tissue masses: histologic basis for decreased signal (short T2) on T2-weighted MR images. *Am J Radiol* 1987; 148:1247-50.
- Lee T.H., Wapner K.L., Hecht P.J. Plantar fibromatosis. *Current Concepts Review. J Bone Joint Surg* 1993; 75A:1080-4.
- Enneking W.F. *Musculoskeletal Tumor. Surgery*, pp. 747-75. New York, Churchill-Livingstone, 1983.
- Sugiura I. Desmoplastic fibroma. Case report and review of the literature. *J Bone Joint Surg* 1976; 58A:126-30.
- Turek S.L. *Orthopaedics. Principles and their application*, pp. 829-30. Philadelphia, J. B. Lippincott, 1967.
- Wu K.K. *Surgery of the Foot*, pp. 154-156. Philadelphia, Lea and Febiger, 1986.
- Carnesale P.G. Soft tissue tumors. In *Campbell's Operative Orthopaedics*, edited by A. H. Crenshaw. Ed. 7. pp. 807-826. St. Louis, C. V. Mosby, 1987.
- Curtin J.W. Fibromatosis of the plantar fascia. Surgical technique and design of skin incision. *J. Bone and Joint Surg* 1965; 47A:1605-8.
- Wapner K.L., Ververeli P.A., Hecht P.J., Becker C.E. Plantar fibromatosis: a review of primary and recurrent surgical treatment. Read at the annual summer meeting of the American Orthopaedic Foot and Ankle Society, Asheville, North Carolina, July 1993.
- Landers P.A., Yu G.V., White J.M., Farrer A.K. Recurrent plantar fibromatosis. *J Foot Ankle Surg* 1993; 32:85-93.
- De Bree E., Zoetmulder F.A., Keus R.B., Peterse H.L., Van Coevorden F. Incidence and treatment of recurrent plantar fibromatosis by surgery and postoperative radiotherapy. *Am J Surg* 2004; 87:33-8.
- Allen P.W. The fibromatoses: a clinicopathologic classification based on 140 cases. *Am J Surg Pathol* 1977; 1:255-70.

Share with us your cases

Share with us your experience

Submit a case report now

