

CASE REPORT

INFANTILE DIGITAL FIBROMATOSIS A CASE REPORT

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ABSTRACT

Introduction: Infantile digital fibromatosis (FDI) is a rare, recurrent, benign tumor that occurs in the fingers and toes at birth or in the first months of life. We report a new case. **Case report:** A 2 1/2 year old girl had light-pink, non-painful, hard, solid tumor formations, sitting on the dorsal surface of the fifth metatarsal bilaterally and the 2nd phalanx of the 5th left finger, resulting in joint deformity and then functional discomfort. X-rays of the front and the profile of the fingers showed a chondrome of P2 of the 5th finger, without anomaly on the X-ray of the knees. On the ultrasound of the soft parts: presence of soft masses very hypoechoic avascular compared to the 5th ray measuring 2.81cm on the right and 1.21cm on the left. The diagnosis of infantile digital fibromatosis was retained. Surgical treatment was undertaken. **Conclusion:** The FDI is a rare and often easy diagnostic entity. Its recognition is important to consider adequate treatment knowing that this lesion can regress spontaneously and that post-surgical recurrences are frequent.

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INTRODUCTION

Infantile digital fibromatosis (FDI) or Reye's disease is a rare benign tumor, accounting for about 2.5% of the child's fibromatosis (1), occurring in the fingers and toes at birth or in the first few months of life. We report a new case with three FDI asynchronous locations.

OBSERVATION

A 2 and a half year old girl with no particular pathological history, consulted us for multiple tumors of asynchronous appearance at the extremities evolving since the age of six months which gradually increased in size and in whom the clinical examination objectified a first nodule measuring 1.5 cm well circumscribed, smooth and dome-shaped, covered with flesh-colored skin, of hard, painless consistency, at the expense of the 2nd phalanx of the 5th left finger (figure1), as well as two other nodules appeared 3 months later non-protuberant, 1cm pale pink each, not painful and sitting on the dorsal surface of the fifth metatarsals bilaterally (Figure 2). There is no reported functional impairment. The rest of the exam was without

abnormalities. The standard radiographs of the extremities were without particularity, notably no bone erosion or osteophyte. Ultrasound of the soft tissues: presence of hypoechoic and homogeneous tissue formations. The diagnosis of infantile digital fibromatosis was retained and confirmed by histology. Therapeutic abstention was advocated in our patient. An onset of spontaneous regression was reported after eight months of surveillance.

DISCUSSION

Infantile digital fibromatosis (FID) is a rare and benign tumor of myofibroblasts occurring on the fingers and toes during infancy [1] as in our patient. The majority of cases are sporadic [2]. Usually, she presents at birth or in the first two years of life, but has been reported to appear even until she is 52 years old. [3]

As a rare entity, available literature describing diagnostic criteria and treatment options for IDF is relatively rare. [1]

Reye defined these tumors for the first time in 1965, noting that they could be distinguished from other fibrous growths on the basis of their common anatomical location, age of onset, and clinico-histological features. [4]

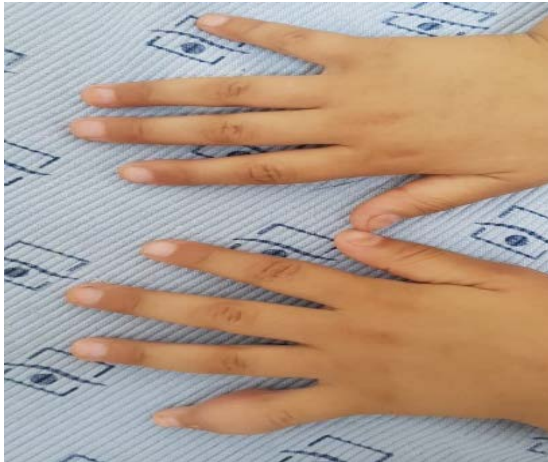


Figure 1: P2 nodule of the 5th left finger



Figure 2: tumor formations Bilateral of the 5th metatarsals

Simultaneous lesions involving several fingers have been reported. [5] Rarely, this lesion can appear outside the fingers. [6] as is the case with our patient. Clinically, IDF occurs in the first year of life as smooth, round, indurated confluent nodules up to 2 cm in diameter. Lesions can be single or multiple. The color of the skin can be pink to reddish. Initially, FDI lesions are indolent. They can follow a phase of rapid growth for several months, then the size of the tumor remains stable until a spontaneous regression occurs in half of the cases. [7,8,9] Some cases show a decrease or loss of joint function with long-term functional repercussions. [9 - 12] The histopathological diagnosis is generally based on some characteristic findings: limited cell proliferation in the dermis; intertwined fascicles extending to the epidermis; and tumor cells surrounding the adnexal structures. [13] The most pathognomonic feature is the presence of eosinophilic paranuclear inclusions consisting of an actin aggregate, although they are not necessary for FID diagnosis if the clinical picture and other histological findings are present. [13]

The conservative approach to spontaneous involution is the best strategy at present. [9] The surgical procedure was largely effective, reserved for symptomatic forms, of continuous growth and compromising the functional prognosis, although the recurrence occurs in 61% to 74% of the cases. [9,13] Other therapeutic approaches included topical steroids, intralesional steroids and intralesional 5-fluorouracil. [1,14,15] Spontaneous regression occurs over a period of several months to several years after the stabilization phase, with most regression occurring within one year (range, six months and five years). [16,17] No metastasis has ever been reported. [1]

CONCLUSION

The FDI is a rare entity and often easy diagnostic. Its recognition is important to consider adequate treatment knowing that this lesion can regress spontaneously and that post-surgical recurrences are frequent.

PATIENT CONSENT

Written informed consent was obtained from the patient for publication of this case report.

COMPETING INTERESTS

The authors declare no competing interests.

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