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Case Report

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Juvenile Fibroma: A Case Report and Review of the Literature

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ABSTRACT

Juvenile aponeurotic fibroma is a rare benign tumor which occurs in young patients and found primarily in the extremities. Clinical presentation is a unique, hard and painless tumour of the palm or sole which has a strong propensity to recur. The treatment commonly accepted for this locally recurrent tumour is complete excision with function preservation. With a close supervision to identify any recurrence or development of metastases at early stages.

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Introduction

Juvenile aponeurotic fibroma (JAF) is a rare, slow-growing, benign fibrous tumor that occurs in early childhood within the juvenile fibromatosis group, with well-characterized histopathological features [1,2]. We report the case of a patient with an aponeurotic fibroma of the finger of the hand in order to insist on the existence of this entity.

Case Report

A 9-year-old boy, with no medical history, presented since birth a small lump on the volar surface of his right hand on the base of the second finger. The family had noticed the lump for three year when it suddenly began to grow. The clinical examination finds a firm, nontender mass, infiltrating the skin, that extended to the radial and ulnar sides of the finger (Fig. 1), producing a 30° flexion deformity of the proximal interphalangeal articulation. A standard X-ray and à CT scan showed a subcutaneous mass extending to both edges of the finger with no anomaly of the flexor tendon or the bone (figure 2). A diagnosis of juvenile fascial fibroma was made after biopsy.

A surgical excision was performed respecting the peritendon and pedicles and sacrificing the skin in front with immediate coverage by a total skin graft, a diagnosis of juvenile fascial fibroma was confirmed on histological proof (figure 3). The patient has regained full flexion of the finger (figure 4). There has been no recurrence to date with a 9-month follow-up.

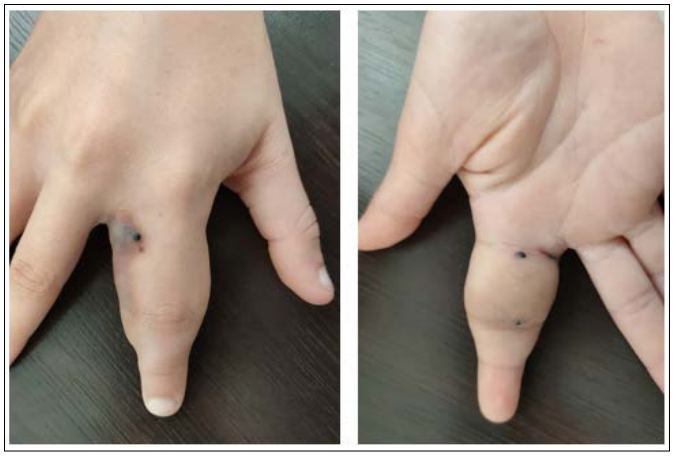

Figure 1: Fibroma of the Finger Infiltrating the Skin, Extended to the Radial and Ulnar Sides.

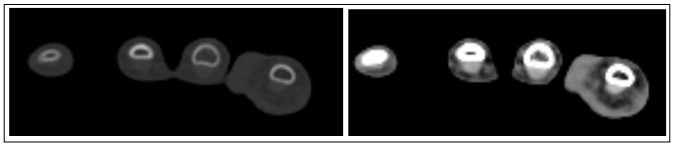

Figure 2: Axial Section of CT Scan Showed A Subcutaneous Mass Extending to both Edges of the Finger with no Anomaly of the Flexor Tendon or the Bone

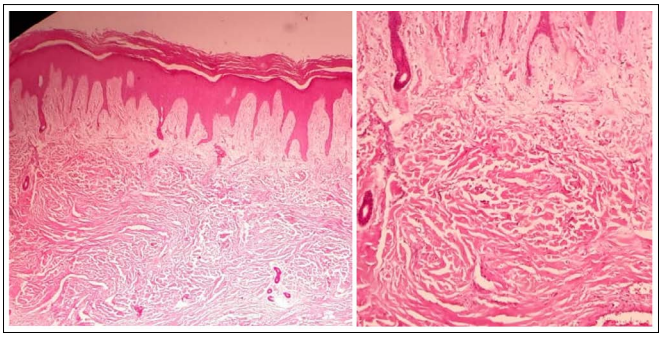

Figure 3: Benign Tumor Proliferation Made of Fibroblastic Cells Without Nuclear Atypia with Dense Collagenous Stroma in Accordance with a Juvenile Aponeurotic Fibroma.

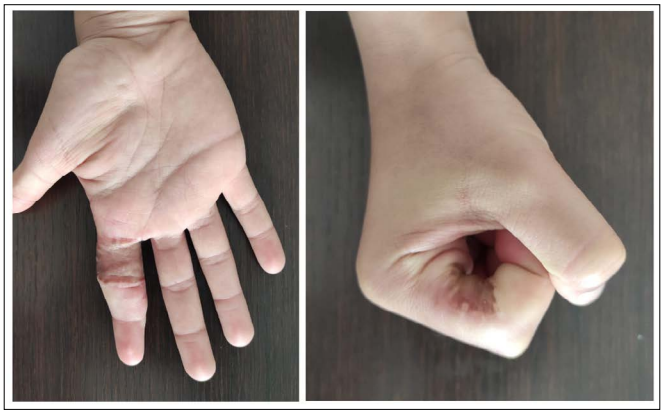

Figure 4: Surgical Excision Sacrificing the Skin in Front With Coverage by a Total Skin Graft(A), Complete Finger Flexion(B).

Discussion

Infantile digital fibromatosis represents 2% of pediatric fibroblastic tumors [3]. It was first described by Keasby who individualized it in the group of juvenile fibromatoses [4].

It usually occurs in children and adolescents, in the first or second decade of life (maximum incidence between 8 and 14 years of age, although more recently a larger age distribution has been described) [5,6]. It seems clear that this tumour develops predominantly in males [7].

It is a tumor that usually affects the deep palmar fascia, tendons and fascia of the hands, rarely the feet. In addition, less frequently affected sites include the neck, mandible, thigh, forearm, popliteal fossa and lumbosacral region [8].

It presents as a firm, grey-white, painless, fixed to the deep plane, slow-growing mass measuring less than 3 cm in diameter. It tend to infiltrate the surrounding tissue [8].

X-ray shows a soft tissue mass, with fine or progressive calcification. Signs of bone involvement may be seen, such as a periosteal reaction and a cortical defect. The scanner can visualize associated calcifications. Magnetic resonance imaging (MRI) is the most precise technique for preoperative planning and for assessing the extent of the lesion and margins [9].

The diagnosis of certainty is histological, this lesion is characterized by a diffusé, highly cellular proliferation of slightly pleomorphic, round-to-ovoid fibroblastic nuclei which do not show increased frequency of mitoses. Infiltration of surrounding fat and skeletal muscle by these cells is en- countered frequently, and there may be areas of dense collagenous tissue. Calcifications within the tumor may be evident microscopically and if present roentgenographically may give a preoperative indication as to the true diagnosis [10].

Complete removal of the tumor, without sacrifice of functional structures, will give the patient the best chance of cure [2]. However, recurrence rates are high (local recurrence exceeds 50%). Presumed malignant transformation and subsequent metastases are very rarely observed [5]. In case of recurrence, a new local excision without sacrifice of functional tissues is always indicated.

Most of the criteria of juvenile aponeurotic fibroma reported in the literature are found in our observation:

- Age of appearance in the first decade [5]
- Male sex [7]
- Location in the palm of the hand [8]
- Clinical appearance of a painless, hard, adherent tumor [8]
- Benign character and histological criteria [10]

The conservative treatment adopted in our patient is justified by the benign nature of the tumor. Close surveillance is recommended to identify any recurrence or early appearance of metastases.

Conclusion

Juvenile aponeurotic fibroma is a unique, rare, and benign soft-tissue fibroma usually with an asymptomatic clinical presentation. The standard of care for JAF includes correct diagnosis and conservative surgical treatment to maintain limb functionality. Surveillance of these patients is warranted to identify any recurrence or development of metastases at early stages.

Declarations

No conflict of interest

Written informed consent was obtained from the patient's legal guardian for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal."

- Availability of supporting data not available
- Competing interests
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