wjpmr, 2024, 10(5), 74-75

SJIF Impact Factor: 6.842

WORLD JOURNAL OF PHARMACEUTICAL AND MEDICAL RESEARCH

www.wjpmr.com

Case Study

ISSN 2455-3301

WJPMR

INFANTILE DIGITAL FIBROMATOSIS : A CASE REPORT

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Article Received on 14/02/2024 **Article Revised on 03/04/2024** **Article Accepted on 24/04/2024**

ABSTRACT

Infantile digital fibromatosis (FDI) is a rare, recurrent benign tumor that occurs in the fingers and toes of young children. We report a case of a 10-year-old child with a localization on the palmar surface of the 3rd right finger.

KEYWORDS: Infantile digital fibromatosis, fingers.

INTRODUCTION

Infantile digital fibromatosis is an uncommon fibroblastic and myofibroblastic tumour occurring in the digits of young children. This benign tumour commonly occurs in the dorsolateral aspects of toes and fingers excluding the thumb and big toes. Patients mostly present in the first two years of life and about a third of cases are congenital. Intracytoplasmic inclusions in the fibroblastic cells are characteristic features in this tumour. Recurrence rate post-surgery is high and spontaneous involution has been described in some cases.

OBSERVATION

- A.K 10-year-old girl, had a tumor of regular growth of the 3rd finger of the hand.
-the clinical examination of the hand found a dorsolateral mass of the 3rd finger straight, painless, not restricting the movements of the fingers measuring about 4 cm, The lesion does not regress.
-Ultrasound was done objectifying a mucoid cyst.
-MRI of the 3rd finger was related to a nodular lesion of the palmar surface of the 3rd ray may be related to a nerve origin.
- An excision with a skin graft was performed.without recurrence of the initial injury. After 3 years of clinical monitoring.
- The histopathological examination was in favor of digital fibromatosis.

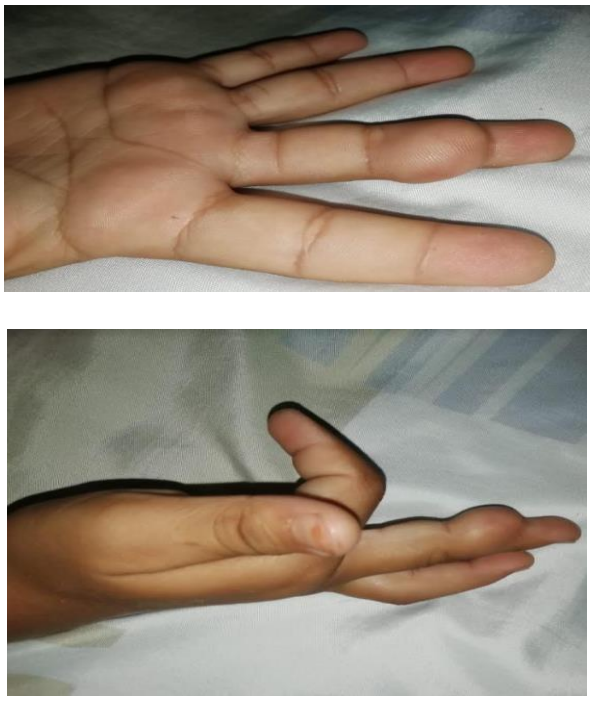

Figure 1: Pictures Showing Infantile Digital Fibromatosis.

DISCUSSION

Infantile digital fibromatosis is also called Reye's Tumour and inclusion body fibromatosis. It is a benign tumour first extensively described by Reye as recurring digital fibrous tumour in 1965. It occurs predominantly in digits, has a high recurrence rate and is characterised by intracytoplasmic inclusions on histology. The aetiology of inclusion fibromatosis is unknown. A viral aetiology was initially suspected because of the presence of cytoplasmic inclusions but polymerase chain reaction and electron microscopy have ruled out this

possibility.Inclusions are now known to be composed of densely packed vimentin and actin filaments and is thought to arise due to aberrant assemblage of microfilaments in myofibroblasts and smooth muscle cells.

The tumour may be single or multiple, firm or gelatinous in consistency. Nodules occur in the dorsolateral aspects of toes and fingers excluding the thumb and big toe. Although, tumours with similar appearance have been described in extra digital sites such as the breasts and thorax; the fingers are moreoften involved than the toes and rarely it may be associated with pain or functional impairment. often involved than the toes and rarely it may be associated with pain or functional impairment.

-Radiographs show a non-specific soft tissue mass, the underlying bone may be involved with either erosion or invasion but only rarely. The tumour is benign, although recurrence following surgery is high with rates of up to 60%.Recurrence occurs at the same site and sometimes a second tumour may develop in an adjacent finger or toe.

-Histological features are unique, interdigitating fascicles of spindle cells and collagen fibres make up the tumour which is located in the dermis and may extend into the subcutaneous tissue.Intracytoplasmic inclusions can be seen on H&E at high power.Histochemical stains are useful in making a diagnosis.

-Due to the benign nature of the tumour, recent studies recommend a conservative management with regular monitoring. This was not done in this case as there was an increase in the size of the tumour and diagnosis was made after surgery. Waiting for spontaneous regression might be a little difficult in this environment as attendance at follow up clinics is notoriously poor and patients would often seek traditional medical practitioners who may further complicate the lesion. Cases with spontaneous involution have been reported in literature, it usually occurs within 2 to 3 years.Other reasons to consider in conservative approach are the highrate of recurrence and the possible occurrence of deformities in the digits following surgery. Surgery is indicated where there is deformity and functional impairment.

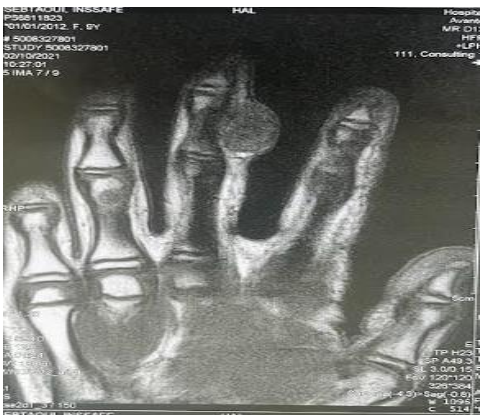

Figure 2: MRI showing Infantile digital fibromatosis of the 3rd finger.

Figure 3: Results 3 Years After Surgery.

CONCLUSION

Infantile digital fibromatosis is a rare entity that is often easy to diagnose because of its clinical and histological features. Its recognition is important to consider an appropriate treatment, knowing that this lesion can regress spontaneously and that post-surgical recurrences are frequent.

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