



Infantile Digital Fibromatosis- Rare Benign Tumour!

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ABSTRACT

Background: Infantile Digital Fibromatosis is a rare benign fibro proliferative tumour of early childhood. Case characteristics: A six-month-old boy with swelling on the right third digit histopathologically fibromatosis. Observation: Congenital onset in our patient. Message: Reassure about benign nature to avoid unnecessary surgical intervention.

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Introduction

A rare benign fibro proliferative tumour of early childhood- Infantile Digital Fibromatosis was first described by Reye as late as in 1965, with just 250 cases reported till date¹. The lesion involves the distal phalanx of the toes and fingers with equal frequency except of the thumbs and great toes. Very rarely involvement of some extra-digital sites have been reported to be involving the hands, arms, feet, nose, breasts and tongue. Congenital onset, although rare, is described also, as observed in our patient. None of the lesions have been reported to have malignant transformation or metastasis.

Case Report

A six-month-old boy presented with progressively increasing diffuse swelling on the dorsum of the right third digit. It was noticed soon after birth. There was no history of trauma or inflammation. No allergic history was reported. The other hand and both feet were unremarkable. None of the family members had experienced similar lesions. There was no functional impairment and the size of the lesion was increasing with age which caused parental anxiety. A 3-mm punch biopsy was performed under local anaesthesia, and the histopathology revealed fibromatosis affecting the reticular and adventitial dermis, arranged as sheets and interlacing bundles of eosinophilic myofibroblasts set in a collagenous background. No mitotic figures and no atypical cells were observed. MRI findings revealed diffuse circumferential soft tissue signal intensity encasing proximal phalanx & metacarpal joint of middle finger appearing hypointense on all sequences suggestive of fibromatosis. The current management of IDF recommends avoiding surgical intervention, as spontaneous involution of IDF is the rule². Parents were reassured and child is under follow up for 1 year in which the tumour has showed no increase in size.



MRI-Diffuse circumferential soft tissue signal intensity encasing proximal phalanx & metacarpal joint of middle finger appearing hypointense suggestive of fibromatosis. The neurovascular bundle are encased. The underlying proximal phalanx is thin in girth possibly remodelling due to pressure effect

Discussion

Benign asymptomatic nodular proliferation of fibrous tissue- Infantile digital fibromatosis, occurs almost exclusively on the dorsal and lateral aspects of the toes or the fingers. Although, the etiology is still a dilemma, it has been suggested that bone morphogenetic protein, which is a member of transforming growth factor- β super family has a role in it. Deregulation of this bone morphogenetic protein leads to various apoptotic pathways, which may explain the location of these lesions at the sites of digital septation. It is the transforming growth factor- β 1 which mediates myofibroblast differentiation from fibroblasts^{2,3}. In this disorder, myofibroblasts are the primary cell type. Mostly the nodules appear in the initial months of life with upto 75-80% in the first year of life, although only one third are

congenital. The lesions tends to spontaneously involute without scarring mostly with rare cases of ulceration, deformity or functional impairment. Skin biopsy is needed for confirming the diagnosis which shows the characteristic perinuclear eosinophilic cytoplasmic inclusion bodies along with interlacing fascicles of spindle-shaped myofibroblast cells and collagen bundles. Immunohistochemical stains positive for cytokeratin, vimentin, calponin, desmin, and alpha-smooth muscle actin⁴.

Importance of early recognition lies in the fact that it will avoid unnecessary surgical intervention, which may be a potentially aggravating factor, unless and until serious cosmetic or functional concerns intervene. Its natural course shows a slow growth in the initial months, followed by rapid growth over a year and finally spontaneous resolution over 1-10 years (average 2-3 years)^{2,4}. Recurrence rate of upto 60% has been observed even with wide local excision⁴. Mohs micrographic surgery is recommended in case of functional impairment, which also has regular recurrences⁵. Henceforth, it is desirable to avoid surgical treatment unless severe dysfunction is observed⁶. With regards to medical management, topical corticosteroids hasn't shown any benefit whereas intralesional corticosteroids and fluorouracil has controversial benefits⁷. Literature supports clinical surveillance without any active intervention, as spontaneous regression is known to occur in 2-3 years. Prognosis being excellent, the parents need to be reassured of the same and its benign nature.

Conclusion

It is important that we paediatricians being the first contact point for the parents are aware of this rare condition which has an excellent prognosis so as to avoid unnecessary surgical referrals and intervention and thus recurrence. Parents should be informed of the benign nature of IDF. Parental reassurance is the most important management.

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