

Case Report

Infantile digital fibromatosis: A rare fibrous tissue disorder of infancy and childhood

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Abstract Infantile digital fibromatosis is a rare benign asymptomatic nodular proliferation of fibrous tissue occurring almost exclusively on the dorsal and lateral aspects of the fingers or the toes and starts mostly in early months of life. A little more than hundred cases have been described in literature. We report such a case in a 3-year-old child, in whom, the disorder started in first year of life and restricted only to the fingers of the hand. A brief review of the literature is also presented.

Key words

Infantile digital fibromatosis, acquired digital fibromatosis, acquired digital fibrokeratoma, pachydermodactyly, reactive digital fibromatosis.

Introduction

Infantile digital fibromatosis (IDF) is a distinctive benign fibrous tumour of infancy. It usually presents in the first year of life and occurs exclusively in the fingers and toes but spares the thumbs and great toes¹. Reye first described it in 1965 as a recurring digital fibrous tumor.² Males and females are equally affected. Most nodules appear in the first few months of life; one third are congenital, and 75-80% are noted during the first year of life. Reports of IDF developing in older children and adults are rare.^{1,3} Patients are asymptomatic, without associated systemic symptoms. Single or multiple, firm, erythematous, dermal nodules with a smooth dome-shaped surface appear on the dorsolateral aspect of the distal phalanges of the digits. They can grow up to 2 cm in diameter, yet they rarely cause functional impairment or deformity. Rare

extradigital sites reported include the hands, the feet, the arms, the nose, the breasts, and the tongue.^{1,3,4} The cause of this condition is unknown. Although a viral etiology was initially suspected because of cytoplasmic inclusion bodies found on histologic examination, subsequent electron microscopy (EM) and polymerase chain reaction (PCR) findings do not support a viral cause.⁵ A skin biopsy is recommended to confirm the diagnosis. Unique histologic features are diagnostic of IDF. Interlacing fascicles of spindle-shaped cells and collagen bundles form a dermal nodule. The nodule may extend into the subcutaneous tissue. The ovoid nuclei of the cells are accompanied by characteristic perinuclear eosinophilic cytoplasmic inclusion bodies. Immunohistochemical stains positive for vimentin, cytokeratin, desmin, and muscle-specific actin, along with ultrastructural studies, point to a myofibroblastic origin for the cells. Radiographic examination reveals a noncalcified soft tissue mass. There is often phalangeal dysplasia but rarely bone erosion.^{1,3,5} IDF is a benign condition

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without evidence of malignant transformation or metastases. Spontaneous involution without scarring over an average of 2-3 years has been reported in approximately 12% of patients.⁶ Following surgery 60% of cases recur locally but the final prognosis is excellent and surgery is recommended only if impairment or deformity of the digits exists.⁷ Topical corticosteroids with or without occlusion have not shown any benefit; however, intralesional corticosteroids may prove beneficial.⁸

Case report

A three years old male child reported in out-patient department having history of multiple asymptomatic nodules on his finger. These started appearing on the dorsolarteral aspects of the fingers at the age of 5-6 months and gradually increased in size and number during next few months but were static for last about one year (**Figure 1**). Thumbs of the hands and all toes of the feet were spared and there were no similar lesions elsewhere over his body. There was some clawing of the fingers due to overlying fibrosis. He neither had a habit of chewing his fingers, nor there was any history of repeated trauma to finger. A likely diagnosis of IDF was considered and skin biopsy of one of the nodules was performed which revealed a dense mass of dermal fibrous tissue with interlacing spindle-shaped cells and collagen bundles (**Figure 2**). Perinuclear eosinophilic inclusion bodies were also seen on higher magnification. Clinical features and the histological picture confirmed our diagnosis of IDF. Considering benign nature of the disease and possibility of spontaneous resolution, his



Figure 1 Multiple, firm, nontender, erythematous nodules on dorsolateral aspects of the fingers are seen in a 3 years old boy.

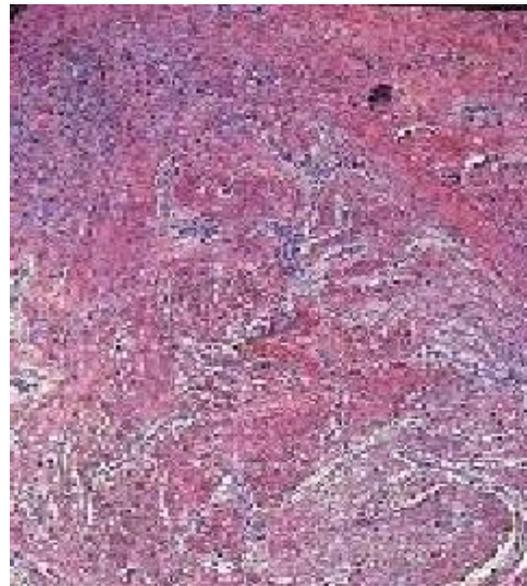


Figure 2 Dense fibrous tissue mass in the dermis showing interlacing spindle-shaped cells and collagen bundles. Perinuclear eosinophilic inclusion bodies are not seen at this magnification.

parents were reassured. No surgical or medical intervention was considered and he was advised yearly follow up in outdoor.

Discussion

Infantile digital fibromatosis, also known as acquired digital fibromatosis is quite rare and only around hundred cases have been described in dermatological literature³. In clinical practice, it is to be differentiated from other common benign acral tumors

typically reported as angiofibromas (AFs) or acquired digital fibrokeratomas (ADFs) or dermatofibromas. A recently described subset of digital fibromas (cellular variant characterized by dense cellular proliferation of spindle cells) can easily be misdiagnosed as dermatofibrosarcoma protuberans (DFSP) if immunohistochemical staining is not done. This variant is strongly CD34-positive as against DFSP or AFs which are strongly positive for factor XIIIa.^{3,5,9} IDF should also be differentiated from commonly occurring knuckle pads and pseudo-knuckle pads. Knuckle pads are discrete benign cutaneous lesions overlying the extensor surfaces of the fingers and hand joints and are unrelated to trauma, whereas pseudo-knuckle pads may be considered as a form of callosity that appears after repeated trauma. This type of knuckle pad has been described in children with obsessive behavior as "chewing pads" and in adults as occupational disorder. Moreover, pachydermodactyly is more generalized term used in literature to describe all these three entities. Other clinical differential diagnoses that should also be kept in mind include granuloma annulare, sarcoidosis, xanthomas, multicentric reticulohistiocytosis and neurilemoma.^{3,5,10,11,12}

Our case did not fit into any of the above differentials. Asymptomatic nodules classically appeared on dorsolateral aspects of the fingers during infancy. There was no evidence of knuckle pads, pseudo knuckle pads, xanthomas or any other symptomatic tumour mass. Clinical course of the disease and histology was almost confirmatory of the diagnosis.

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