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Case Report

A rare case of inclusion body fibromatosis

Maria Reji[1,*], **Sonia Mary Thomas**[1], **Jessy M M**[1]

[1] *Dept of Pathology, Pushpagiri Institute of Medical Science and Research Centre, Thiruvalla, Kerala, India*

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ABSTRACT

Infantile digital fibromatosis is a marked fibrous proliferation of infancy characterized by occurrence in the fingers and toes and a marked tendency for local recurrence. It accounts for 0.2% of registered soft tissue tumors. The lesion rarely exceeds 2 cm and tends to regress spontaneously. The presence of characteristic inclusion bodies in the cytoplasm of neoplastic fibroblasts distinguishes it from other fibromatosis. Current treatment recommendations include function-preserving excision and intralesional steroid injections for symptomatic lesions. Although lesions recur more than 60% of the time after excision, the ultimate prognosis is excellent.

1. Introduction

Inclusion body fibromatosis, more commonly known as infantile digital fibromatosis, most commonly presents as a small digital nodule in young children of both sexes.[1] In up to a third of patients, the lesion is congenital, and about 15% have multiple lesions that may affect both hands or feet, either synchronously or asynchronously. Strangely, the thumb and big toe always seem to be spared. No evidence of any family tendency can be seen. Recurrence is common after local excision, as well as the subsequent development of new lesions. Spontaneous regression will eventually occur mostly.[1–3]

2. Case History

A 1-year-old girl had a swelling on the dorsal aspect of her right middle finger since birth. Over the past 3 months, the swelling has gradually increased. (Figure 1). Physical examination revealed 1.5 x 1 cm nodule that was firm in consistency and was fixed to the skin. A USG performed showed soft tissue swelling extending to extensor expansion with ill-defined margins. The swelling was excised and sent for histopathological examination. Macroscopy showed a skin-covered nodule measuring 1.5 x 1.2 x 0.6 cm. The incision was ill-defined, pale white, and firm. Microscopy showed an ill-defined dermal neoplasm composed of spindle-shaped cells arranged in whorls, short bundles, and storiform arrays. Individual cells had slightly eosinophilic cytoplasm, elongated cytologically indistinct nuclei with pink to red intracytoplasmic paranuclear inclusions. The tumor extended from the superficial dermis to the subcutaneous tissue. Masson's Trichrome stain highlighted the red inclusion bodies. (Figure 2)

3. Discussion

Inclusion body fibromatosis is a benign, sometimes multicentric, myofibroblastic tumor with characteristic eosinophilic intracytoplasmic inclusions and the potential for local recurrence.[1] This tumor accounts for 0.2% of registered soft tissue tumors and 2% of pediatric fibroblastic tumors.[1] The classic subtype involves the dorsal or dorsolateral aspect of the distal or medial portion of the

* *Corresponding author*.
E-mail address: airammaria.reji@gmail.com (M. Reji).

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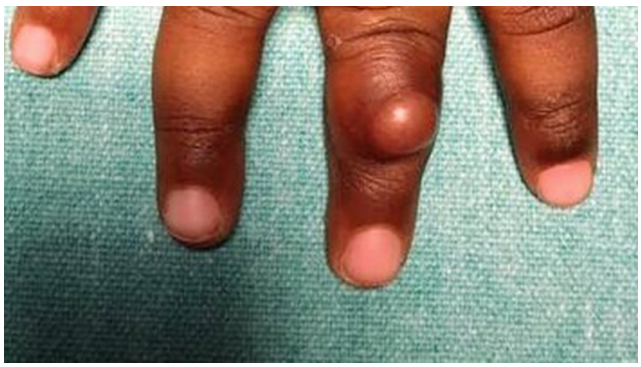

Fig. 1:

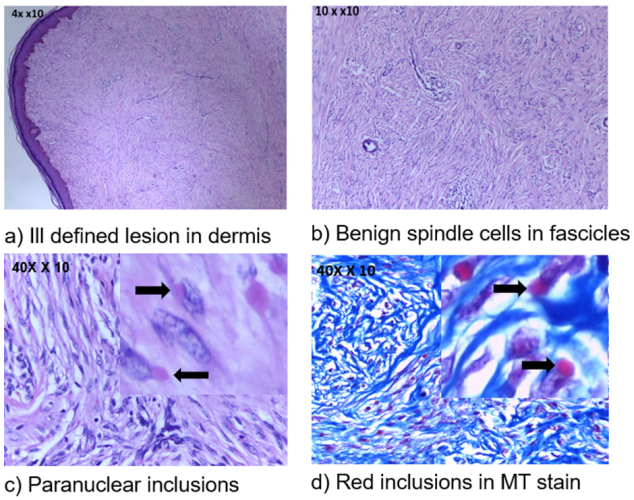

Fig. 2:

second, third, and fourth digits followed by the fifth digit, usually sparing the first digit, hand, and foot.[1,2] It presents as an asymptomatic dome-shaped or polypoid skin nodule, typically no larger than 2 cm.[3] Tumors are firm or rubbery, ill-defined protuberant or polypoid dermal nodules. The cut surface is fibrous and almost white to gray.[1–3] Microscopic images show spindle-shaped cells with slightly eosinophilic cytoplasm and an elongated cytologically unremarkable nucleus. Cells proliferate in whorls, interspersed with short fascicles or storiform fields in a variably collagenous dermis.[4] The cells contain 1.5–24 micrometer, rounded, pale pink to red intracytoplasmic inclusions that are often paranuclear deepening the nucleus and are enhanced with trichrome (red), phosphotungstic acid hematoxylin (dark purple), and Movat stains (pink).[2–4] Ultrastructurally, tumor cells contain intracytoplasmic bundles of actin-rich myofilaments. Recurrence rates between 61 and 75% have been reported in large series.[5] Current treatment recommendations include function-sparing excision or intralesional steroid injection for symptomatic lesions and clinical observation after diagnosis of asymptomatic tumors, as lesions may spontaneously regress.[6]

4. Conclusion

Inclusion body fibromatosis is a rare, benign, dermal myofibroblastic lesion with characteristic cytoplasmic eosinophilic inclusions usually found on the fingers of infants. Although lesions recur more than 60% of the time after excision; the final prognosis is excellent. This tumor does not metastasize. In our case, the lesion recurred 1 month after excision. A biopsy of the same showed similar histopathological findings. Regular check-ups were recommended to the patient.

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6. Conflict of Interest

None.

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Author biography

Maria Reji, PG Resident https://orcid.org/0000-0003-1419-0389

Sonia Mary Thomas, Assistant Professor

Jessy M M, Professor

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