Inclusion Body Fibromatosis

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Definition

Inclusion body fibromatosis (IBF) is a benign lesion classified among the fibroblastic—myofibroblastic tumors.

It is also known as infantile digital fibroma, infantile digital fibromatosis, and recurring digital fibrous tumor of childhood.

Epidemiology and Presentation

IBF accounts for 0.1% of soft tissue tumors and 2% of pediatric fibroblastic tumors. It generally develops within the first 2 years of life (30% of cases are congenital). This neoplasm, which equally affects males and females, rarely occurs in adults. IBF classically involves the digits (except the first digit) and (less frequently) the hand and foot. Extra-digital cases have been rarely reported.

Usually it presents as an asymptomatic dome-shaped or polypoid cutaneous nodule (generally <2 cm). Synchronous and metachronous lesions may occur.

Pathology

IBF is a predominantly myofibroblastic tumor characterized by eosinophilic paranuclear inclusions (hence the name). Spindle cells characteristically grow perpendicular to the epidermis within a variably collagenous dermis. The paranuclear inclusion is highlighted by trichrome (red), phosphotungstic acid-hematoxylin (dark purple), and Movat (pink) stains. Mitoses are rare, and atypia is absent.

Differential diagnosis may be needed with the following: infantile fibrosarcoma (not digits, usually >2 cm, more cellular, more mitotic figures, no inclusions) and infantile desmoid fibromatosis (rare on hand, usually >2 cm, more cellular, no inclusions).

S. Mocellin, *Soft Tissue Tumors*, https://doi.org/10.1007/978-3-030-58710-9_129

Biomarkers

Spindle cells express muscle-specific actin (MSA), calponin, and desmin, whereas they are negative for cytokeratin, estrogen receptor, progesterone receptor, and beta-catenin.

Prognosis

This is a benign tumor, although with a high potential for local recurrence (up to 60–70%). Spontaneous regressions have been reported.

Therapy

Surgical wide excision is the treatment of choice.

Suggested Readings

Fletcher (2020) WHO classification of tumours of soft tissue and bone, 5th ed.
Marks (2016) Infantile digital fibroma: a rare fibromatosis. Arch Pathol Lab Med 140(10):1153–1156