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Case for diagnosis

Caso para diagnóstico

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CASE REPORT

A healthy, 4-month-old male infant from São Paulo presented with a firm papule measuring 0.5 cm in diameter on the distal phalanx of the fourth toe of his right foot. This painless lesion was first noted one month previously and two weeks later another similar papule was found close to the initial one. His mother reported that there had been no other local symptoms. On physical examination, a firm, normochromic papule of 0.5 cm in diameter was found on the distal phalanx of the fourth toe of the child's right foot (Figure 1).

Radiographic examination showed no bone involvement. Histology revealed a proliferation of bundles of spindle cells in the superficial and deep dermis (Figure 2). A large number of pink-stained intracytoplasmic inclusions were shown at high magnification. The inclusions stained red with Masson's trichrome stain (Figure 3). Immunohistochemical analyses showed the lesion to be composed of myofibroblasts. Cells expressed HHF35 (muscle-specific actin), 1A4 (smooth muscle actin) and desmin. They were negative for CD68 and CD 34.



FIGURE 1: A normochromic, firm papule of 0.5 cm in diameter was observed on the distal phalanx of the fourth toe of the patient's right foot

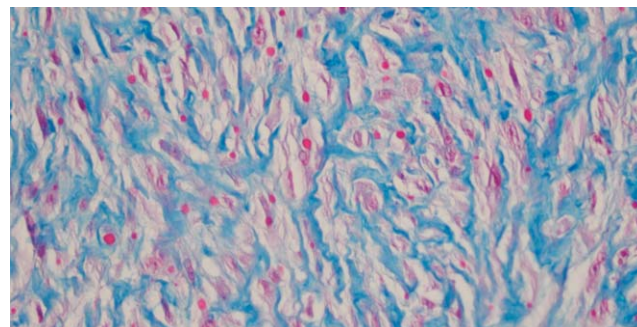


FIGURE 3: Inclusions stained red with Masson's trichrome stain

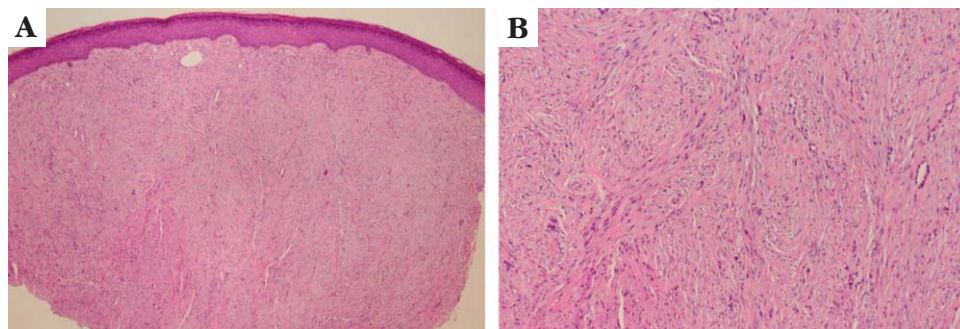


FIGURE 2: Histologic examination showing a proliferation of bundles of spindle cells in the superficial and deep dermis (HE 40X, HE 100X)

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DISCUSSION

Infantile digital fibromatosis, also referred to as Reye's tumor or inclusion body fibromatosis, is a rare, benign, fibroproliferative process that was first described by Reye in 1965.¹ This uncommon tumor accounts for 2.5% of all cases of fibromatosis.² The etiopathogenesis of the condition has yet to be fully clarified. In one-third of cases, the lesion is already present at birth,^{1,3} while in the remaining cases the lesions usually develop during the first year of life. The disorder is characterized by firm, asymptomatic, erythematous or normochromic, single or multiple nodules that develop on the digits of infants and young children. Classically, the condition spares the big toes and thumbs and rarely exceeds 2 cm in diameter.^{1,2,3} Histopathological examination shows a proliferation of spindle cells identified as myofibroblasts that are arranged in interlacing fascicles in the dermis. The finding that distinguishes this disease from other

forms of fibromatosis is the presence of eosinophilic intracytoplasmic inclusions in a varying number of myofibroblasts, which stain red with Masson's trichrome stain.^{2,3} The bodies of the intracytoplasmic inclusions are composed of actin and vimentin. Following diagnosis, treatment may consist of conservative, expectant monitoring, as the lesions tend to regress spontaneously. However, the exact rates of regression have yet to be defined.¹ Excision of the nodule is reserved for cases in which the function of the finger or toe is affected, but recurrence rates are high.¹⁻⁶ Cryotherapy is another treatment option.⁶ In the present case, after confirmation of the diagnosis, we opted for a conservative approach, since the lesion was not causing any functional impairment. After 6 months of follow-up, the nodules showed no signs of increasing in size. □

Abstract: Infantile digital fibromatosis or Reye's tumor is a benign fibroproliferative tumor, the etiopathogenesis of which has yet to be fully clarified. It typically presents at birth or in the first year of life and is characterized by a firm, flesh colored or erythematous nodule or nodules located on the digits. These lesions tend to regress spontaneously. **Keywords:** Child; Digitalis; Fibroma; Pediatrics; Solitary fibrous tumors

Resumo: A fibromatose digital infantil ou tumor de Reye é um tumor fibroso benigno de etiologia desconhecida, presente ao nascimento ou com aparecimento no primeiro ano de vida. Caracteriza-se por nódulos firmes, únicos ou múltiplos, normocrômicos ou eritematosos, localizados nas falanges, com tendência à regressão espontânea. **Palavras-chave:** Criança; Digitalis; Fibroma; Pediatria; Tumores fibrosos solitários

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