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Case Report

Monostotic fibrous dysplasia of the metacarpal: a case report☆

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ABSTRACT

Fibrous dysplasia is a bone disease characterized by abnormal differentiation of fibrous tissue in the bones; it is often asymptomatic. It may affect one bone (monostotic) or several bones (polyostotic). The monostotic form primarily affects the ribs, but hardly ever affects the hand. It is important to make the differential diagnosis with malignant bone tumors. This article describes the treatment and outcome of a rare case of a patient admitted with a history of tumor growth in the right hand, diagnosed as fibrous dysplasia of the right second metacarpal. Male patient, 14 years of age, admitted to the Sarah Hospital with lesion on the dorsum of the right hand without pain complaints, previous history of trauma, nor local signs of inflammation. Physical examination revealed swelling on the dorsum of the second metacarpal, painless, with unaltered mobility and sensitivity. Radiography, computed tomography, and magnetic resonance imaging indicated the involvement of the entire length of the second metacarpal: only the distal epiphysis was preserved, with areas of bone lysis. After biopsy confirmation, the patient underwent surgery, using a long cortical graft for reconstructing the metacarpal. During the follow-up period of five years there were no signs of recurrence, and proper digital growth and functionality of the operated hand were observed.

Displasia fibrosa monostótica em metacarpo – Relato de caso

Palavras-chave:
Doença
Metacarpo
Osteíte fibrosa cística
Displasia fibrosa óssea

RESUMO

A displasia fibrosa é uma doença óssea que se caracteriza pela diferenciação anormal de tecido fibroso nos ossos e é muitas vezes assintomática. Pode acometer um osso (monostótica) ou vários ossos (poliostótica). A forma monostótica acomete principalmente as costelas, mas raramente acomete a mão. O diagnóstico diferencial com tumores ósseos malignos é importante. O artigo descreve o tratamento e evolução de um caso raro de

☆ Study conducted at Hospital Sarah, Brasília, DF, Brazil.

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paciente admitido com história de crescimento tumoral na mão direita, na qual foi diagnosticada displasia fibrosa do segundo metacarpo direito. Paciente do sexo masculino, 14 anos, admitido no Hospital Sarah com lesão no dorso da mão direita, sem queixa álgica, antecedente traumático ou alteração flogística local. No exame físico, apresentava aumento de volume no dorso do II metacarpo, indolor, mobilidade e sensibilidade inalteradas. Foram feitos exames de radiografia, tomografia e ressonância magnética, evidenciou-se o comprometimento de toda a extensão do segundo metacarpo; apenas a epífise distal estava preservada, com áreas de lise óssea. Fez-se tratamento cirúrgico após a biópsia de confirmação, com o uso de enxerto longo cortical para reconstrução do metacarpo. Durante o tempo de seguimento de cinco anos não foram verificados sinais de recidiva; adequado crescimento digital e funcionalidade da mão operada foram observados.

Introduction

The term fibrous dysplasia was introduced by Lichtenstain in 1938 to describe the anomalous replacement of medullary bone by fibrous tissue. It is a benign lesion that may involve one (monostotic) or more bones (polyostotic), or be accompanied by other systemic alterations and endocrine disorders, such as in the McCune–Albright syndrome.[1,2] The etiology has been described as a mutation in the gene encoding the subunit α of the Gs protein located on chromosome 20q13.213.[3] The natural history of this lesion depends on its presentation; many lesions are asymptomatic, while others may cause pain, bone deformity, fractures, functional and cosmetic changes, and malignant degeneration. The monostotic form occurs with greater predilection for the long bones, ribs, and radius; few cases have been described in the hand.[1–4] The authors describe the treatment results and evolution of a rare case of monostotic fibrous dysplasia located on the second metacarpal of the right hand.

Case report

A male 14 year old patient was admitted to the Sarah Hospital with a lesion on the dorsum of the right hand with slow progression during the course of two years; the patient had no pain complaints, no previous history of trauma, and no local signs of inflammation. Physical examination revealed painless swelling on the dorsum of the second metacarpal with unaltered mobility and sensitivity. Radiography, CT scan and magnetic resonance imaging indicated the involvement of the entire length of the second metacarpal: only the distal epiphysis was preserved with areas of bone lysis. (Fig. 1). The levels of C3 199.0, C4 38.5, and alkaline phosphatase, as well as the chest X-ray were normal. The patient underwent an incisional biopsy disclosing fibrous dysplasia. The patient then underwent general anesthesia, plexus block, blod emptying on the upper limb using an Esmarch bandage and tourniquet positioning with pressure of 200 mmHg, and removal of the lesion on the second right metacarpal. Reconstruction was made using a 5-cm bone graft taken from the right fibula. The metacarpalphalangeal joint of the second finger was preserved, with a local margin from the edge of 1 mm; the fibula graft was proximally fixated with two transcortical titanium screws and distally with two crossed 1-mm Kirschner wires (Fig. 2). The procedure was bloodless. The material was sent to for anatomopathological examination, culture, and antibiogram. Prophylactic antibiotics were administered for 48 h. Patient was submitted to radiographic postoperative control (Fig. 3) and immobilization with circular antebrachiopalmar plaster for six weeks. Kirschner wires were removed after six weeks after bone graft healing, when physical therapy program was initiated. The transcortical screws were removed after five years, due to local pain complaints. The result of the histopathological exam indicated fibrous dysplasia with 46,XY,add(6)(q27),t(14:21)(q22;p1?11.2)[4]/46,XY[12] karyotype (Fig. 4). Follow-up continued for five years without recurrence, showing normal function of the operated hand.

Discussion

Fibrous dysplasia represents 7% of benign bone tumors, and its exact etiology is unknown. The monostotic form is more common and the radiographic findings are nonspecific.[5–7]

The etiology of the tumor remains unclear, but it appears to be linked to a single nucleotide mutation in the Gsα gene on the long arm of chromosome 20 (20q13.2-3), which results in a disturbance of the tissue differentiation process.[1,8] This mutation occurs in somatic cells some time after fertilization, and therefore is not inherited. Chromosome 12 has also been implicated in the pathogenesis of fibrous dysplasia; however, to date, no chromosomal abnormalities have been consistently demonstrated. The lesions in the long bones usually appear in the metaphysis as an intramedullary expansion with cortex thinning and hazy aspect; however, depending on the extent of the fibrous tissue and dysplastic changes in bone, as well as the degree of calcification, the findings may vary from sclerotic to radiolucent.[3,7] Clinically, these lesions are either characterized by volume expansion or asymptomatic. As in several tumors, the differential diagnosis should include sarcomas.

Radiographically, the differential diagnosis may include Paget's disease, solitary bone cysts, aneurysmal bone cyst, enchondroma, adamantinoma, low-grade intramedullary

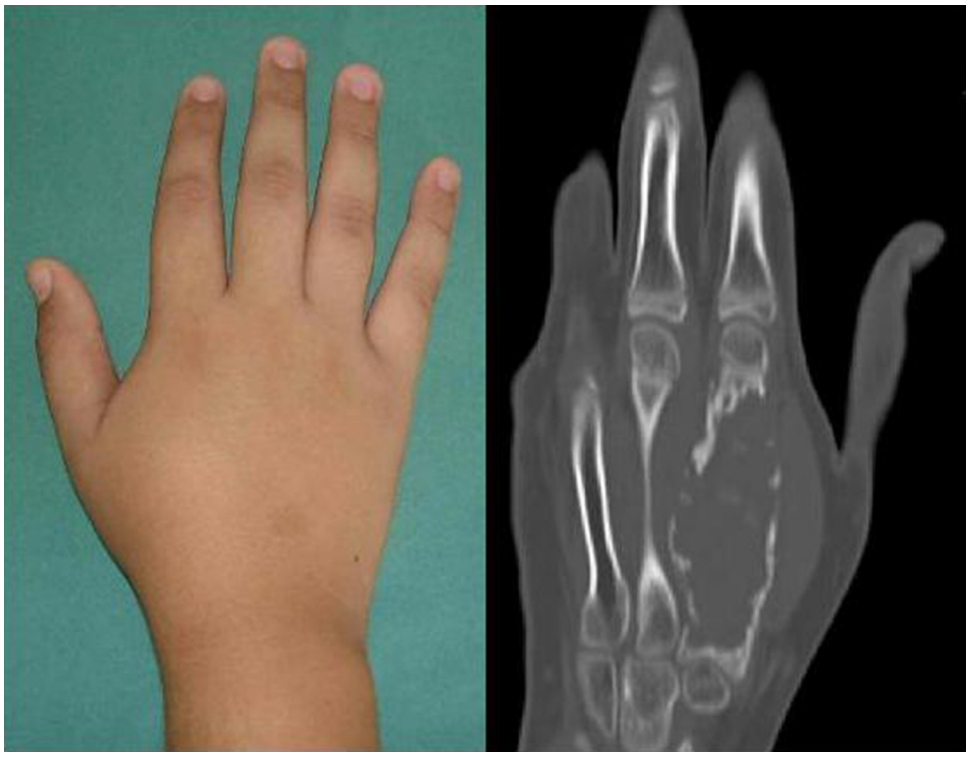

Fig. 1 – Physical and radiological examination showing the tumor on the second metacarpal.

osteosarcoma, osteofibrous dysplasia, and giant cell tumor.[3] The radiological findings suggestive of malignancy include lytic regions in previously mineralized areas, intralesional calcification, periosteal reaction, cortical disruption, and soft tissue invasion. Some aspects of these alterations were observed preoperatively in the present case. Moreover, the need for preoperative biopsy for the diagnosis of bone tumors should be emphasized.

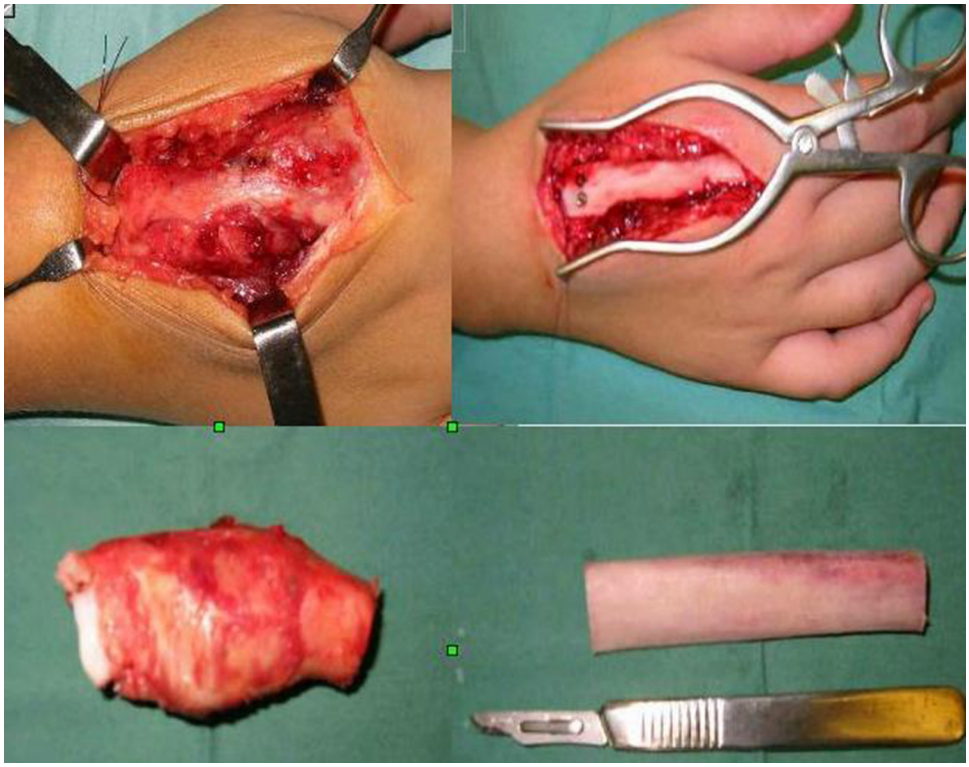

Fig. 2 – Intraoperative tumor excision and reconstruction with fibular graft.

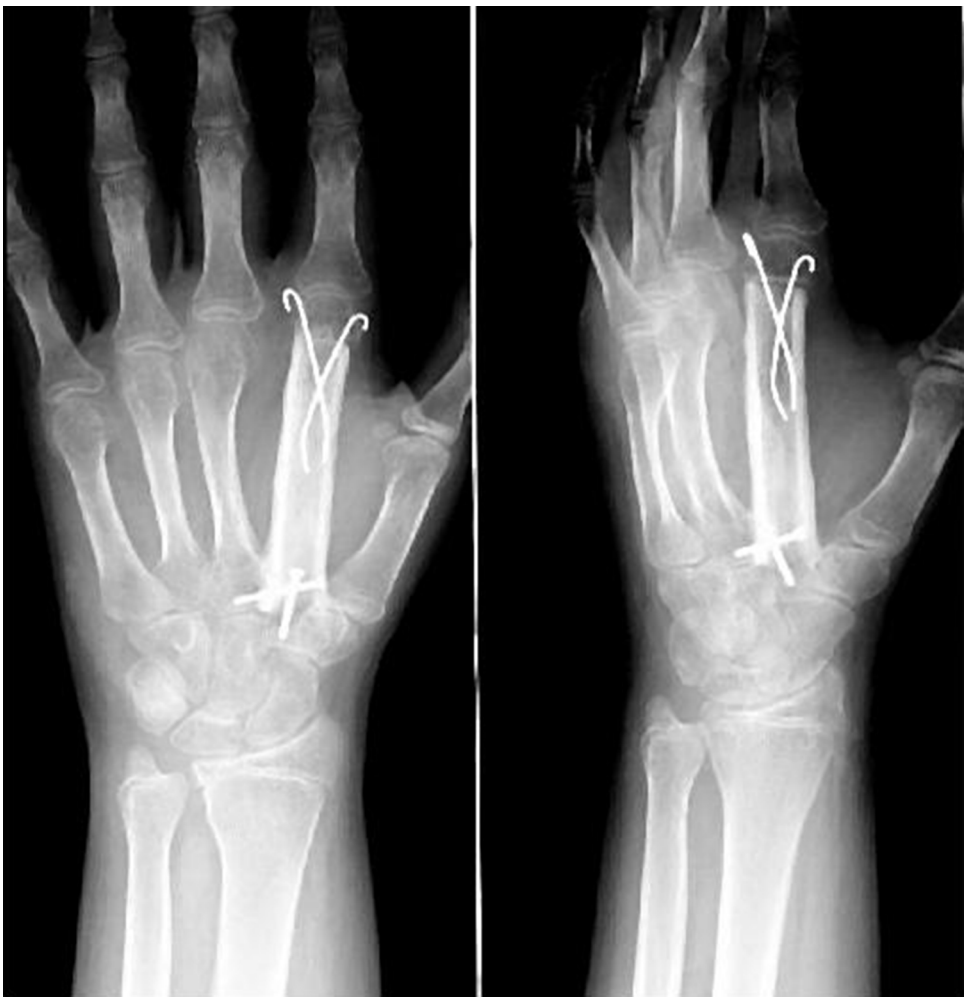

Fig. 3 – Radiological follow-up of the hand in the postoperative period.

Malignant transformation occurs with rapid bone growth in approximately 0.5% of patients with monostotic fibrous dysplasia and in 4% of those with McCune–Albright syndrome,[1,2] with osteosarcoma being the most common. Other tumors, such as fibrosarcoma, chondrosarcoma, or malignant fibrous histiocytoma, may also be observed. Histologically, low-grade osteosarcoma is more cellular, more atypical, and presents more mitosis, having a higher activity than fibrous dysplasia. Furthermore, the regularly spaced bony spicules seen in fibrous dysplasia are not present in osteosarcoma.[2]

The treatment of fibrous dysplasia for asymptomatic and stable lesions is regular follow-up. Surgery is indicated only for

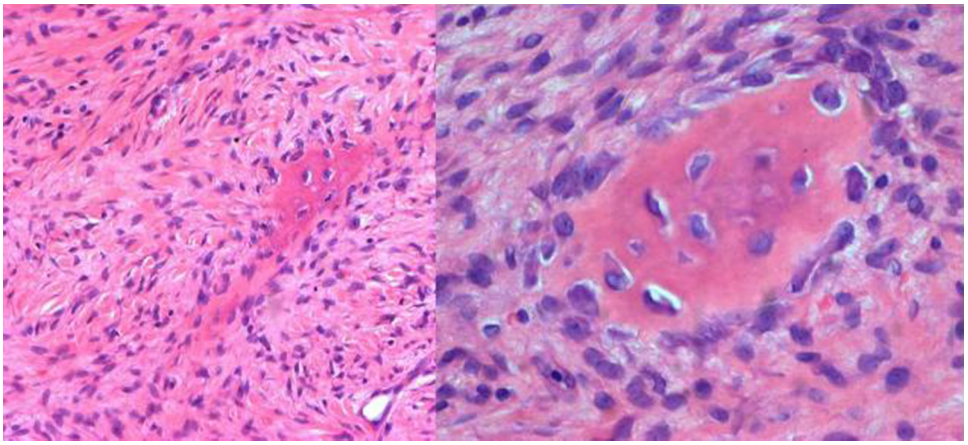

Fig. 4 – Anatomopathological exam (HE, 200×) showing fibrous tissue intermingled with fibrous bands.

confirmation biopsy, correction of deformities, non-operative therapy failure, prevention of pathological changes, and/or eradication of symptomatic lesions.[1,9–11] In cases of fractures, the treatment can be done with closed fixation. Other treatment options include curettage, curettage plus bone graft, or internal fixation.[9–11] More extensive cases may require bone graft or vascularized bone graft.[12]

In the present case, three important aspects should be highlighted: the first is the occurrence of second metacarpal monostotic dysplasia in the upper limb, a less common area; the second is the importance of the differential diagnosis with other lesions, including malignant degeneration; the third aspect is the treatment using free cortical bone graft, allowing for adequate bone length of the finger and normal function of the hand.

Conflicts of interest

The authors declare no conflicts of interest.

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