

# A Rare Case of Radiculopathy: Monostotic Fibrous Dysplasia of the Sacrum

## Um Caso Raro de Radiculopatia: Displasia Fibrosa Monostótica do Sacro



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### ABSTRACT

Fibrous dysplasia is a bone disease characterized by an osteoblastic dysfunction resulting in a fibrous replacement of the normal medullary bone. We describe the case of a 33-year-old who presented with low back pain irradiating to her right leg. Both the computed tomography scan and magnetic resonance imaging showed an osteolytic, multicystic lesion of the right hemi-sacrum with invasion of the right S1 foramen. She underwent foraminotomy and curettage of the lesion. Histological diagnosis was fibrous dysplasia, without features of malignant transformation. Three years after surgery the patient is asymptomatic and imaging is stable. This is the fifth known case of monostotic fibrous dysplasia involving the sacrum, a rare entity that must be considered in the differential diagnosis when approaching patients with sacral lesions.

**Keywords:** Fibrous Dysplasia, Monostotic; Radiculopathy; Sacrum

### RESUMO

A displasia fibrosa é uma doença óssea caracterizada por uma disfunção osteoblástica que resulta na substituição do osso medular normal por tecido fibroso. Descrevemos o caso de uma doente de 33 anos que se apresentou com lombalgia e cialgia direita. A tomografia computadorizada e ressonância magnética lombares revelaram uma lesão osteolítica e multiquística no hemissacro direito com invasão do foramen de S1. A doente foi submetida a foraminotomia e curetagem da lesão. O diagnóstico histológico foi de displasia fibrosa, sem sinais de transformação maligna. Três anos após a cirurgia, a doente está assintomática e os estudos de imagem apresentam estabilidade da lesão. Este é o quinto caso conhecido na literatura de displasia fibrosa monostótica com atingimento do sacro, uma entidade rara que deve ser considerada no diagnóstico diferencial de doentes com lesões do sacro.

**Palavras-chave:** Displasia Fibrosa Monostótica; Radiculopatia; Sacro

### INTRODUCTION

Fibrous dysplasia is a genetic, non-inherited bone disease, characterized by an osteoblastic dysfunction resulting in a fibrous replacement of the normal medullary bone and it accounts for approximately 7% of all benign bone lesions.<sup>1</sup> This disorder is subdivided in monostotic and polyostotic forms. Lesions are most commonly found in the long bones,<sup>2</sup> and spinal involvement in fibrous dysplasia is extremely rare.<sup>3</sup> In this article, we present a case of monostotic fibrous dysplasia affecting the sacrum and a review of the literature.

### CLINICAL CASE

A thirty-three-year-old female presented to our Department referring low back pain and right sciatica. One month earlier she had been evaluated in another institution and underwent lumbar computed tomography (CT) and lumbar magnetic resonance imaging (MRI) (Fig. 1) that demonstrated an expansive osteolytic lesion of the right hemi-sacrum with cystic component and compression of the right S1 nerve root. A CT-guided biopsy was then performed. The result was inconclusive and no diagnosis was established. The symptoms increased gradually and one month after onset she presented to our Department. She had permanent low back pain without alleviating position and right S1 radiculopathy associated with hypoesthesia and paresthesia.

No motor deficit was observed. The relevant medical history consisted of smoking habits. Laboratory studies showed normal blood cell count and normal C-reactive protein levels with no other remarkable changes. We proposed a surgical decompression of the right S1 nerve and biopsy of the lesion.

A right L5-S1 laminotomy, S1 foraminotomy and curettage of the lesion were performed with no complications.

Histology (Figs. 2 and 3) showed a dense fibrous tissue composed of spindle cells without cytologic anaplasia containing irregular trabeculae of metaplastic bone with scanty or no osteoblastic activity. The diagnosis of fibrous dysplasia was made.

The patient was discharged on the day after surgery with mild S1 paresthesia, no radicular pain and improved low back pain. A scintigraphy study using <sup>99m</sup>Tc revealed no other bone involvement besides the right hemissacrum. At 3 years of follow-up the patient referred occasional mild low back pain that was successfully treated with non-steroidal anti-inflammatory drugs. She had no radicular pain or paresthesia. Control MRIs showed stability of the sacral lesion at this point. No further treatment was considered.

### DISCUSSION

We found reference to four other patients with sacrum involvement of monostotic fibrous dysplasia. In the first

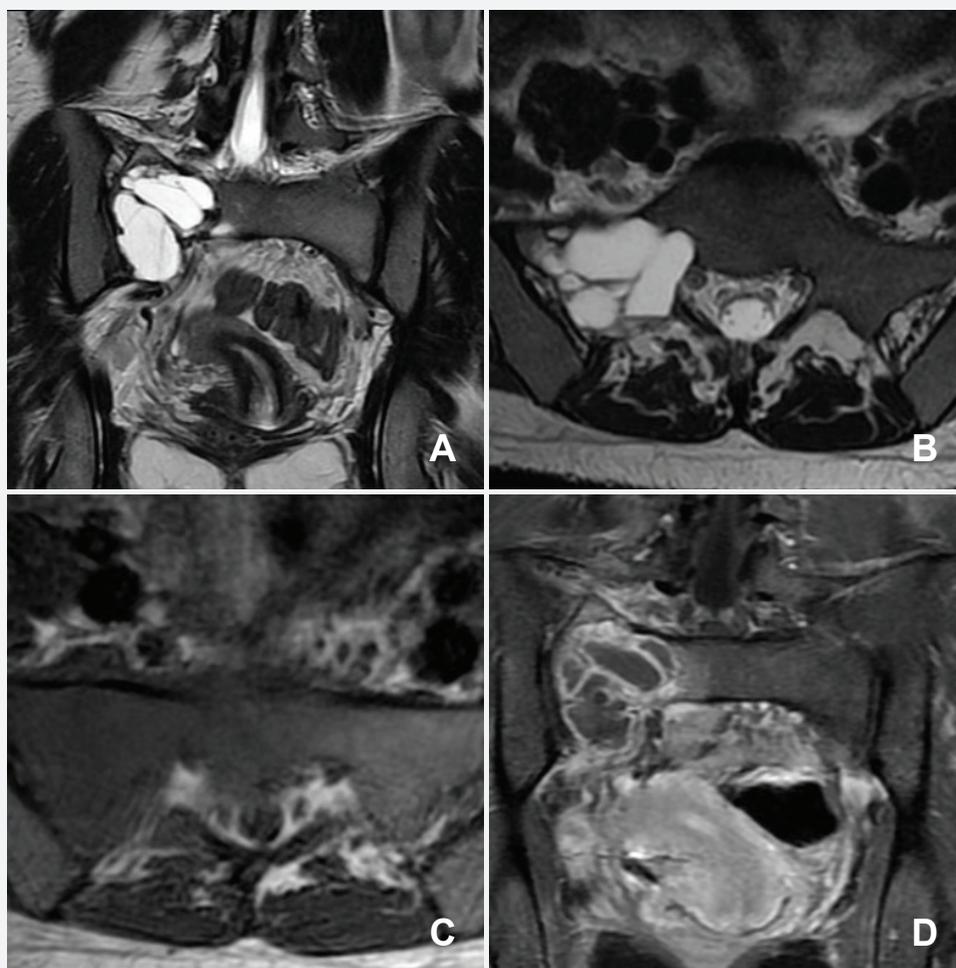
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**Figure 1** – Expansive lesion of the right hemisacrum with associated cystic component and compression of the right S1 nerve root. **(A)** Coronal T2 Weighted Image. **(B)** Axial T2 Weighted Image. **(C)** Axial T1 Weighted Image. **(D)** Coronal T1 Weighted Image post gadolinium

and Stutzman's review, a single case of monostotic sacrum fibrous dysplasia was mentioned with no other clinical information added.<sup>4</sup> Schoenfeld *et al* described three patients with monostotic disease of the sacrum.<sup>5</sup> Two of these patients underwent curettage and bone-grafting. A 51 year old female remained asymptomatic after 4 years and experienced reduction in the lesion size on plain radiograph. The other patient was a 32 year old female who referred mild paresthesia in the left foot, but no change on imaging was elicited after 14 years. One other patient, a 22 year old male that underwent biopsy and conservative treatment was asymptomatic and with no imaging changes at 8 years of follow-up.

Fibrous dysplasia comprises 2.5% of all bone neoplasms and about 7% of all benign bone lesions.<sup>4</sup> The specific origin of fibrous dysplasia is not completely understood. Differential diagnosis includes bone tumors, Paget's disease, hemangioma, aneurysmal bone cyst and giant cell tumor.

The monostotic form accounts for 70% to 85% of the patients.<sup>6</sup> Lesions are most commonly found in the femur, tibia, humerus, ribs, clavicle and craniofacial bones. Despite the higher frequency of the monostotic form, the axial skeleton is more often found in the polyostotic disease.<sup>7</sup>

Bisphosphonates, primarily pamidronate, have been used most extensively for treating polyostotic disorder. Several studies have demonstrated their benefit in reducing clinical pain.<sup>8</sup> Other drugs like opioids and non-steroidal anti-inflammatory drugs are also used to control pain. New therapies like the monoclonal antibodies denosumab (RANKL blocker) and tanezumab (anti-NGF) are still being studied for this disease in clinical trials.<sup>9</sup> Surgery can sometimes be necessary for confirmatory biopsy, correction of deformity, prevention of pathologic fractures or nerve root decompression.

Malignant transformation is a rare condition, occurring in less than 1% of the cases, with only one case reported in the monostotic disease of the spine. The most common transformation is osteosarcoma, followed by fibrosarcoma and chondrosarcoma. It has been suggested that the probability of transformation to osteosarcoma increases after radiation treatment.<sup>10</sup>

According to our literature search, this is the fifth case reported in the English literature. Our approach had a double objective: treating a neurological symptom and attaining a histological diagnosis. After the diagnosis, we requested a bone scintigraphy to determine whether the disease was

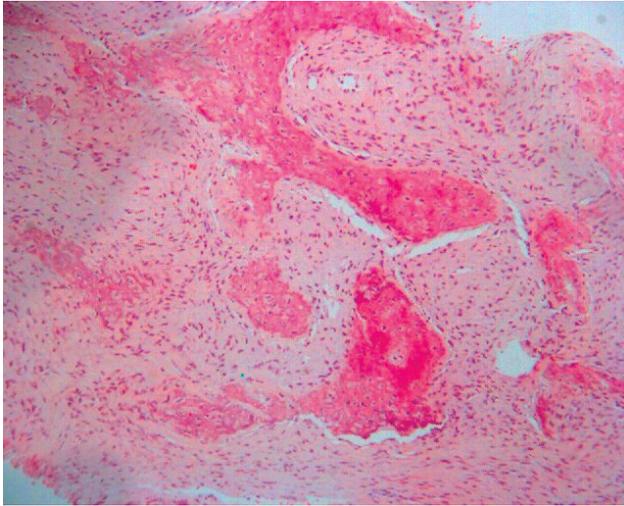


Figure 2 – H.E.x10. Small magnification of the lesion displaying fibrous tissue and bone trabeculae

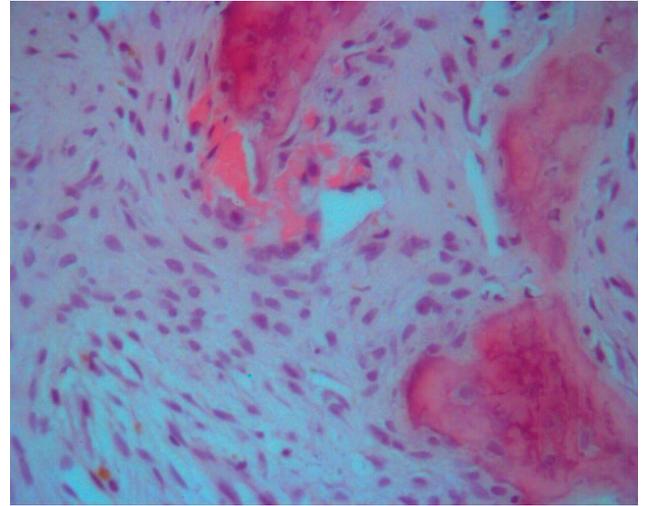


Figure 3 – H.E.x40. Spindle cells with a whorled arrangement and trabeculae of immature woven bone devoid of osteoblasts

monostotic or polyostotic. If in the presence of polyostotic form, the patient would be referred to a multidisciplinary team.

Although rare, FD must be considered as a possible diagnosis when approaching lytic lesions of the sacrum. In the known reported cases of monostotic sacrum FD, a minimally aggressive treatment was sufficient to obtain symptom relief when compared to aggressive treatments indicated in other sacral tumors. Despite its usual benign course, this disease requires watchful waiting due to the possibility of lesion progression and/or malignant transformation.

#### PROTECTION OF HUMANS AND ANIMALS

The authors declare that the procedures were followed according to the regulations established by the Clinical Research and Ethics Committee and to the Helsinki Declaration of the World Medical Association.

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#### DATA CONFIDENTIALITY

The authors declare having followed the protocols in use at their working center regarding patients' data publication.

#### PATIENT CONSENT

Obtained.

#### CONFLICTS OF INTEREST

All authors report no conflict of interest.

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