Fibrous dysplasia of the lumbar spine in a middle-aged woman. Case presentation

Julia M. Saidman, Damián Di Memmo, Alejandro Rasumoff
Diagnostic Imaging Department, Hospital Italiano de Buenos Aires (Buenos Aires, Argentina)

ABSTRACT

Fibrous dysplasia (FD) is a benign skeletal disorder characterized by abnormal development of fibrous tissue in a whorled pattern and containing trabeculae of immature non-lamellar bone. FD has two forms of clinical presentations: monostotic and polyostotic. Spinal involvement is seen mostly in the polyostotic form and is very unusual in the monostotic form. We present a 46-year-old woman that complained of right low back pain with a 2-month evolution. The definitive diagnosis was FD of the lumbar spine. Imaging testing revealed a lytic-cystic monostotic lesion with internal septa located in the posterior arch of the fifth lumbar vertebra, suggestive of Aneurysmal Bone Cyst (ABC). However, the anatomical pathology revealed FD as the final diagnosis. Conservative treatment was undertaken due to minimal symptoms and the absence of complications. Although monostotic FD of lumbar spine is rare, it should be taken into account among the differential diagnoses of a single osteolytic lesion. However, histopathology testing cannot rule out the coexistence of FB and ABC or a setting of FB secondary to an ABC.

Key words: Fibrous dysplasia; aneurysmal bone cyst; lumbar spine.
Level of Evidence: IV

Displasia fibrosa de columna lumbar en una mujer de mediana edad. Presentación de un caso

RESUMEN

La displasia fibrosa es una lesión ósea benigna caracterizada por el desarrollo anormal del tejido fibroso de disposición arremolinada, con trabéculas de hueso inmaduro no laminar. Se distinguen dos formas: monostótica y poliostótica. El compromiso de columna vertebral se asocia, con más frecuencia, a la variedad poliostótica; la forma monostótica es infrecuente. Presentamos a una mujer de 46 años que consultó por lumbocruralgia derecha de dos meses de evolución, con diagnóstico definitivo de displasia fibrosa de columna lumbar. Los estudios por imágenes mostraron una lesión monostótica lítico-quística con tabiques internos localizada en el arco posterior de la quinta vértebra lumbar. Sus características en los estudios por imágenes sugirieron un quiste óseo aneurismático, mientras que la anatomía patológica fue reveladora frente al diagnóstico final de displasia fibrosa. Los síntomas menores y la ausencia de complicaciones llevaron a indicar un tratamiento conservador. Si bien el compromiso de columna lumbar por displasia fibrosa monostótica es infrecuente, debería considerarse entre los diagnósticos diferenciales de una lesión lítica solitaria en dicha localización. No obstante, no se descarta mediante histopatología que pueda tratarse de un caso de coexistencia displasia fibrosa y quiste óseo aneurismático o que la displasia fibrosa se haya desarrollado sobre un quiste óseo aneurismático.

Palabras clave: Displasia fibrosa; quiste óseo aneurismático; columna lumbar.
Nivel de Evidencia: IV

Received on April 13th, 2019. Accepted after evaluation on October 7th, 2019 • JULIA M. SAIDMAN, MD • julia.saidman@hospitalitaliano.com.ar
How to cite this paper: Saidman JM. Fibrous dysplasia of the lumbar spine in a middle-aged woman. Case presentation. *Rev Asoc Argent Ortop Traumatol* 2020;85(2):167-173. http://dx.doi.org/10.15417/issn.1852-7434.2020.85.2.980

INTRODUCTION

The most frequent tumoral lesions of the spine are osseous metastases, multiple myeloma, and lymphoma.[1,2] Osseous lesions known as "tumor-like" lesions represent a small percentage of the bone conditions and include a wide range of benign processes that usually pose a challenge regarding diagnosis and treatment for the interdisciplinary medical team.[3,4]

FD is an uncommon fibro-osseous disorder, which results from the abnormal development of fibrous tissue characterized by a whorled pattern and containing trabeculae of immature non-lamellar bone. FD occurs in both children and adults, has an equal gender distribution, and accounts for 7% of bone tumors.[1,5]

The lesion is usually asymptomatic and an incidental finding, but many are discovered because of pain and local swelling, or complications ranging from a pathologic fracture to the rare case of sarcomatous transformation.[1,4,6]

FD has two forms of clinical presentations: monostotic and polyostotic. Although these forms share some radiographic and histologic features as well as anatomic sites of involvement, there are some differences.[1,4] The monostotic form of FD comprises 80% of all cases and most commonly affects ribs, femurs, tibiae, mandible, skull, and humeri.[6] The polyostotic form of FD most commonly involves the skull, facial bones, pelvis and spine, and may be associated with several syndromes, including the McCune-Albright syndrome (neuroendocrine abnormalities and cutaneous pigmentation) and the Mazabraud syndrome (intramuscular myxomas).[4,6]

The imaging characteristics suggestive of FD compose a highly variable types presentations of these lesions.[1,2]

Plain X-rays and CTs may show from sclerotic images with a characteristic "ground-glass" background and osteolytic lesions with sclerotic rims to expansile cyst-like lesions with a "blown-out" cortical shell.[1,6] FB MRIs have intermediate to low signal intensity on T1-weighted images and intermediate to high signal intensity on T2-weighted images, with heterogeneous enhancement following the IV administration of gadolinium.[1,6]

Histologically, the lesion characteristically appears well-circumscribed, composed of uniformly cellular fibrous tissue containing a proliferation of bland and uniform spindle cells with sparse mitotic activity. Scattered across the fibrous matrix are lamellae or rounded nests of woven bone without significant osteoblastic rimming.[4]

Treatment may be conservative since in many cases lesions remain stable over time. Radiation therapy involves a high risk of malignant transformation and should be avoided.[4]

Among the tumor-like osseous lesions, it's worth mentioning the ABC, which is one of the differential diagnosis of FD. ABC is a relatively rare lesion that represents 1.4-2.3% of primary bone tumors, it usually occurs in the second decade of life.[1]

The most common anatomic sites are the metaphysis of long bones (distal femur and proximal tibia) and the spine (3-20% of the cases).[4] Involvement of the thoracic spine is the most common, followed by the lumbar and cervical spine.[1,3] Spinal involvement is typically in the posterior arch and may extend into the adjacent vertebrae or intervertebral disk, the ribs, and the paravertebral soft tissue.[1]

Clinical presentation may vary according to the lesion location and extension. Most patients may present pain and swelling, and vertebral involvement may cause symptoms related to compression of the spinal cord or nerve roots.[1,2]

Imaging tests reveal its characteristics features. Radiographically, ABC commonly appears as an expanded, radiolucent lesion, with septa and well-defined margins. CT and MRI scans reveal fluid-fluid levels, indicative of hemorrhage with sedimentation.[1,2,4] MR imaging is the most sensitive method for detecting well-defined lesions with internal septation and different fluid-fluid levels, which are commonly represented as areas of increased signal intensity on T1-weighted images due to methemoglobin.[1,2] It's worth mentioning that fluid-fluid levels are not pathognomonic for ABC but are very suggestive of this diagnosis.[1,2]

Histologically, ABC is composed of blood-filled cystic spaces separated by cell stroma with osteoclast-like giant cells and osteoid.[1]

Treatment options largely depend on the localization and extension of the lesion. Although treatment by curettage and reconstructing the defect with bone graft remain the standard of care for ABCs, there is a range of treatment options that have been adapted to reduce the morbidity and mortality risk: the use of adjuvants (argon beam coagulation or phenol), alternative strategies (preoperative elective arterial embolization and adjuvant radiotherapy), and some promising emerging techniques, including percutaneous doxycycline, bisphosphonates, and denosumab.[7]

CLINICAL CASE REPORT

A 46-year woman whose relevant history only includes bilateral hearing impairment, the use of hearing aids since she was 12, and a cataracts surgery when she was 42.

She consulted at our Center for right low back pain, stabbing in nature, and with a 2-month evolution, for which she had been treated as an outpatient with kinesiotherapy and pain management. The patient reported no history of trauma/falls or cancer.

The case history and physical examination performed by the Department of Orthopedics revealed no further findings.

A lumbosacral spine CT showed the presence of a process of lytic appearance with sclerotic rims and complete thin internal septa, expansile and centered on the right pedicle of the fifth lumbar vertebra and extension into the vertebral body, facet and the right transverse process. Cortical thickening and thinning was observed with no extra-osseous components and with no intervertebral foramen involvement (Figure 1).

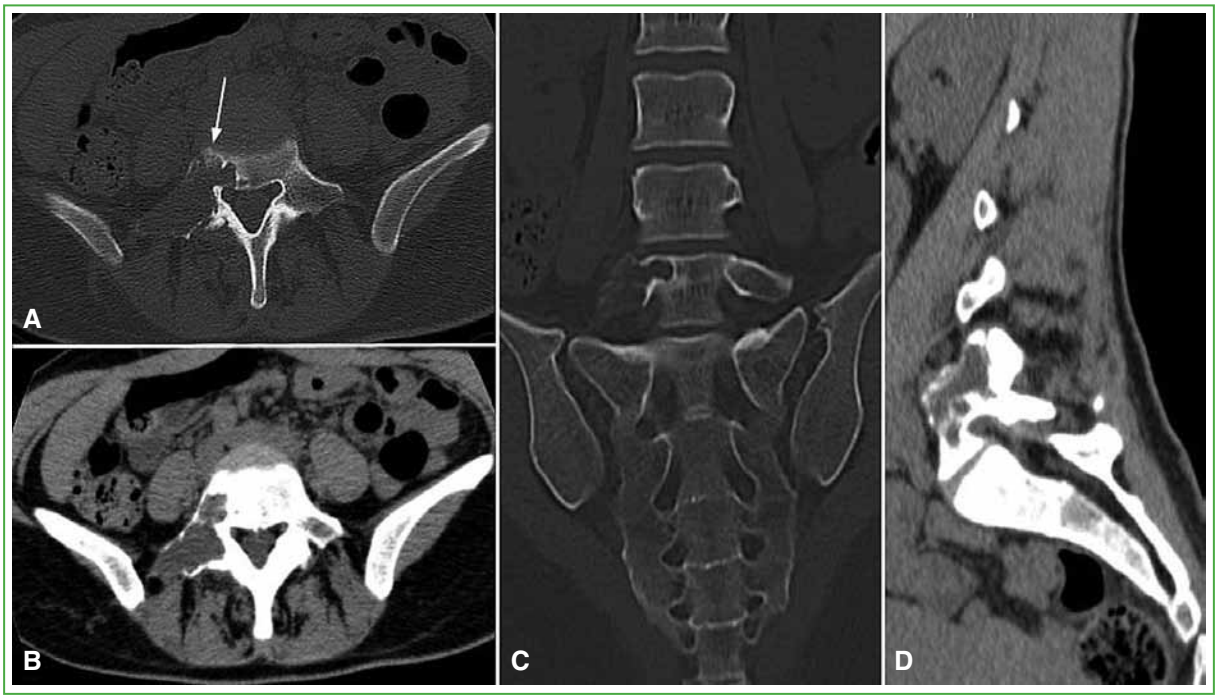

Figure 1. Computed tomography **A.** Bone window, axial section. **B.** Soft tissues, axial section. **C.** Bone window, coronal section. **D.** Soft tissues, sagittal section. Lesion of osteolytic appearance with sclerotic rims and complete thin internal septa, expansile with thinning of the cortex and centered on the right pedicle of L5 and extension into the vertebral body, facet and the right transverse process. Small "ground-glass" area (arrow).

An MRI with IV contrast of the lumbosacral spine showed the same expansile lesion of 24mm x 33mm x 16mm, containing fluid, internal septa and thinning of cortical bone, centered on the right pedicle of L5 and extension into the vertebral body, facet and the right transverse process (Figure 2).

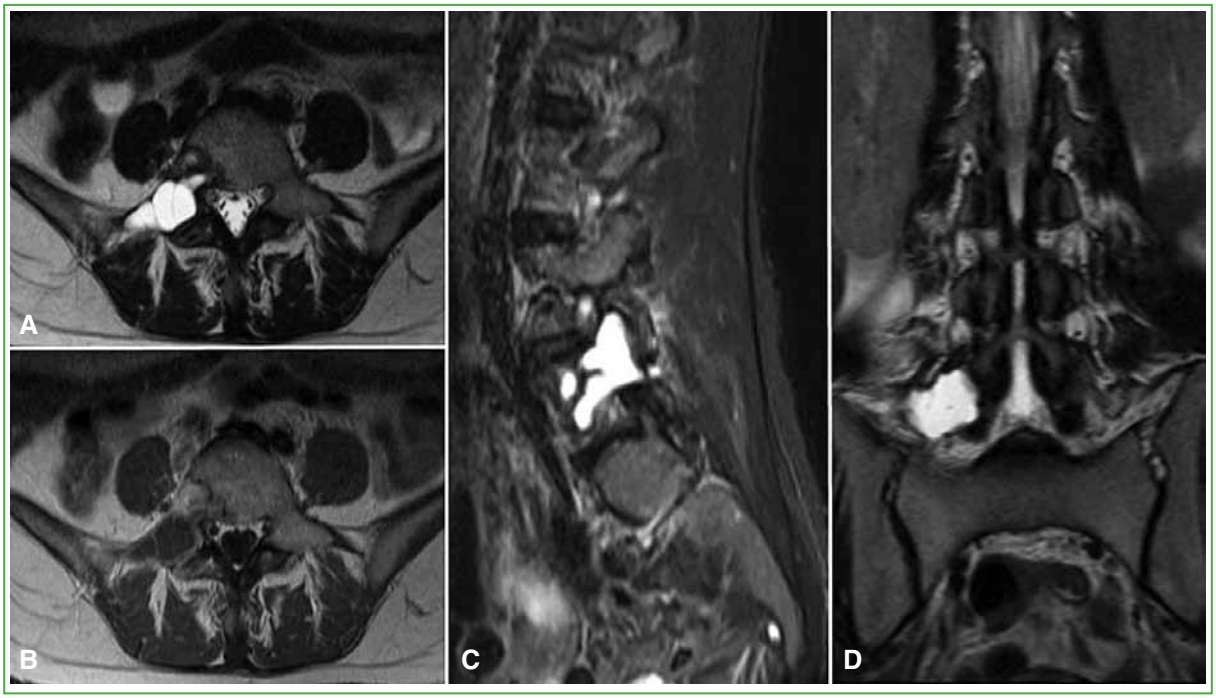

Figure 2. Magnetic resonance imaging. **A.** T2-weighted sequence, axial section. **B.** T1-weighted sequence, axial section. **C.** STIR, sagittal section. **D.** T2-weighted sequence, coronal section. Lesion of fluid signal intensity (hyperintense on T2-weighted and STIR sequences and hypointense on T1-weighted sequences) with internal septa, expansile with thinning of the cortex and centered on the right pedicle of L5 and extension into the vertebral body, facet and the right transverse process.

CT and MRI findings together with the lesion location led to considering an ABC setting.

A needle biopsy under CT guidance was performed through a right posterolateral access and produced a syringe containing 10 cm^3 of serosanguineous material for cytodiagnosis and a small bone biopsy block for histologic analysis. There were no complications during the procedure. The centrifugal cytology revealed abundant RBCs and blood elements, and bone with hypocellular blood material. The histopathological analysis revealed trabeculae of reticular bone, without significant osteoblastic rimming, in relation to fibrous nodular spaces, which led to the final diagnosis of FD (Figure 3).

The established diagnosis of FD caused the adoption of a watchful waiting strategy with a periodic clinical and imaging follow-up. The patient remains under follow-up evaluation 14 months after diagnosis, with an adequate symptomatic management and without associated complications.

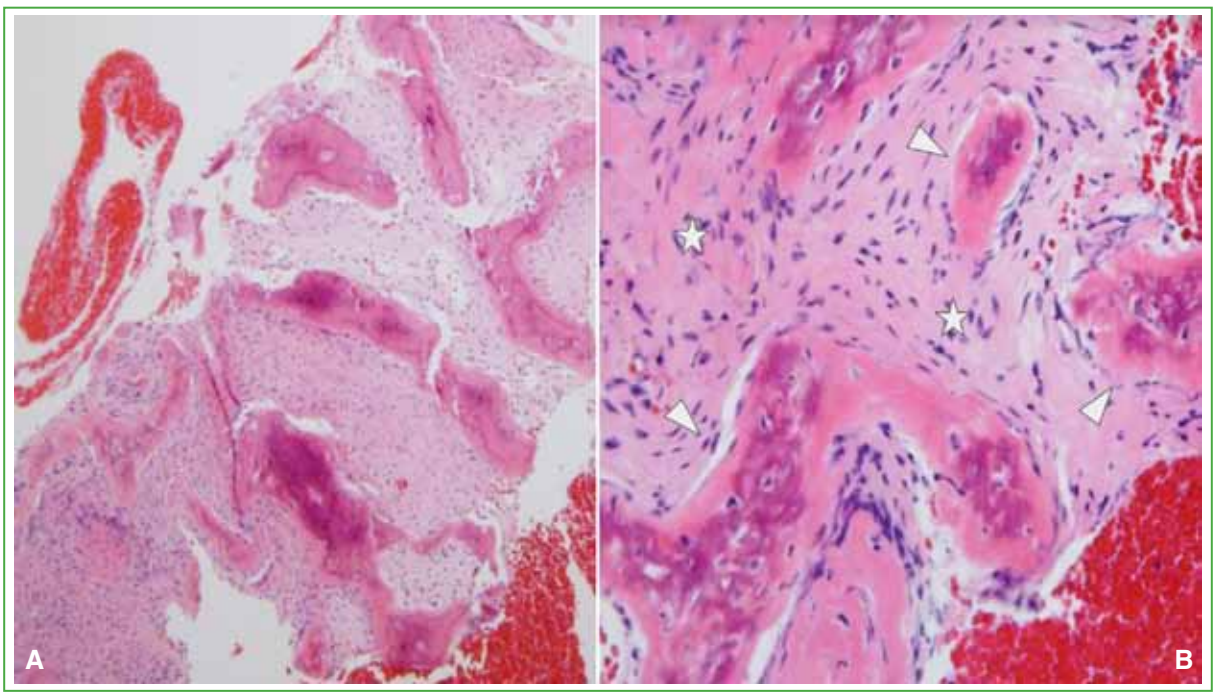

Figure 3. Anatomical pathology. **A.** 40x: **B.** 100x. Histologic sections reveal trabeculae of reticular bone, without significant osteoblastic rimming (arrow heads), in relation to fibrous nodular spaces (*), findings compatible with fibrous dysplasia.

DISCUSSION

FD and ABC belong to the group of benign tumor-like bone lesions.[3,4,8,9] Their forms of presentation, imaging features and locations may significantly vary, and, at the same time, may share imaging results. Thus, differential diagnosis must be done between these two conditions and the rest of the bone tumor disorders, until diagnostic confirmation by histopathologic finding.[3,4,10,11]

ABC may be a primary lesion or a secondary to a bone pathology with cystic degeneration (one-third of ABC cases), the most common of these conditions being giant cell tumor, followed by osteoblastoma, angioma, and chondroblastoma, FD and eosinophilic granuloma.[1] Thus, it should be taken into consideration that a lesion such as FD may undergo cystic degenerative changes and become an ABC formation.[1]

A third possible scenario is the coexistence of FD and ABC components in a single lesion, a very uncommon scenario with only a few reported cases. Buraczewski and Dabska reported 2 cases of FD and ABC coexistence from a total of 25 cases; one was localized on the posterior aspect of the third rib and the other one at the angle of the right mandible.[9] Martinez and Sissons studied 42 FD patients and found an ABC component on the fifth rib of one of them.[12] Lee *et al*. also reported a similar case of FD and ABC on the right frontoparietal region.[11]

Spine involvement of FD is rare, and, when it happens, it's usually the polyostotic form. Our case was that of a monostotic FD with spine involvement, thus adding up to a very rare scenario. Of the FD cases with spine involvement, only 13 involved the lumbar spine,[5,13] which enhances the uniqueness of the present case.

The differential diagnosis for monostotic bone lesions on the posterior spinal arch should include osteoid osteoma, osteoblastoma, osteochondroma, and ABC.[1] Osteoid osteoma manifests as a sclerotic lesion with a low-attenuation nidus and causes nocturnal pain; an osteoblastoma may be impossible to distinguish from an osteoid osteoma, although its central nidus is usually larger; and the osteochondroma is commonly an eccentric sessile-pedunculated lesion. Some malignant lesions should be considered as exceptions: chondrosarcoma, osteosarcoma, and Ewing sarcoma.[1]

The main limitation of our study is that the possibility of an intralesional ABC component cannot be ruled out since the biopsy sampling was performed through a posterolateral access of a single segment peripheral to the lesion; and thus, the FD-ABC coexistence also cannot be ruled out. Additionally, we cannot dismiss the possibility of an ABC to FD transformation within the evolution period, due to lack of previous studies and consultations.

CONCLUSIONS

We report a case of a monostotic lesion on the posterior spinal arch. The data obtained by the medical team to achieve a presumptive diagnosis (clinical picture and imaging testing) posed the strong suspicion that it was a classic ABC presentation. However, the definite pathologic diagnosis of FD was an eye-opening development. Although we must consider the most common epidemiological features and differential diagnoses when encountering a lesion on the posterior spinal arch, the possibility of lesions of rare localization and presentation should not be dismissed.

Conflict of interests: The author claims she does not have any conflict of interests.

D. Di Memmo ORCID ID: https://orcid.org/0000-0002-1675-964X
A. Rasumoff ORCID ID: https://orcid.org/0000-0002-7505-501X

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