Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Radiology

Radiologic Features of Monostotic Humeral Fibrous Dysplasia: Case Report and Literature Review

Adam Hajjine[1*], Meriem Ouali Idrissi[1], Badr Boutakioute[1], Najat Cherif Guennouni Idrissi[1]

[1]Radiology Department of Mohamed VI University Hospital, Marrakech, Morocco

DOI: 10.36347/sjmcr.2022.v10i03.023 | **Received:** 21.02.2022 | **Accepted:** 25.03.2022 | **Published:** 30.03.2022

***Corresponding author:** Adam Hajjine
Radiology Department of Mohamed VI University Hospital, Marrakech, Morocco

Abstract **Case Report**

Fibrous dysplasia is rare benign sporadic non inherited disease that can affect any bone, but a predilection for ribs, craniofacial and lower limb bones is described in the literature. Only few cases concerning the humerus have been reported. Radiographically, typical lesions appear as an area of radiolucent ground glass matrix**.** Here, we present a case report of a fibrous dysplasia located in the humerus revealed by chronic elbow pain in a 63 years-old woman. The imaging findings are displayed along with a literature review. When it's symptomatic, fibrous dysplasia could be revealed by common symptoms such as pain and bone deformity. Therefore, the radiologist plays a crucial role in diagnosis assessment, also in monitoring the lesion for early detection of complications especially malignant transformation.

Keywords: Fibrous dysplasia, imaging, monostotic, humerus.

INTRODUCTION

Fibrous dysplasia (FD) is a benign sporadic non inherited disease caused by a mutation of the α-subunit of the Gs-protein, resulting in replacement of normal bone by a fibrous tissue containing a newly formed mineralized product [1].

It's a rare condition accounting for 2,5% of osseous lesions, probably underestimated due to asymptomatic forms. However, when it's symptomatic, clinical features include pain, deformity and functional impairment [2].

In 80% of cases, FD concerns one bone (monostotic form) with rib, femur and skull as the most common sites of involvement. It may also present as polyostotic form or occur as a part of McCune Albright syndrome or Mazabraud syndrome. Radiographically, typical lesions appear as an area of radiolucent ground glass matrix. A cystic or sclerotic feature can also be encountered [3].

Here, we present a case report of a fibrous dysplasia located in the humerus revealed by chronic elbow pain in a 63 years-old woman. The imaging findings are displayed along with a literature review.

CASE REPORT

A 63 years-old female patient was referred to radiology department of Mohamed VI University hospital with a one year history of chronic right elbow pain. The patient didn't report any trauma. On physical examination, no deformity or joint motion limitation was noticed.

The laboratory test didn't show any abnormalities. Plain X-Ray of the right humerus including shoulder and elbow showed an oblong intramedullary lytic lesion with sclerotic rim within a ground glass opacity area. It occupied the middle third of the humeral diaphysis responsible of bone enlargement with no cortical rupture or periosteal reaction. No joints abnormalities were found (Figure 1). On Computed Tomography (CT), the lytic intramedullary lesion measured 12,4 x 1,2 x 1,3 cm without enhancement and its mean density was 30 UH suggesting its cystic nature (Figure 2). For further characterization, a Magnetic Resonance Imaging (MRI) was performed which confirmed the cystic nature of the lesion as it was hypointense T1 and hyperintense on both T2 weighted images and STIR (Figure 3). Both CT and MRI discarded an extension to surrounding soft tissues.

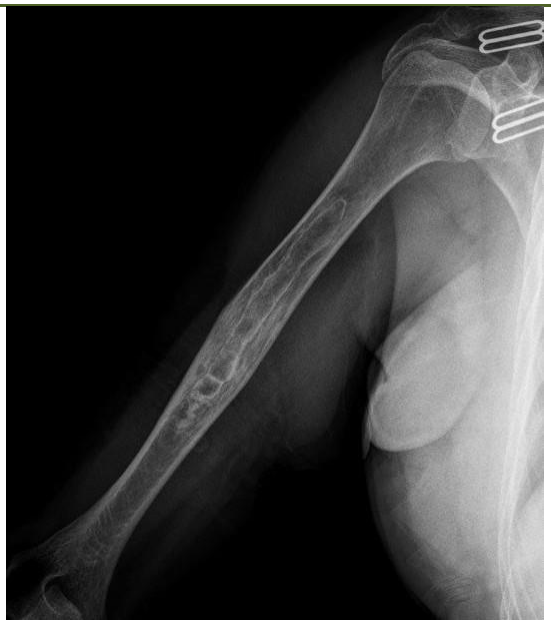

Figure 1: Plain X-Ray of the right humerus showing an oblong intramedullary lytic lesion with sclerotic rim within a ground glass opacity area, responsible of bone enlargement with no cortical rupture or periosteal reaction

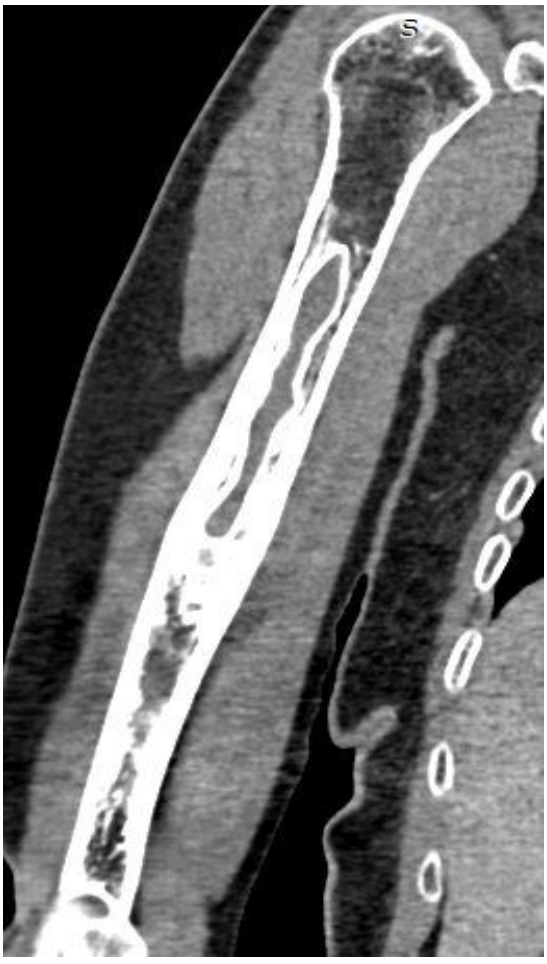

Figure 2: Computed Tomography (CT) frontal view exhibiting a hypodense lytic intramedullary lesion without enhancement suggesting its cystic nature

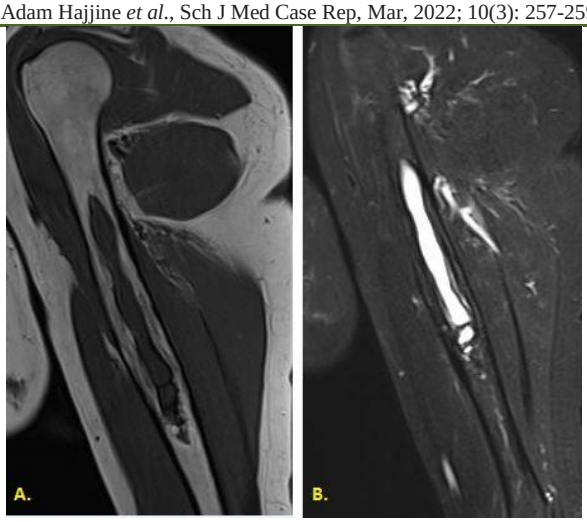

Figure 3: MRI frontal view confirming cystic nature of the lesion as it was hypointense T1 (A) and hyperintense on STIR (B). No extension to surrounding soft tissue was noted

DISCUSSION

Fibrous dysplasia is a developmental disease that can affect any bone, but a predilection for ribs, craniofacial and lower limb bones is described in the literature. Only few cases concerning the humerus have been reported [4].

FD is frequently localized to a single bone, it's the monostotic form which is often asymptomatic with an incidental imaging finding. While in the polyostotic form with involvement of several bones, usually located on the same side, the diagnosis is made in the first decade of life as the lesions can be responsible of pain, deformities and fractures. FD can also present as McCune Albright syndrome when it's associated to skin pigmentation and endocrine disorders or Mazabraud syndrome when intramuscular myxomas are present. Typically syndromic patients have polyostotic forms [3, 5]. In our case, the chief complaint was an isolated chronic pain and the imaging concluded to the diagnosis of monostotic fibrous dysplasia.

There is a wide spectrum of radiologic features of fibrous dysplasia, but the lesions can be classified into three primary lesions: cystic, sclerotic and mixed. The size of lesions can range from small to large affecting all the bone length. On radiographs, the epicenter of the lesion is medullary, a characteristic that signs marrow replacement by fibro-osseous tissue. However, the lesion may be eccentric within the affected bone but it is not cortical. Classically, FD lesion presents as an area of radiolucent ground glass matrix causing a cortical thinning with a smooth outer cortical contour. The margins could be sclerotic which is known as a rind sign. Other features include: dense and sclerotic lesions, delicate fine trabeculae or calcifications within the lesion. The periosteal reaction is not usually present unless it is associated with a pathological fracture [3, 6].

CT is superior to plain radiographs in: better characterization of fibrous dysplasia such as the margins, internal structure, and effect on surrounding structures; evaluation of craniofacial locations; and detection of sarcomatous transformation [6, 8].

MRI is not particularly useful in diagnosis of FD since signal intensity and contrast enhancement are very variable depending on the amount and degree of bony trabeculae, cellularity, collagen, and cystic and haemorrhagic changes. Typically, FD lesions show well-defined borders with intermediate to hypoinensity on T1-WI and intermediate to hyper-intensity on T2-WI . The higher the number of bony trabeculae, the lower the T2 signals, and vice versa. When cystic areas are present, the T2 signal is brighter. Some degree of enhancement on post-contrast T1-WI is usually seen. The main use of this technique remains characterization of extension of the lesion into the surrounding soft tissues and detection of malignant changes [3, 7].

The radionuclide bone scintigraphy is useful to demonstrate the extent of disease, to tell apart monostotic forms from polyostotic ones and to detect signs of malignancy [2].

FD exhibits a dynamic pathologic nature characterized by age-related histological, radiologic, and clinical transformations, therefore, a radiologic monitoring is recommended [9].

CONCLUSION

Fibrous dysplasia is a rare developmental disease that could be revealed by common symptoms such as pain and bone deformity. Therefore, the radiologist plays a crucial role in diagnosis assessment.

Radiologic monitoring is also useful for detection of complications especially malignant transformation.

Conflict of Interest: The authors declare no conflict of interest.

REFERENCES

1. Anitha, N., Sankari, S. L., Malathi, L., & Karthick, R. (2015). Fibrous dysplasia-recent concepts. *Journal of pharmacy & bioallied sciences*, *7*(Suppl 1), S171- S172.
2. Couturier, A., Aumaître, O., Mom, T., Gilain, L., & André, M. (2016). Craniofacial fibrous dysplasia. *La Revue de Medecine Interne*, *37*(12), 834-839.
3. Kushchayeva, Y. S., Kushchayev, S. V., Glushko, T. Y., Tella, S. H., Teytelboym, O. M., Collins, M. T., & Boyce, A. M. (2018). Fibrous dysplasia for radiologists: beyond ground glass bone matrix. *Insights into imaging*, *9*(6), 1035-1056.
4. Kelle, B., Kelle, A. P., Erdoğan, K. E., & Kozanoğlu, E. (2016). A rare cause of shoulder pain: monostotic fibrous dysplasia. *Archives of Rheumatology*, *31*(2), 184-187.
5. Fitzpatrick, K. A., Taljanovic, M. S., Speer, D. P., Graham, A. R., Jacobson, J. A., Barnes, G. R., & Hunter, T. B. (2004). Imaging findings of fibrous dysplasia with histopathologic and intraoperative correlation. *American Journal of Roentgenology*, *182*(6), 1389-1398.
6. Kransdorf, M. J., Moser Jr, R. P., & Gilkey, F. W. (1990). Fibrous dysplasia. *Radiographics*, *10*(3), 519-537.
7. Kinnunen, A. R., Sironen, R., & Sipola, P. (2020). Magnetic resonance imaging characteristics in patients with histopathologically proven fibrous dysplasia—a systematic review. *Skeletal Radiology*, *49*(6), 837-845.
8. Sontakke, S. A., Karjodkar, F. R., & Umarji, H. R. (2011). Computed tomographic features of fibrous dysplasia of maxillofacial region. *Imaging science in dentistry*, *41*(1), 23-28.
9. Hwang, D., Jeon, J., Hong, S. H., Yoo, H. J., Choi, J. Y., & Chae, H. D. (2020). Radiographic follow-up of fibrous dysplasia in 138 patients. *American Journal of Roentgenology*, *215*(6), 1430-1435.