ISSN- O: 2458 - 868X, ISSN–P: 2458 – 8687
Index Copernicus Value: 68 . 16
PubMed - National Library of Medicine - ID: 101731606

International Journal of Medical Science and Innovative Research (IJMSIR)
IJMSIR : A Medical Publication Hub
Available Online at: www.ijmsir.com
Volume – 5, Issue –4, August - 2020, Page No. : 85 - 88

Rare Case of Osteo-Fibrous Dysplasia Arising In Humerus – A Case Report

[1]Dr. Faizan Akhtar, Pathology Resident, Dept. of Pathology, NKP Salve Institute of Medical Science and Research center, Digdoh Hills, Nagpur 440019, Maharashtra

[2]Dr. Sabiha Maimoon, Professor Dept. of Pathology, NKP Salve Institute of Medical Science and Research center, Digdoh Hills, Nagpur 440019, Maharashtra

Corresponding Author: Dr. Faizan Akhtar, Pathology Resident, Dept. of Pathology, NKP Salve Institute of Medical Science and Research center, Digdoh Hills, Nagpur 440019, Maharashtra.

Citation this Article: Dr. Faizan Akhtar, Dr. Sabiha Maimoon, "Rare Case of Osteo-Fibrous Dysplasia Arising In Humerus – A Case Report", IJMSIR- August - 2020, Vol – 5, Issue - 4, P. No. 85 – 88.

Type of Publication: Case Report

Conflicts of Interest: Nil

Abstract

Fibrous dysplasia is a benign condition of bone where the bone is replaced by fibrous tissue, clinically resulting in a bone that is weak and prone to fractures, deformity, or any functional impairment. It is found to be associated with McCune-Albright syndrome. Literature has defined a variant of fibrous dysplasia called Osteo-fibrous dysplasia (OFD). OFD is classically described to occur almost exclusively in the tibial bone. This study reported a case of OFD arising in a humerus. A 15-year-old male child suffered from a fracture of the right humerus while playing cricket. He came to the Orthopaedic OPD at NKPSIMS and RC, Nagpur. A biopsy was taken and sent to the Pathology Department.

Keywords: Osteofibrous dysplasia, Humerus.

Introduction

Osteofibrous dysplasia (OFD) is a rare fibro-osseous lesion of the long bones of unknown pathogenesis described first as a separate clinical entity by Campanacci in 19761.

Clinically the condition presents with painless enlargement of the bone with anterior or anterolateral bowing.

Histologically, osteofibrous dysplasia is characterized by woven bone trabeculae with a rimming of osteoblasts and a cellular proliferation of fibroblast-like cells and has long been thought to be related to adamantinoma of long bones.2–4

Fibrous dysplasia sometimes resembles osteofibrous dysplasia histologically. Osteofibrous dysplasia has been considered to be a congenital lesion or a variant of fibrous dysplasia.5

Surgical treatment for OFD is controversial due to the rarity of the condition and variation in its progression. Osteo-fibrous dysplasia (OFD). OFD is classically described to occur almost exclusively in the tibial bone but rare cases have been reported to occur in the Humerus. We report such a case of OFD arising in the Humerus.

Case Report

A 15-year-old male child suffered from a fracture of the right Humerus while playing cricket. He came to the Orthopaedic OPD at NKPSIMS and RC, Nagpur.

The patient complained of excruciating pain at the right arm which was relieved on immobilization and medication.

He was diagnosed with fracture shaft humerus on plain X-ray.

On Magnetic Resonance Imaging, Bicortical displaced fracture of shaft of the humerus is noted.

An altered signal intensity area is noted in the proximal fractured fragment approximately 5.2 cm away from the metaphysis and in the proximal part of the distal fragment of shaft of humerus appearing isointense on T1WI and hyperintense on T2WI and STIR. No evidence of periosteal reaction noted.

Altered signal intensity is noted along the brachialis, brachioradialis, biceps brachi, and triceps in the subcutaneous plane, suggestive of muscle edema with adjacent free fluid.

A closed reduction and internal fixation (CRIF) with TEN (titanium elastic nail) procedure was performed and a biopsy was taken from the fracture site and sent for histopathological examination.

Histopathological examination

On histopathological examination, an irregular tissue fragment is seen showing many trabeculae composed of woven (immature) bone, at most places, lined by rim of osteoblastic cells.

The stroma is composed of fibrocollagenous tissue showing presence of many spindle cells and mononuclear inflammatory infiltrate.

Considering clinical, radiological and histopathological features, a diagnosis of Osteofibrous dysplasia was given.

Discussion

OFD is a benign fibro-osseous lesion which most commonly occurs in the anterior cortex of the proximal or middle third of the tibia. The occurrence of OFD in other bones is extremely rare.6

Unusual sites like humerus have also been reported albeit very rarely.

OFD is most commonly detected during the first two decades of life6

The histopathology of OFD includes fibro-osseous lesions, with irregular fragments of woven bone rimmed by well-defined osteoblasts.1,6. Zonal architectures, which are characterized by delineation with thin and woven bone or fibrous tissue predominantly in the centre of the lesion with more abundant anastomoses and lamellar bone peripherally, are also observed.1,5. The fibrous tissue also has a whorled, storiform pattern.1,5

In our case, an irregular tissue fragment is seen showing many trabeculae composed of woven (immature) bone, at most places, lined by rim of osteoblastic cells.

The stroma is composed of fibrocollagenous tissue showing presence of many spindle cells and mononuclear inflammatory infiltrate.

However, zonal architecture and storiform pattern was not evident in our case. But, OFD could be diagnosed in view of other features.

In general, the main differential diagnosis for OFD is FD or adamantinoma.1

FD is a benign fibro-osseous lesion that occurs in adults and children.6 Although it occurs in almost any part of the bones, the craniofacial bone and the femur are the two most common sites.1,6. Its histopathology shows fibro-osseous lesions. The osseous component comprised irregular, curvilinear, trabeculae of woven

bone. The woven bone is not rimmed by osteoblasts, in contrast with OFD.5

Adamantinoma is a low-grade malignant bone tumor which frequently occurs in the tibia in young to middle-aged adults.1 There are two types of adamantinoma, including classical (the more aggressive type) and differentiated (the more benign form). The latter one resembles OFD (OFD-like adamantinoma).1 The relationship between OFD and adamantinoma is important because they are in the same spectrum of diseases, and OFD is known to progress into adamantinoma in some cases.

In our case, FD was ruled out because the tumor was composed of woven bone rimmed by osteoblasts histologically.

Until bone maturity, it is understood that OFD should not be treated surgically because the lesion will heal spontaneously and there is a high risk of recurrence after surgery.

Lee et al reported 16 cases in which recurrence occurred in 6 cases after curettage, and therefore recommended extraperiosteal excision as the surgical treatment for OFD. On the contrary, Ozaki et al reported good results in six cases treated with curettage and bone grafting.

Recently, the safety and effectiveness of anhydrous ethanol for adjuvant therapy after intraregional curettage of bone tumor has been reported.

Conclusion

We reported a rare case of osteofibrous dysplasia (OFD) occurring in humerus of a 15-year-old child. Humerus is a rare site for the presentation of OFD, so it is imperative to keep OFD in mind as a differential diagnosis when there is presence of osteoblastic rimming in woven bone histologically.

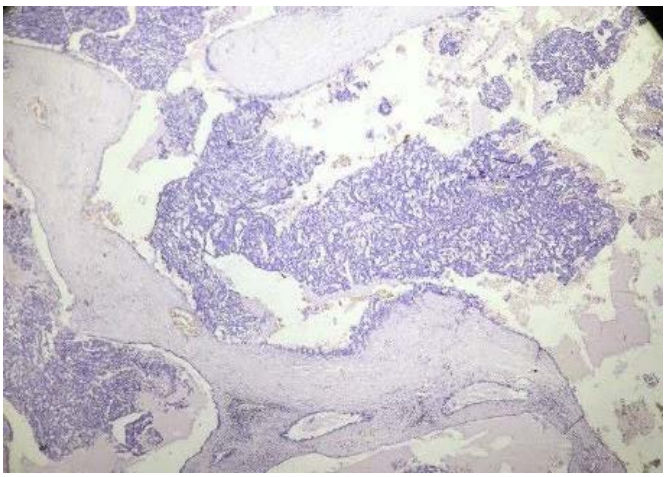

Figure 1: photomicrograph (H&E x 100)showing irregular woven bone lined by osteoblastic cell.

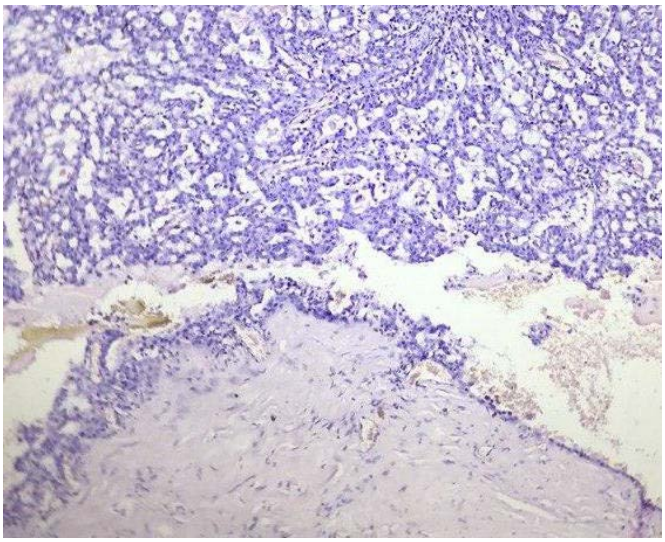

Figure 2: photomicrograph (H&E x 100) showing stroma composed of fibro collagenous tissue showing presence of spindle cell

References

1. Campanacci M. Osteofibrous dysplasia of long bones a new clinical entity. Ital J Orthop Traumatol. 1976 Aug;2(2):221–37.

2. Alguacil-Garcia A, Alonso A, Pettigrew NM. Osteofibrous dysplasia (ossifying fibroma) of the tibia and fibula and adamantinoma. A case report. Am J Clin Pathol. 1984 Oct;82(4):470–4.

3. Keeney GL, Unni KK, Beabout JW, Pritchard DJ. Adamantinoma of long bones. A clinicopathologic study of 85 cases. Cancer. 1989 Aug 1;64(3):730–7.

4. Markel SF. Ossifying fibroma of long bone: its distinction from fibrous dysplasia and its association with adamantinoma of long bone. Am J Clin Pathol. 1978 Jan;69(1):91–7.

5. Park YK, Unni KK, McLeod RA, Pritchard DJ. Osteofibrous dysplasia: clinicopathologic study of 80 cases. Hum Pathol. 1993 Dec;24(12):1339–47.

6. Bridge JA, Hogendoorn P, Fletcher CD, Bridge JA, Hogendoorn PC, Fletcher CDM. WHO Classification of Tumours of Soft Tissue and Bone. 4th ed. Lyon: International Agency for Research on Cancer; 2013.