Volume 20 Issue 3
Originally issued as H1

Global Journal of Medical Research: H
Orthopedic and Musculoskeletal System
Volume 20 Issue 1 Version 1.0 Year 2020
Type: Double Blind Peer Reviewed International Research Journal
Publisher: Global Journals
Online ISSN: 2249-4618 & Print ISSN: 0975-5888

Pathological Proximal Femoral Fracture in Mc-Cune Albright Syndrome: A Case Report

By Apoorv Sehgal, Pratyush Shahi & Aarushi Sudan

University College of Medical Sciences

Abstract- A 25-year-old female present to us in the emergency department with atraumatic fracture of the proximal femur. She had a cafe-au-lait spot on the face and gave history of precocious puberty. A skeletal survey showed polyostotic fibrous dysplasia. A clinico-radiological diagnosis of McCune-Albright syndrome was made which was further confirmed on biopsy. An extra-medullary fixation was done and at 6 months follow-up, the fracture had united and the patient could walk without support.

GJMR-H Classification: *NLMC Code: WE 175*



Strictly as per the compliance and regulations of:

© 2020. Apoorv Sehgal, Pratyush Shahi & Aarushi Sudan. This is a research/review paper, distributed under the terms of the Creative Commons Attribution-Noncommercial 3.0 Unported License http://creativecommons.org/licenses/by-nc/3.0/), permitting all non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

Pathological Proximal Femoral Fracture in Mc-Cune Albright Syndrome: A Case Report

Apoorv Sehgal [α], Pratyush Shahi [σ] & Aarushi Sudan [ρ]

Abstract- **A 25-year-old female present to us in the emergency department with atraumatic fracture of the proximal femur. She had a cafe-au-lait spot on the face and gave history of precocious puberty. A skeletal survey showed polyostotic fibrous dysplasia. A clinico-radiological diagnosis of McCune-Albright syndrome was made which was further confirmed on biopsy. An extra-medullary fixation was done and at 6 months follow-up, the fracture had united and the patient could walk without support.**

I. Introduction

Fibrous dysplasia/McCune-Albright syndrome (FD/MAS) occurs due to somatic gain-of-function mutations of the *GNAS* gene. These mutations cause over activity in the target tissues leading to clinical features that vary in severity and age of onset.[1] Since the disease is rare and can have atypical presentation, the diagnosis can be missed. We report the case of a young female with McCune-Albright syndrome presenting to us with pathological fracture of the proximal femur and highlight the challenges one can face in fixation of such fractures.

II. Case Presentation

A 25-year-old female patient presented to us with a history of sudden onset of pain in her right hip and inability to bear weight over the affected limb. There was no history of trauma, fever or weight loss. On examination, the right lower extremity was externally rotated and shortened. Movements at the right hip elicited pain. The patient was of short stature and had acafé-au-lait skin macule with jagged, irregular borders (coast of Maine) on her face with short stature.*(Figure 1)* On further inquiry, she gave a history of precocious puberty.

Radiographs revealed lytic expansile lesion with ground-glass appearance and thinning of cortex in metaphyseal region of the right proximal femur associated with a pathological subtrochanteric fracture. Other sites like skull, radius and phalanges also had similar lytic lesions.*(Figure 2)*

Based on clinical and radiological features, a diagnosis of McCune-Albright syndrome with pathological right subtrochanteric fracture was made. The patient was put on skin traction for the right lower limb and advised to undergo surgery for the subtrochanteric fracture of right femur in view of the nature of the fracture and the requirement of early ambulation. After proper consent, the patient was taken up for surgery. Fracture was reduced on a traction table after opening the fracture area. Biopsy was taken from the fracture site followed by reduction and fixation with distal femoral locking plate of opposite side in reverse configuration. The initial plan was of internal fixation with cephalomedullary nail, but due to very narrow diameter of the medullary canal, an extramedullary fixation had to be done. *(Figure 3)* The patient was discharged on post-operative day 5 after confirming healthy state of the wound. She was started on partial weight-bearing with crutches at 2 weeks continuing to full weight-bearing at 6 weeks. At 6 months follow-up, the patient could walk without support and with no residual complaints.

Author α σ ρ: University College of Medical Sciences, Delhi, India.
e-mail: apoorvsehgal2006@gmail.com

© 2020 Global Journals



Figure1: Showing the cafe-au-lait spot









© 2020 Global Journals



Figure 2: (A-D) showing lytic ground-glass lesions in the skull, radius, phalanges and proximal femur, (E) showing a pathological subtrochanteric fracture, (F) after fixation

III. Discussion

McCune–Albright syndrome is a rare sporadic disease caused by somatic postzygotic activating mutations in the *GNAS* gene. It is characterised by polyostotic fibrous dysplasia, café-au-lait skin spots and variable hyperfunctional endocrinopathies.[2]

A fibrous dysplasia lesion predisposes a person to have a pathological fracture and if involving a large area can lead to deformity of the bone. Although a small lesion can be left as such, a larger lesion causing pain or threatening to fracture should be curetted and bone grafted even though there is a high tendency for it to recur. If the site and extent of the lesion necessitates, prophylactic fixation should be done. The management of fibrous dysplasia becomes more complicated when it is associated with extra-skeletal manifestations.[1,3]

A lesion of the proximal femur causes more pain, fractures and deformity than any other skeletal localization of the disease.[2-5] A pathological fracture of the proximal femur secondary to fibrous dysplasia poses several intra-operative challenges. A surgeon should do proper pre-operative planning and have backup fixation devices. Also, good post-operative wound care and early rehabilitation should be done.

References Références Referencias

1. Lichtenstein L. Polyostotic fibrous dysplasia. Arch urg. 1938;36:874–898. doi: 10.1001/archsurg.1938.01190230153012.
2. Leet AI, Chebli C, Kushner H, Chen CC, Kelly MH, Brillante BA, Robey PG, Bianco P, Wientroub S, Collins MT. Fracture incidence in polyostotic fibrous dysplasia and the McCune-Albright syndrome. J Bone Miner Res. 2004; 19:571–577. doi: 10.1359/JBMR.0301262
3. Harris WH, Dudley HR, Jr, Barry RJ. The natural history of fibrous dysplasia. An orthopaedic, pathological, and roentgenographic study. J Bone Joint Surg Am. 1962; 44-A:207–233. doi: 10.2106/00004623-196244020-00001
4. Stanton RP, Ippolito E, Springfield D, Lindaman L, Wientroub S, Leet A. The surgical management of fibrous dysplasia of bone. Orphanet J Rare Dis. 2012;7(Suppl 1):S1. doi: 10.1186/1750-1172-7-S1-S1.
5. O'Sullivan M, Zacharin M. Intramedullary rodding and bisphosphonate treatment of polyostotic fibrous dysplasia associated with the McCune-Albright syndrome. J Pediatr Orthop. 2002;22:255–60.

© 2020 Global Journals