ISSN: 2230-9926

Available online at http://www.journalijdr.com

IJDR

International Journal of Development Research
Vol. 08, Issue, 02, pp.18863-18864, February, 2018

CASE STUDY OPEN ACCESS

ONCOGENIC OSTEOMALACIA ASSOCIATED WITH PHOSPHATURIC MESENCHYMAL TUMOR OF THE THIGH: A CASE REPORT

***Dr. Shravan Shetty, Dr. Sagar Sharma, Dr. Deepak Parikh, Dr. Yash Devckar and Dr. Ankur Pareek**

Department of Surgical Oncology, Somaiya Ayurvihar Asian Cancer Institute, Mumbai

ARTICLE INFO

Article History:
Received 20th November, 2017
Received in revised form
23rd December, 2017
Accepted 13th January, 2018
Published online 28th February, 2018

Key Words:
Oncogenic Osteomalacia,
Phosphaturic Mesenchymal Tumor,
Hypophosphatemia,
Fibroblast Growth Factor 23(FGF 23).

ABSTRACT

Oncogenic Osteomalacia (OOM) is an uncommon metabolic and bone disease caused by Fibroblast Growth Factor 23 (FGF 23),a phosphaturic factor produced by phosphaturic mesenchymal tumors characterized by phosphate leakage from the kidneys and subsequent hypophosphatemia. We present the case of oncogenic osteomalacia in a 50 year old woman who presented to our hospital with complaint of being unable to walk since 5 years. She had been initially diagnosed with low phosphorus level and treated with oral phosphate powder for 3 years. After a complete diagnostic evaluation, she was diagnosed with a phosphaturic mesenchymal tumor of the thigh. The patient underwent surgery. Post surgery, the patient had complete recovery.

Citation: Dr. Shravan Shetty, Dr. Sagar Sharma, Dr. Deepak Parikh, Dr. Yash Devckar and Dr. Ankur Pareek. 2018. Oncogenic Osteomalacia Associated with Phosphaturic Mesenchymal Tumor of the Thigh: A Case Report, *International Journal of Development Research*, 8, (02), 18863-18864

INTRODUCTION

Oncogenic Osteomalacia(OOM) is an acquired disorder characterized by marked mineral derange and adjustment of skeletal metabolism (Econs *et al*., 1994; Folpe, 2004). Its clinical expressions are hypophosphatemia, osteomalacia,bone distress, proximal muscle weakness,pathological fractures leading to functional disability.OOM is commonly associated with small and slow growing mesenchymal tumors. Both biochemical signs and bone disease can be completely solved by complete removal of the tumor.

Case Report

A 50 year old woman presented to our hospital with the complaints of unable to walk since 5 years.She had initially been diagnosed with low phosphorus level and was being treated with oral phosphate powder for 3 years. Bone scan showed abnormal uptake in both shoulders,bilateral elbows, bilateral wrists, bilateral hips, bilateral knee joints, right ankle joint, left femoral shaft and bilateral ribs. These were all suggestive of an extensive large joint arthropathy with adjacent enthesopathy. On examination she had hypophosphatemia (1.9gm/dl) with normal serum calcium & elevated PTH (163pg/ml). FDG PET-CT showed a low grade metabolically active soft tissue lesion in proximal right thigh, just lateral to the femoral vessels which needs biopsy evaluation.Trucut biopsy was consistent with phosphaturic mesenchymal tumor.MRI scans revealed a well defined lobulated heterogeneous enhancing mass lesion in the anterior aspect of the right thigh.Ultrasound neck was normal.She underwent complete wide excision of the tumor in the right thigh.The final histopathology report was phosphaturic mesenchymal tumor.One month after surgery the patient was completely asymptomatic and her phosphate was normal (3.5mg/dl)

DISCUSSION

Phosphaturic mesenchymal tumors have been well described histologically and the entity of oncogenic osteomalacia is a known,albeit uncommonly encountered cause of osteomalacia.

Corresponding author: **Dr. Shravan Shetty,**
Department of Surgical Oncology, Somaiya Ayurvihar Asian Cancer Institute, Mumbai.

There is often a delay in the diagnosis and definitive treatment of patients with a phosphaturic mesenchymal tumor due to the varying clinical presentation and the typically cryptic nature of symptoms. Phosphaturic mesenchymal tumors produce FGF23 causing systemic phosphate depletion and leading to oncogenic osteomalacia. Sites of occurrence are the soft tissue and bone,almost half of the cases in the lower extremities. A long history of osteomalacia,bone pain and fractures is usually present. Renal phosphate wasting and elevated serum levels of FGF23 are found. Radiologically, the tumor may present a varied appearance. Histologically, 3 variants are seen: osteoblastoma- like, non-ossifying fibroma-like and mixed connective tissue variant,the commonest.This is characterized by a hemangiopericytoma-like tissue pattern and areas of matrix with peculiar calcifications. The optimal treatment is the complete surgical removal of the tumor which results in a quick & complete recovery from symptoms,normalization of biochemical alterations and re-mineralization of the bone. These tumors may have different appearance which may be due to their misdiagnosis with various other mesenchymal tumors.These type of tumors are difficult to be detected because often of small dimensions and localized elsewhere in the body (Suryawanshi, 2011).

Preoperative localisation is essential and could be reached by many imaging modalities such as CT scans or MRI, or somatostatin receptor scintigraphy or PETCT (Fukumoto, 1999; Dupond *et al*., 2005; Hesse *et al*., 2007). From a biochemical point of view, OOM is characterised by hypophosphatemia, elevation of alkaline phosphatase, low circulating 1,25 $(OH)_2D3$ and increased concentration of FGF23. In this paper we describe the case of a patient affected with OOM originating from a right thigh tumor in which we obtained complete clinical recovery after tumor removal;this result confirms the ones described in literature.

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