

Kahler's Disease Revealed by a Fracture of the Humeral Diaphysis

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Abstract

Kahler's disease or multiple myeloma is a malignant hematological pathology, characterized by the malignant proliferation of plasma cells. Reporting a case of a humeral shaft fracture in a non-traumatic context that revealed Kahler's disease during paraclinical investigations. Management involves histological confirmation of the pathology, osteosynthesis of the fracture by static centromedullary nailing, with medical treatment made of biphosphonate combined with a chemotherapy protocol targeting the disease. The management of a pathological fracture is a diagnostic emergency, but never an operating emergency. A diagnosis of certainty must be made before any intervention.

Keywords: Kahler Disease; Pathological Fracture; Humeral Diaphysis; Centromedullary Nail; Chemotherapy

Introduction

Kahler's disease or multiple myeloma is a malignant hematological pathology, characterized by the malignant proliferation of plasma cells. The most common symptomatology is osteolytic bone damage, severe bone pain, hypercalcemia, kidney damage and anemia. The occurrence of pathological fractures in the humeral diaphysis as the first clinical sign of multiple myeloma is rare.

We reported a case of a fractured humeral shaft that revealed Kahler's disease during paraclinical investigations.

Materials and Methods

This is a 73-year-old male with a history of high blood pressure under treatment, who had consulted the emergency department for acute left arm pain with total functional impotence of the left upper limb without any notion of trauma. The clinical examination on admission found a conscious and hemodynamically and respiratory stable patient with a swollen, deformed and painful left arm on palpation, the vascular and nervous examination of the left upper limb was normal. An X-ray of the arm taking the shoulder and elbow from the front and profile was performed, which showed the presence of a fracture of the upper third of the left humeral diaphysis on an osteolysis image with fuzzy limits without invasion of the soft tissues (Figure 1).

Research Article

Volume 3 Issue 3 Received Date: April 05, 2019 Published Date: April 26, 2019 DOI: 10.23880/jobd-16000181

Journal of Orthopedics & Bone Disorders

An X-ray of the skull showed incomplete "cookie cutter" lesions (Figure 2). Without obvious lesions on a pelvic X-ray, a complement by an MRI of the left arm confirmed a fracture on a tumour process of the humeral diaphysis (Figure 3). The non-traumatic mechanism of the fracture as well as radiographic images led us to diagnose an unstable diaphyseal fracture on pathological bone. Serum protein electrophoresis showed the presence of a monoclonal peak migrating into the beta-2 globulin zone with a monoclonal band of IgA lambda isotypy (Figure 4). Kahler disease was confirmed on surgical biopsy. The patient was treated surgically with intramedullary osteosynthesis by locking nail with immobilization by elbow sling to the body for 03 weeks followed by functional rehabilitation (Figure 5). Bisphosphonate treatment has been initiated.



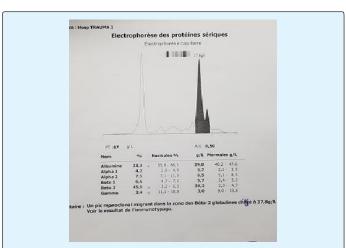
Figure 1: X-ray of the left humerus showing the fracture and associated bone lacunar images.



Figure 2: Skull x-ray: incomplete "cookie cutter" lesions characteristic of multiple myeloma.



Figure 3: MRI of the left arm shows a fracture on a tumor process.



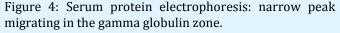




Figure 5: X-ray of the left humerus from the front showing good bone stabilization with a static nail.

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Results

The patient was referred to the oncologist for an induction chemotherapy protocol (bortezomib, thalidomide and dexamethasone). After a 6-month retreat, the patient was seen again in consultation in good general condition, the fracture is consolidated with a good functional recovery of his left upper limb. The patient's autonomy was decisive for the possibility of performing an auto graft of the bone marrow after induction chemotherapy and therefore for the control of his malignant hematopathy.

Discussion

Myeloma is a disease characterized by the development in the skeleton of multiple osteolytic plasma cell tumours (plasmocytomas) secreting a single monoclonal immunoglobulin in 80% of cases, either type G (twothirds of cases) or type A (one-third of cases). It is considered to be the most common malignant hematopathy after lymphoma, accounting for about 10% of blood cancers [1]. Bone involvement secondary to bone marrow infiltration is the most common manifestation and results in osteolytic lesions, bone pain and pathological fractures.

Fractures caused by myeloma can be managed in the same way as those caused by carcinoma, and their biological behaviour and mechanical implications are similar to metastatic fractures [2].

Bone metastases affecting the proximal humerus can be treated by arthroplasty resection or curettage of the pathological lesion, polymethyl methacrylate cement (PMMA) and plate osteosynthesis [3]. Prosthetic replacement provides excellent analgesic control, but often sacrifices part of the shoulder function [4]. This option will be indicated for patients with a sufficiently long vital prognosis, especially in the case of single metastasis. Stabilization of the pathologic fracture by PMMA-reinforced locked plate provides adequate local tumor control, good mechanical stabilization and satisfactory preservation of function [5,6]. Centromedullary nailing makes it possible to treat the majority of diaphyseal metastases, possibly reinforced by PMMA. It offers a good analgesic effect, allows rapid mobilization and with fewer complications [7-12]. This is the case for our patient but without PMMA. Bisphosphonate supplement therapy is essential to prevent and treat bone damage during kahler disease, whether oral clodronic acid or intravenous zoledronic or

pamidronic acid [8,9]. The American Society for Clinical Oncology set the terms of this treatment in 2007, with a preference for intravenous bisphosphonates and minimum treatment duration of 2 years to be reconsidered based on the hematological response to auto-transplantation [10].

The anatomical and functional results in our patient after a six-month retreat, obtained by a stable (locked centromedullary nail), effective and durable osteosynthesis, resulted in a return to autonomy [11]. Katagiri prognostic survival score for patients with bone metastases is the most widely used in the literature. In our context a survival rate is estimated at 98% at 06 months and 75% at 24 months [13,14].

Conclusion

Kahler's disease is a plasmocytic, malignant, clonal proliferation. Tumor plasma cells stimulate bone resorption responsible for bone gaps with the risk of pathological fracture. The management of a pathological fracture is a diagnostic emergency, but never an operating emergency. A diagnosis of certainty must be made before any intervention. The management of these fractures requires fixation by stable osteosynthesis, which is effective with fewer complications and allows the patient to regain his autonomy combined with an appropriate chemotherapy protocol

Conflict of Interest

The authors have no conflict of interest to declare

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Dehayni B, et al. Kahler's Disease Revealed by a Fracture of the Humeral Diaphysis. J Ortho Bone Disord 2019, 3(3): 000181.

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