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Case Report

Presentation and management of bilateral fracture shaft humerus in a multiple myeloma patient: An extremely rare case report and review of literature

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ABSTRACT

Multiple myeloma (MM) is a malignancy of plasma cells and the second most common hematologic malignancy (13%); these cells accumulate in bone marrow and overproduce a monoclonal protein. The bone disease develops in 80–90% of patients with MM includes bone pain, pathologic fractures (40%), spinal cord compression (5%), and hypercalcemia. Pathological fractures can occur in extremities, but bilateral involvement is extremely rare. We report a case of 65-year-old male presented with a chief complaint of spontaneous bilateral humerus shaft fractures. On subsequent workup, MM was confirmed. The patient was managed with internal fixation bilaterally and showed favorable union at 1-year follow-up.

Key words: *Bilateral pathological fracture, Humerus, Multiple myeloma*

Multiple myeloma (MM) is a malignancy of plasma cells; these cells accumulate in bone marrow and overproduce a monoclonal protein. The bone disease develops in 80%–90% of patients with myeloma and includes bone pain, pathologic fractures (PFs) (40%), spinal cord compression (5%), and hypercalcemia. These skeletal-related events compromise mobility and independence, adversely affect the quality of life and are associated with decreased survival. Other major clinical manifestations of MM are anemia, hypercalcemia, renal failure, and an increased risk of infections [1-3]. MM is highly treatable malignancy mainly presents in the axial skeleton. Unlike other malignancies that metastasize to bone, the osteolytic bone lesions in MM exhibit no new bone formation [4]. PFs can occur in extremities, but bilateral involvement is extremely rare [5].

We present a case of unusual, spontaneous bilateral humerus shaft fractures in a MM patient, managed surgically and followed up to 1 year. No similar case was found on the literature search.

CASE REPORT

A 65-year-old man presented to the department of orthopaedic with a chief complaint of spontaneous-onset pain in the right arm and inability to use the same upper limbs for 3 days. The patient also complained of back pain for 3–4 months. There was no history of trivial or other traumas and pain in the left arm. He was on regular medication for hypertension and chronic obstructive pulmonary disease. Informed consent was obtained from the patient for taking the photographs. On examination, the patient had severe pallor; the midarm region was swollen and tender. Tenderness was also noted in the spine and pelvic bones. Other systemic examinations were unremarkable.

Radiograph of arm revealed ill-defined osteolytic lesions in the acromion, clavicle, scapulae, and shaft and proximal humerus bone with fractures (Fig. 1). A chest radiograph showed multiple ill-defined osteolytic lesions in multiple ribs, with fibrocalcific haze in the right upper and middle zone (Fig. 2). A differential diagnosis of metastasis or MM was considered on initial screening. The upper limb was splinted. A further skeletal survey of skull, spine, and pelvis revealed lytic areas in the skull, proximal femur, pelvis, and spine, with a collapse of the 8th and 12th thoracic vertebra (Figs. 3 and 4). After 5 days of admission, while on workup, the patient complained mild pain in the left arm and inability to use the same upper limb. There was no history of trivial or any other traumas X-ray showed ill-defined osteolytic lesions in the shaft and proximal humerus with fracture (Fig. 5), the left upper limb was also splinted.

Laboratory tests showed hemoglobin 75 g/L, total leukocyte count 5.7×10^9/L (79% granulocytes, 18% lymphocytes, 1% monocytes, and 2% eosinophils), erythrocyte sedimentation rate 54 mm/1st h, C-reactive protein 12.8 mg/L, β2 microglobulin 64.23 mg/L, serum urea 29.4 mmol/L, serum creatinine 1.0 mg%, serum calcium 9.2 mg%/dl and serum alkaline phosphatase 142 U/L, total protein 12.20 g/dl (normal 6.40–8.10 g/dl), serum albumin 3.55 g/dl (normal 3.50–5.64 g/dl), and serum globulin 6.60 g/dl (normal 0.62–1.53 g/dl). Serum protein electrophoresis revealed an M band pattern. Urinalysis was negative for Bence Jones proteins. Ultrasound whole abdomen revealed bilateral basal pleural effusion with a trace of ascites in hepatorenal angle. Contrast-enhanced computed tomography chest and abdomen

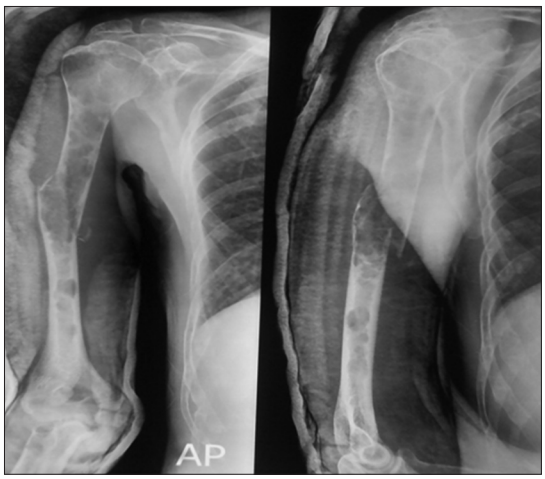

Figure 1: Ill-defined osteolytic lesions in acromion, clavicle, scapulae, shaft, and proximal humerus with fracture

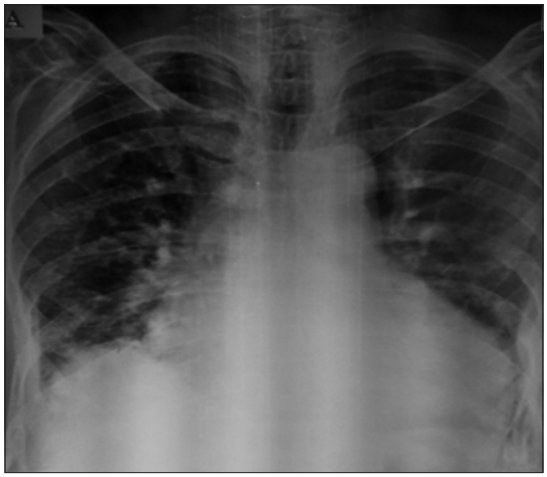

Figure 2: Multiple ill-defined osteolytic lesions in multiple ribs, with fibrocalcific haze in the right upper and middle zone

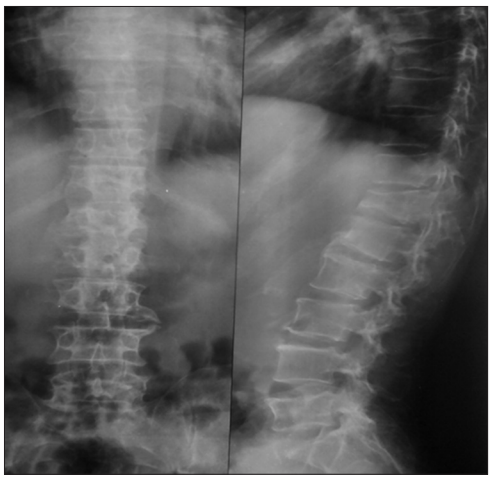

Figure 3: Lytic areas in the spine with collapse of the 8th and 12th thoracic vertebra

Figure 4: Lytic lesions in the pelvis and proximal femur

showed osteolytic lesions with a cortical break in most of the visualized bones with compression fracture DV8, DV12, and LV2 bodies. Workup for the primary was negative. Examination of a bone marrow aspirate showed moderately increased hypercellular marrow with increased plasma cells. A diagnosis of Stage IIIB myeloma was made according to the international staging system. The patient was given 5 units of whole blood to improve the hemoglobin level.

A surgical procedure was performed under scalene block anesthesia. In beach-chair position, a deltoid-splitting incision was used at the anterior aspect of the acromioclavicular joint and was extended about 2.5 cm. The site of entry for the nail into the humerus was in the sulcus between the greater tuberosity and the articular surface of the humeral head, directly in line with the medullary canal. Then, the supraspinatus tendon was sharply incised parallel to its fibers to expose the entry site. The proximal entry site was perforated, and guidewire was passed down into the canal. The appropriate size nail was driven with small rotary movements over the guidewire without reaming the medullary canal. Proximal interlocking was performed with the assistance of a nail-mounted drill guide. Two interlocking screws were used to enhance the stability of the fixation. Distal interlocking was performed by the free-hand technique by carrying out a limited dissection through a 5-cm incision to protect the radial nerve. After through wash, wound was closed with skin staples. Same technique was followed for the left humerus fracture. Post-operative period was uneventful.

The patient was referred to an oncology center for chemotherapy after 1 week when he was stabilized and able to use his arms during daily activities with advice of gentle range of motion of both shoulder and elbow joint. Lesions at other sites including spine and proximal femur were managed conservatively. A chemotherapy regimen of thalidomide and dexamethasone was started. The patient was followed up for 3 months in the outpatient department up to 1 year. At final follow-up, laboratory parameters were measured as C-reactive protein 18.0 mg/L, β2 microglobulin 65.19 mg/L, and serum albumin 3.2 g/dl. There was no limitation of range of motion at the elbow joint. The abduction of the right and left shoulder was 0–80° and 0–110°, respectively. The forward flexion of the right and left shoulder was 0–90° and 0–120°, respectively. An inappropriate length of nail left outside at the proximal entry site was responsible for limited abduction

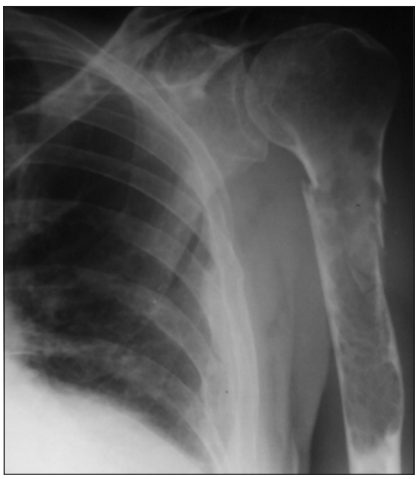

Figure 5: Ill-defined osteolytic lesions in acromion, clavicle, scapulae, shaft, and proximal humerus with fracture

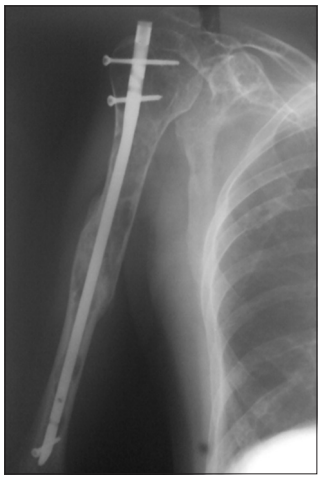

Figure 6: Bony union at 1 year with internal fixation *in situ* right

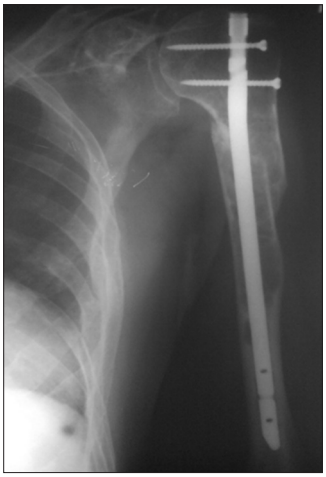

Figure 7: Bony union at 1 year with internal fixation *in situ* left

in the right shoulder. Humerus fracture showed satisfactory union on both sides (Figs. 6 and 7).

DISCUSSION

MM is the second most common hematologic malignancy (13%) and accounts for 1% of all cancers. The diagnosis of MM is incidental in 30% of cases [6]. PFs have been associated with 23–32% increased risk of mortality in prostate cancer, breast cancer, and MM [7]. The diagnosis of MM requires ≥10% clonal bone marrow plasma cells or a biopsy-proven plasmacytoma plus evidence of one or more MM defining events consists of CRAB (hypercalcemia, renal failure, anemia, or lytic bone lesions) features felt related to the plasma cell disorder, bone marrow clonal plasmacytosis ≥60%, serum involved/uninvolved free light chain (FLC) ratio ≥100 (provided involved FLC is ≥100 mg/L), or >1 focal lesion on magnetic resonance imaging [8]. Bilateral spontaneous fracture is extremely rare. Only three cases have been reported so far on literature search Table 1 [6,9,10]. Similarly, bilateral spontaneous fracture of shaft humerus in MM patient was unusual in our report. In the recently published meta-analysis, it was concluded that PFs correlate with reduced survival in patients with malignant bone disease [11] supported by other similar studies. Our initial workup was in favor of metastasis with primary in lungs. In our case, chest radiograph showed fibrocalcific haze, normal range laboratory parameters, and urinalysis was negative for Bence Jones proteins. However, diagnostic certainty came from serum electrophoresis, bone marrow examination. Collins found Bence Jones proteins in about 49% of cases [12]. In 83% of MM cases, serum electrophoresis produces a single band called a monoclonal spike or M band [13]. We have managed our case with internal fixation with close unreamed interlocking nailing on both sides. The closed interlocking humeral nailing is a simple and quick procedure and is associated with minimal morbidity and low failure in PFs [14]. In similar published studies, there is a lack of internal fixation as a management of PFs associated with MM. MM has shown improved long-term survival rates due to recent advances in treatment options. Survival with conventional therapy ranges from 1 to 10 years. The chemotherapy regimens generally used are melphalan and prednisone or vincristine, doxorubicin and dexamethasone [15]. Autologous stem cell therapy achieves higher remission in young patients with low β2 microglobulin levels. Bisphosphonates are also useful in cases of hypercalcemia and for the prevention of skeletal-related events [13]. We did not use bisphosphates and stem cell therapy in our case as calcium level was in normal range. The presented case showed favorable bony union

Table 1: Review of literature

Study	Age & Gender	Treatment	Chemotherapy	Follow up & Result
Alexa *et al*[6]	70, Male	Conservative	Not mentioned	Not mentioned
V Gupta *et al*[9]	52, Male	Conservative	Not mentioned	Not mentioned
Singh *et al*[10]	49, Male	Conservative	Vincristine, doxorubicin dexamethasone bis	1 week, died due to severe pulmonary infection
Present study	65, Male	Close Interlocking nailing	Thalidomide dexamethasone	1 year

at 1-year follow-up. In similar studies, they are not mentioning about follow-up. Gainor and Buchert [16] reported a union rate of 67% of fractures in PFs associated with MM. However, further studies are needed to determine healing of PFs in such cases.

CONCLUSION

Pathological fractures of long bones associated with MM can be satisfactorily treated with internal fixation with adequate adjuvant therapy.

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