

A SMOLDERING / INDOLENT MYELOMA WITH EXTENSIVE ABDOMINAL PRESENTATION- CASE REPORT.

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ABSTRACT

A case of indolent/smoldering myeloma in a 70-year-old man is reported. He presented with an unusual multiple symptomatic myeloma with extramedullary impairment and absence of bone pain. He was treated with pulses of high-dose dexamethasone with commendable clinical improvement.

Key words: Myeloma, Dexamethasone

INTRODUCTION

Plasma cell dyscrasias are a group of disorder that arise from clonal proliferation of neoplastic plasma cells or associated B cells. Typically clinical manifestation varies as a result of the heterogenous biology and span the entire spectrum from the early phase of indolent/smoldering myeloma (SM) to highly aggressive overt multiple myeloma (MM).¹ Two subsets of SM have been identified namely evolving SM and non-evolving SM with stable M- paraprotein that abruptly increases when symptomatic MM develops. SM is characterized by M-protein ≥ 30 /dl and /or bone marrow clonal cells $\geq 10\%$ but no related organ or tissue impairment (ROT1)(end-organ damage), which is typically manifested by increased calcium, renal insufficiency, anaemia, or bone lesions (CRAB) attributed to the plasma cell proliferative process.² A proportion of the patients have to be followed up indefinitely for appropriate treatment to avoid complications.³ This however is often difficult in a developing country like ours where culture attitude and follow up is a problem. This paper reports the first case in 10 years in our institution of unusual pattern of a myeloma case that was successfully treated with high-dose of dexamethasone.

Case Report

A 70 year-old farmer was referred to the haematology clinic July 2004 with two-year history of increasing abdominal swelling, pain on the left side of the abdomen with dragging sensation and easy satiety. He also complained of tiredness/weakness, sudden episodes of fainting attacks and weight loss. Two months prior to presentation, he gave a 2 week history of haematuria for which he was treated in a private clinic. There was no history of bone pains, fever, night sweats or bleeding diathesis. Physical examination revealed a chronically ill-looking elderly man. He was conscious and alert, He

had herniorrhaphy several years ago otherwise his personal and family history was unremarkable. severely pale, anicteric with generalizes lymphadenopathy (1cm). There was massive splenomegaly (24cm midclavular line), hepatomegaly (6cm) and moderate ascites. We suspected differential cause of massive splenomegaly. Laboratory evaluation revealed the following results: haemoglobin (Hb) 10g/dl, white blood cell count (WBC) $2.6 \times 10^9/l$ with differentials of 35% neutrophils, 65% lymphocytes, platelet count $90 \times 10^9/l$ and ESR 130 mm/hr. Coombs direct and indirect antiglobulin test and retroviral tests were negative. The peripheral blood film showed hypochromia, microcytosis, target and pencil shaped cells with no rouleaux formation. There was normal morphology but apparent reduction in granulocytes and platelets. A Perl's stain done was negative. Bone marrow cytology revealed a hypercellular marrow and few megaloblasts. Plasmacytosis (12%) evenly dispersed throughout the marrow was seen. Some of these plasma cells were binucleated with inconspicuous nucleoli. A few others showed accumulation of prominent cytoplasmic vacuoles. Bone marrow histology confirmed mild plasmacytosis. A comprehensive myeloma work-up was initiated. Total skeletal survey with chest thoracolumbar, pelvic x-ray were all normal. There was a slight reduction of normal circulating immunoglobulin on serum protein electrophoresis. Urine analysis and urinary Bence jones protein (BJP) was normal. Serum calcium was 8.0mg/dl, creatinine 1.5mg/dl, urea 25mg/dl, HCO_3 20mmol/l and fasting blood sugar was 49mg/dl. No evidence of systemic myelomatosis was detected. An abdominal ultrasound scan confirmed hepatosplenomegaly with spleen containing kidney showed multiple cysts (the largest measuring 12.4cm *1.9cm). Supportive measures were instituted: two units of packed erythrocyte transfusion was given over 2 day period, intravenous perflacine 200mg 12hrly for 10 days, haematinics and adequate fluid hydration.

Post transfusion Hct was 0.361/l, WBC $3.0 \times 10^9/l$ and platelet count was $100 \times 10^9/l$. owing to the atypical/unusual clinical presentation of massive splenomegaly, reduced performance status with a previous history of haematuria and lack of typical features of MM (renal failure, hypercalcemia and bone pain), polychemotherapy could not be instituted. Ten days later he was commenced on a 4-day pulse of high-dose dexamethasone 12mg 8hrly/day 500 ml of 5% dextrose/water, and 40mg/d p.o on 15-18 every 4 weeks. The patient's condition improved gradually and he was discharge 28 days after admission. Laboratory features at discharge showed Hb 12.5/dl, WBC $3.7 \times 10^9/l$, platelets $115 \times 10^9/l$ and clinically spleen had decreased to 19cm. Therapy was maintained as described above and the patient was given a monthly clinic appointment. 3 months later on routine clinic visit, the spleen had decreased to 10cm. liver was nonpalpable and patient was remission with Hb 12.0g/dl, WBC $4.0 \times 10^9/l$, platelets $120 \times 10^9/l$ and a repeat bone marrow revealed 6% plasma cells.

DISCUSSION

The diagnosis of smoldering myeloma (SM) is challenging and early identification of the disease is crucial as treatment is controversial in regards to obtaining remission. In this case report, the clinical presentation was unusual as patient presented with features of SM associated with a huge abdominal tumor involving the spleen, liver and left kidney. A similar case report of indolent myeloma in a 62-year-old woman who was under observation for 6 years period also developed a huge abdominal tumor.⁴ This imply subtype in terms of the natural history of MM.

Treatment is generally unsatisfactory in patients with SM who have a low tumor mass and show disease progression. This is because those at risk for early or late disease progression could not be distinguished in order to prevent complications.⁵ They are often observed for a period of time before therapy is instituted.⁶ In advance economy, improvement in diagnostic techniques such as MRI, CT scan have resulted in early diagnosis and prompt treatment. In resources poor economy like ours, it is likely we may be seeing only symptomatic cases for a long time to come. Therefore, the "watch and wait" approach may not be feasible in our environment for now. The conclusion of dimopoulos et al is therefore applicable at present where the extent of disease at diagnosis and the subsequent rate of disease evolution are considered major factors in the total survival time.⁵

We felt compelled to treat this patient with a nonchemotherapeutic agent because of the evidence of mild bone marrow plasmactosis, massive

splenomegaly including lymphadenopathy in the absence of active features of MM as manifested by (CRAB). In addition, a long-term follow-up of SM case in a recent study revealed that 25% of patients who received alkylating agents developed myelodysplastic syndrome (MDS) or acute leukemia.³ Also, chemotherapy is known to be effective in reducing bone pains and local radiotherapy for localized bone resistant to chemotherapy.⁷ Hence, the more active approach with the use of nonchemotherapy agents in selected patients and use of bisphosphonates in patients with or to prevent bone lesions or osteoporosis.³ Our patient was given high-dose dexamethasone. The use of dexamethasone is known to reduce tumor burden in selected patients who do not yet have overt MM or who are not candidates for chemotherapy.³ Also, the rate and extent of fall in paraprotein concentration can be increased by using high-dose which produces impressive serological response^{9,8}. The role of dexamethasone has also been recently established and more research are still ongoing in multicentre randomized trials (IFM 95-01) on the use of only dexamethasone.⁵ Initiating dexamethasone promptly to prevent paraplegia have also been found to be critical in the management of these patients.¹⁰ Our patient showed a remarkably improvement within the first few months of therapy with dexamethasone as the haematological parameters increase and there was clinical improvement.

We conclude that high-dose of dexamethasone is useful in case of SM in the elderly particularly those with unusual presentation of multiple symptomatic features. It is an effective, well-tolerated treatment protocol for patients who do not yet require chemotherapy.

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