A YOUNG PATIENT WITH MULTIPLE MYELOMA

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Summary

Multiple myeloma, generally a disease of the elderly, is a complex disorder that causes a multitude of clinical symptoms and signs and therefore presents significant diagnostic challenge to the clinician specially if the age of onset is unusually early. Here we present one young patient suffering from multiple myeloma.

Key words

Multiple myeloma; MGUS; plasma cell leukaemia

Introduction

The plasma cell dyscrasias are broad range of clinical disorders with monoclonal gammopathy of unknown significance (MGUS) and plasma cell leukaemia at the extremes of clinical presentation [1]. Multiple myeloma, a malignant plasma cell disorder is a disease of the elderly with a peak age incidence between 60-70 years [1]. Fewer than 3% of patients with multiple myeloma are younger than 40 years [2]. A case of multiple myeloma presenting in a young patient is described.

Case Report

A 20-year man presented with pain in the central chest, weakness and anorexia for 3 months. He was working in Saudi Arabia, where he went 8 months back and used to work a gardener with good health until 3 months back when he developed his symptoms. He was having gradual onset of intermittent sharp, moderate to severe central chest pain which was precipitated by heavy work and movement. The pain disturbed his sleep. He also had tiredness and loss of appetite. For these he had to take leave from his duty. He consulted with medical facility there but no specific diagnosis was made. He returned home 15 days before presenting to us and consulted with a general practitioner for his problem who found him to be mildly anaemic and was having a very high ESR during evaluation.

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The patient was admitted in Medicine Department of Bangabandhu Memorial Hospital, USTC. We evaluated the patient. He was having average body built. Physical examination was normal except for the presence of pallor. Complete blood count shows haemoglobin 8.2 gm/dl, ESR 140mm at the end of 1st hour, Total count of WBC 9,5000/cm, differential count : neutrophils 46%, lymphocytes 30%, monocytes 10% , eosinophils 04%, Basophils 00%, myelocytes 06%, Atypical cells 04% , platelet count 70,000/cmm, hematocrit 25%. Peripheral blood film showed leucoerythroblastic blood picture with thrombocytopaenia and marked red cells rouleaux formation, his chest x ray & Ultrasonogram of whole abdomen was normal. After initial evaluation we listed 3 possibilities which are leukaemia, multiple myeloma and bone marrow metastasis from unknown primary site. We did X-ray of skull lateral view (fig-1) which showed multiple rounded lytic lesion of variable size, which can be found in multiple myeloma and metastasis from unknown primary. Bone biochemistry (serum calcium, serum phosphate & serum alkaline phosphatase) was normal which made metastasis from unknown primary very unlikely. Urine for Bence Jones protein was negative. Serum protein electrophoresis revealed a monoclonal peak in the gamma region (fig-2). Bone marrow examination from the sternum showed grossly hypercellular marrow with replacement of marrow by sheets of plasmablast along with fair number of atypical plasma cells with depression of erythropoietic, granulopoietic and megakaryocytic activity.

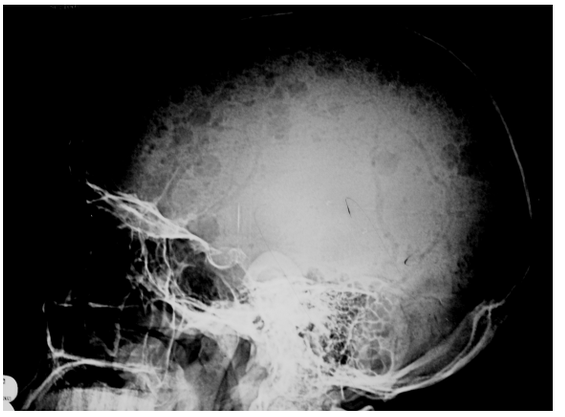
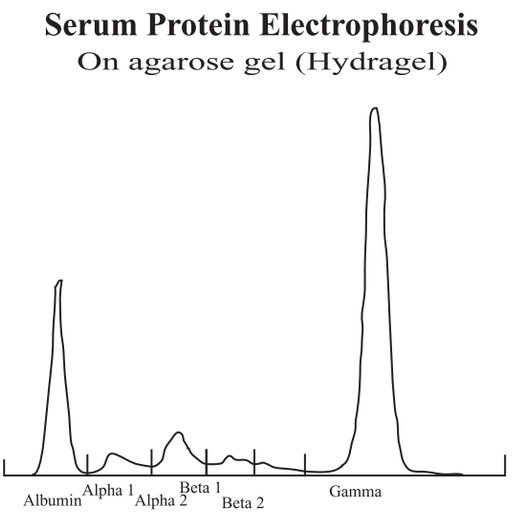


Fig 1 : X-ray skull lateral view showing multiple rounded lytic lesions of various sizes



Fraction	%	g/l	Normal %	g/l
Albumin	20.7	18.8	60-71	39-46
Alpha 1	4.7	4.3	1.4-2.7	0.9-1.7
Alpha 2	8.0	7.3	7-11	5-7
Beta 1	4.1	3.7	6-9	4-6
Beta 2	2.1	1.9	2-5	1-3
Gamma	60.4	55.0	8-16	5-11

Comments : Monoclonal Gammopathy.

Fig 2 : Serum protein electrophoresis showing monoclonal peak in the gamma region

Discussion

Multiple myeloma is the most common primary bone malignancy, accounting for 10% of all haematologic malignancies and 1% of all cancers [3]. In the United States, there are an estimated 16000 new cases and more than 11000 deaths yearly due to multiple myeloma [3]. The frequency of multiple myeloma in patients younger than 30 years accounts for 0.3% of all myelomas [4]. In another study Hewell and Alexanian reported a frequency of 1% for patients aged less than 30 years with multiple myeloma [5]. A literature search by Geetha and colleagues revealed only 11 patients diagnosed at or below age 20 years since 1973 [6]. The classic presentation of multiple myeloma is anaemia, back pain, and an elevated sedimentation rate in an older man [3]. In addition, pain in the rib cage and pain associated with pathologic fracture, most commonly of the hip, prompt the patient the patent to seek attention [3]. Physical examination may reveal pallor, an elevated temperature, bone tenderness, and soft tissue masses [3]. In addition; spinal cord compression may also occur because of tumor masses and may initiate signs and symptoms of a neurologic deficit [7].

Replacement of bone marrow initially causes anemia and progresses later to overt bone marrow failure [8]. Destruction of bone results in bone pain, osteoporosis, hypercalcaemia, and pathologic fractures [9]. Although multiple myeloma is a well defined clinical entity , establishing the diagnosis can be difficult especially when it appears in a young individual. However, important clues that alerts the clinician to the presence of the entity may appear in the history and clinical examination of the patient.

This case reflects the challenges involved in diagnosing the onset of multiple myeloma occurring at an early age. In an analysis of 178 cases of multiple myeloma occurring over a period of 7 years (1993-1999), Usha et a1 uncovered only 14 cases in patients younger than 40 [10]. Such cases generally involved men as the predominating sex, with typical presenting complaints of backache, pain in the pelvis, and weakness, accompanied by anaemia and an elevated sedimentation rate.

Multiple myeloma may present with various complications relative to the disease and treatment. Although it is beyond the scope of this article to delineate and discuss very potential complications, some of the principle affectations need to be recognized in clinical practice.

Our patient presented with chest pain, lethargy, anaemia and a very high ESR. Infections like tuberculosis, rheumatic disease and haematological or other malignancy can give rise to a very high ESR and anaemia. But absence of fever and joint pain made first two possibilities unlikely. More over peripheral blood film showed leucoerythroblastic blood picture which can occur in haematological malignancies like leukaemia multiple myeloma or secondary in the marrow from an unknown primary site. XR skull showed multiple lytic lesions which may occur in multiple myeloma and metastatic malignancy. But bone biochemistry was completely normal. Bone marrow study and serum protein electrophrosis confirms the diagnosis of multiple myeloma.

Therapeutic options for patients with multiple myeloma in young depend on disease stage and the patient's general state of health. High-dose chemotherapy with haematopoietic stem cell rescue has been reported to confer superior event-free and overall survival. Induction chemotherapy consists of VAD (vincristine, doxorubicin plus dexamethasone), or dexamethasone plus thalidomide [11]. Newer agents such as proteasome inhibitor bortezomib are now being considered for first line therapy. The benefits of thalidomide include oral administration and little haematological toxicity, but this must be weighted against the risk of neuropathy and venous thrombosis.

The clinical behavior of multiple myeloma in adolescents and young adults has been suggested to be more indolent [12,13,14]. In patients with multiple myeloma, conventional chemotherapy results in a response rate of 50-60%, and the median survival is 2-3 years [1]. The median duration of survival in patients younger than 30 years is found to be higher. Blade et al reported a median survival of 87 months in 10 patients with multiple myeloma diagnosed under 30 years of age, 69% of these patients had a predicted survival of more than 5 years and 31% more than 10 years [4]. These Results support the beneficial effect of a very young age on survival in patients with multiple myeloma.

Our patient was referred to hematologist for management and got treatment for several months under his supervision. But unfortunately patient died 2 years after diagnosis.

Conclusion

A high level of clinical suspicion is needed to diagnose multiple myeloma at an early age. Failure to consider the diagnosis may lead to medico legal pitfalls in practice. Early diagnosis of multiple myeloma is a relatively young patient offers good prognosis if optimum treatment is given.

Disclosure

All the authors declare no competing interest.

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