Hematology Quiz – Case 30

An 88 year-old Greek male was admitted at the Outpatient of our Institution due to osseous pain. Symptoms began 9 months ago with intense pain at the right proximal antibrachial region. The pain was continuous and aggravated with movements and weight lifting. Four months later, the patient visited an Orthopedic Surgeon who proposed conservative treatment with an antibrachial cast and NSAIDs for three weeks. Despite the immobilization and medical treatment pain was not relieved. Five months later, he was admitted at the Internal Medicine Department of a County Hospital due to fatigue. The patient was submitted to thorough clinical, laboratory, and plain radiographic exams that work up a lesion at the right antibrachium along with highly elevated ESR (115 mm/h), an IgG monoclonal protein and wild renal insufficiency (1.9 mg/dL). Thoracic and abdominal CT scans as well as an MRI of the antibrachium were performed and the patient was referred to our Department (fig. 1).

His medical history included an acute myocardial infarction 34 years ago with a preserved EF of 60%, benign prostate hyperplasia under medical treatment and osteoarthritis of the left surgically treated ten years ago.

On admission, the patient was asymptomatic and had no abnormal clinical findings, expect of local swelling. During clinical examination, liver and spleen were not palpable.

Complete blood counts were as follows: Hb 9.9 g/dL, Ht 30.5%, WBC 6.59×10^9/L (differential count: neutrophils 89%, lymphocytes 9%, monocytes 2%) and platelets 123×10^9/L. The erythrocyte sedimentation rate was 100 mm/h. The biochemical profile was as follows: BUN 112 mg/dL, creatinine 1.8 mg/dL, SGOT 11 IU/L, SGPT 6 IU/L, LDH 164 IU/L (upper normal limit 220 IU/L), ALP 72 IU/L, γ-GT 9 IU/L, Na+ 137 mmol/L, K+ 3.8 mmol/L, PO4 2.9 mg/dL, Mg++ 1.7 mg/dL, Ca++ 8.8 mg/dL, total serum protein 7.8 g/dL, albumin 3.3 g/dL, β, microglobulin 6.0 mg/L. Serum protein electrophoresis demonstrated a spike at the area of γ-globulins. The immunoelectrophoresis revealed a monoclonal IgGκ peak of of 3320 mg/dL. The 24h total urine protein was 1,052 g. A bone marrow aspirate and biopsy were diagnostic (fig. 2).

Comment

Multiple myeloma (MM) is the most common clonal cell disorder.
The diagnosis of MM is based on the presence of at least 10% clonal bone marrow plasma cells and a serum and/or urinary monoclonal protein in serum and urine. Myeloma is classified as asymptomatic or symptomatic depending on the absence or presence of myeloma-related organ or tissue dysfunction, including hypercalcemia, impaired renal function, anemia and bone disease (CRAB).

The International Staging System defines three risk categories determined by the serum concentration of $\beta_2$-microglobulin and albumin. The staging system helps to determine the therapy and the prognosis of the disease.

Chemotherapy is indicated in patients with symptomatic myeloma. A watch and wait policy can be applied in patients with asymptomatic myeloma. Treatment strategy depends on patient’s eligibility for autologous stem cell transplantation. Age, co-morbidities and performance status determine further treatment selection. The introduction of novel agents, as bortezomib, thalidomide and lenalidomide in combination with dexamethasone and/or chemotherapy have increased complete or partial response. Oral melphalan and prednisone remain the gold standard for elderly patients, aiming to achieve durable responses with the least possible toxicity.

The frequency of myeloma arises as the population ages. All investigations are based in the development of new therapies using proteasomes inhibitors, epigenetics agents and monoclonal antibodies.

Solitary bone or soft tissue plasmacytoma is a special manifestation of the disease requiring the presence of either more than one osseous lesion or an extramedullary tumor of monoclonal plasma cells and the absence of systemic disease criteria. The disease most often affects the axial skeleton, especially the vertebral body, causing pain and symptoms due to spinal cord or nerve root compression. Alternatively, osseous lesions can also be detected at the sternum, clavicle and scapula.

Optimal treatment mandates local radiotherapy of the affected sites. Recent literature data suggests that adjuvant chemotherapy may prevent disease progression to multiple myeloma.

Although even for solitary plasmacytoma, serum electrophoresis is pathological, monoclonal protein concentration remains constantly lower of that encountered in multiple myeloma. Progression of the disease should be monitored at regular follow ups with thorough radiological screening for the detection of osseous lesions as well as with electrophoresis and immunofixation to determine the type and concentration of the monoclonal protein.

References


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