Multiple Myeloma Presenting as Evans Syndrome

Dear Editor,

Multiple myeloma (MM) is a clonal malignancy of plasma cells which can be associated with several hematologic autoimmune manifestations. Evans syndrome (ES) is characterized by the simultaneous or sequential development of autoimmune hemolytic anemia (AIHA) and immune thrombocytopenia (ITP) and/or immune neutropenia.

A 41-year-old homemaker was admitted with fatigue and ecchymotic patches over the right thigh and gluteal region for 1 week. She was diagnosed to have ITP 10 years back and was taking prednisolone during relapse. She did not receive any other immunosuppressant drugs and had an irregular follow-up. Examination showed severe pallor and two ecchymotic patches over the right thigh and gluteal region.

Hemoglobin was 7.1 g/dl, total leukocyte count was 9070/μl, platelet count was 76,000/L, and erythrocyte sedimentation rate was 92 mm in 1 h. Peripheral smear showed hemolysis, increased rouleaux formation, normal white blood cells, and moderate thrombocytopenia. Direct Coombs test showed 2+ anti-IgG. Liver function tests showed hyperglobulinemia with reversal of albumin globulin ratio (total protein – 10.2 g/dl, 2.0 – albumin g/dl, and globulin – 8.2 g/dl). Serum lactate dehydrogenase was 850 U/L (100–190 U/L). The corrected calcium for albumin was 12.1 mg/dl.

Bone marrow showed 70% plasma cells with kappa restriction [Figure 1]. Urinary Bence Jones protein was positive. The serum IgG M component was 62.4 g/L (5.4–16.1 g/L). Free light chain assay showed κ chain concentration of 324 mg/L (3.3–19.4 mg/L) and λ 148 mg/L (5.7–26.6 mg/L), and the κ/λ ratio was 2.18 (range 0.26–1.75). Serum Beta 2 microglobulin was 4.40 μg/mL (normal range 0–3 μg/mL). Skeletal survey showed lytic lesions in skull, thoracic, and lumbar vertebrae. She had positive warm IgG antibody and antiplatelet antibodies. Platelet-associated IgG level was 226.0 ng/10^7 platelets. Antinuclear antibody was negative. A diagnosis of ES complicating MM (international staging system Stage 2) was made and was started on dexamethasone, thalidomide, and bortezomib regimen. The patient was started on oral prednisolone 40 mg daily and was tapered over the next 5 months. In the patient when followed up after 5 months, hemoglobin was 11.4 g/dl, platelet count was 142,000/L, and had a negative direct Coombs test.

The association between plasma cell dyscrasias and autoimmune disorders was previously described.[1] Chronic inflammation plays an important role in the development of lymphoproliferative disorders by randomly introducing pro-oncogenic mutations in rapidly dividing cells.[2] The spectrum of autoimmune hematologic conditions associated with MM include AIHA, autoimmune neutropenia, ITP, pure red cell aplasia, pernicious anemia, and ES.[3]

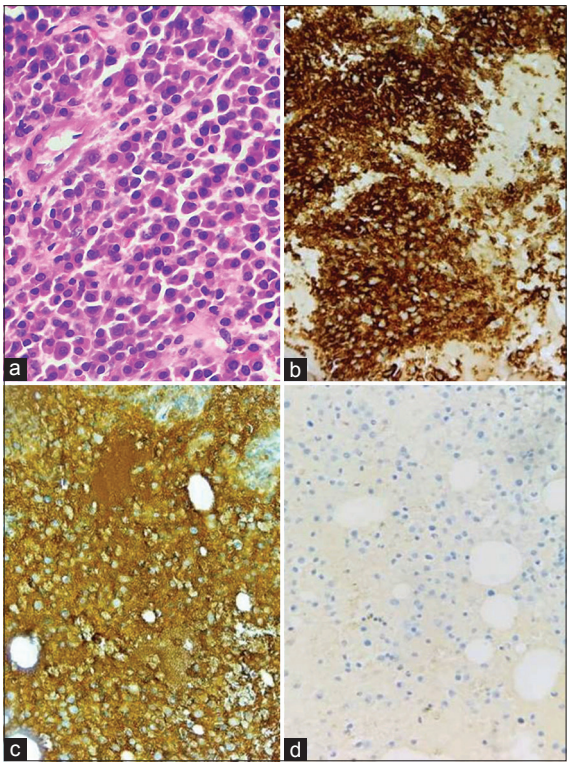

Figure 1: Bone marrow showing plasmacytosis (H and E, ×100) (a). The plasma cells were positive for CD138 (CD138 IHC, ×400) (b), positive for kappa IHC stain with the membranous/surface pattern (kappa IHC, ×400) (c) and negative for lambda IHC stain (d)

AIHA and ITP occurred simultaneously in 37 cases (54.5%) of ES, ITP preceded the onset of AIHA in 20 cases (29.5%), whereas AIHA was the first manifestation in only 11 patients (16%) in a previous study. When occurred sequentially, the mean delay between both cytopenias was 4.2 plus or minus 3.5 years.[4] ES was described in patients with MM previously.[5] The diagnosis of ES preceded the diagnosis of MM by 4 years in one.[5] Three cases had a diagnosis of ES and MM simultaneously. Our patient was diagnosed to have ITP 10 years before the diagnosis of AIHA and MM. ITP preceding the diagnosis of MM by several years was previously reported.[3] We describe a patient with MM and ES to prompt the treating physician to look for an underlying plasma cell dyscrasias in patients with such extremely rare presentations.

Informed consent

Informed signed written consent was taken from the patient involved.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understand that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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Quick Response Code:

Website: www.ccij-online.org

DOI: 10.4103/ccij.ccij_35_19

How to cite this article: Abdulla MC. Multiple myeloma presenting as Evans syndrome. Clin Cancer Investig J 2019;8:174-5.