Case presentation

Amyloid Purpura – One Of Many Systemic Manifestations Of Light Chain Amyloidosis

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Abstract

An 83-year-old female was admitted to the hospital in 2019 following the investigation of a normocytic, normochromic anemia and an elevated ESR. The patient complains of paresthesia of the upper limbs, fatigue and dysphagia for the last two months. She has a history of systemic arterial hypertension and stage 2 chronic kidney disease (CKD).

The clinical examination reveals: numerous purpuric macules on her chest, periorbital purpuric, nonpruritic lesions, macroglossia, submandibular gland hypertrophy and peripheric, white, puffy edema on her legs and right arm.

Following her presentation, the patient was referred to the hematology department for further testing. The diagnosis of IgG kappa secreting MM stage I (ISS) was based on bone marrow aspiration; whereby secondary AL amyloidosis was confirmed by abdominal wall fat pad biopsy. At her presentation the patient had multiple neuropathies, with no pathological findings at the cardiac examination.

Multiple myeloma (MM) is defined by neoplastic proliferation of monoclonal plasma cells. A precent of 10-15% of patients with MM can develop AL amyloidosis, one of the most common and most sever forms of systemic amyloidosis[1]. Mucocutaneous involvement is typically found in 30% of patients with amyloid light chain amyloidosis. The ability of amyloid fibrils to bind factor X, resulting in factor X deficiency, is a one of hypothesis of amyloid purpura[3], while coagulation factor inhibitory paraprotein and hyperfibrinolysis add to the bleeding diathesis[4].

Macroglossia results in firmness of the tongue and difficulties in speech in swallowing[2]. The “racoon eyes” sign, or bilateral, periorbital purpura, is an unmissable hint leading to the diagnosis. The purpuric lesions can be also found on the neck, chest and gastrointestinal tract, leading to malabsorption[3]. Peripheral neuropathy, postural hypotension, fatigue, oedema and weight loss all add up to the clinical picture of amyloidosis[4].

The treatment of choice for this patient included Bortezomib, Melphalan and Prednisone (VMP) and was discontinued after three cycles – due to the lack of adherence to the treatment schedule.

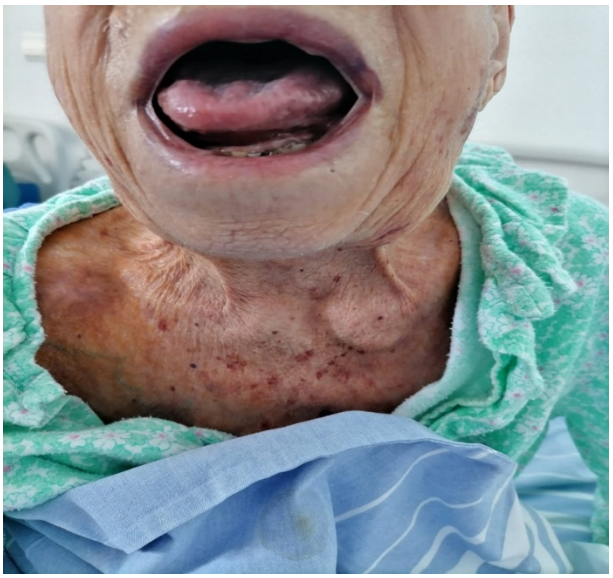

Figure 1. Characteristic purpura seen on the neck and chest and macroglossia.

This case highlights the rare presentation of secondary amyloidosis, that can burden the prognosis of multiple myeloma, but simultaneously pave the way to a faster diagnosis, owed to the specific clinical presentation.

References

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