Multiple flesh-colored papules on the waist and multiple purpuric nodules on the tongue

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Case report

An 83-year-old man presented with a 1-month history of multiple flesh-colored waxy papules over his bilateral waist, and bilateral inguinal and suprapubic areas. He had also noted multiple blister-like nodules over his buccal mucosa and various-sized purpuric tumors on his tongue for over a year. Both types of lesions were asymptomatic. His medical history included hypertension, chronic renal insufficiency, and previous cerebrovascular accident, and he was regularly followed up at our hospital. There was no relevant family history and no constitutional symptoms, except for a 1-month history of right hip and right shoulder pain.

Two skin biopsy specimens were obtained from the abdomen and lip. Histopathologic examination of the abdominal papule revealed a large amount of pink amorphous deposits in the superficial dermis. Histopathologic examination of the lip showed similar deposits in the submucosa and small vessel walls. Congo red staining of both specimens revealed green birefringence under polarized light.

Protein electrophoresis revealed a pattern indicative of monoclonal gammopathy, with a spike in the beta region. Immunofixation electrophoresis of blood and urine disclosed IgA-lambda paraprotein and lambda light chain paraprotein, respectively. Biopsy results from the bone marrow revealed sheets of plasma cells, including some immature forms, and occasional Dutcher bodies. The plasma cells accounted for up to 70% of all nucleated cells focally. No evidence of bone or renal involvement was found in skull X-ray or renal echo examinations. However, magnetic resonance imaging of the heart revealed abnormal thickening and akinesia of the septal wall of the left ventricle.

Figure 1 (A,B) Multiple flesh-colored, waxy papules over abdomen, bilateral waist, and bilateral inguinal and suprapubic areas.

Figure 2 (A,B) Multiple blister-like nodules over the buccal mucosa, lip and various-sized purpuric tumors on the tongue.
Diagnosis

Systemic amyloid light chain (AL) amyloidosis with multiple myeloma.

Discussion

After diagnosis was confirmed, melphalan-prednisolone therapy was initiated, with gradual resolution of the lesions.

Systemic amyloidosis is indicated by pink amorphous material in the dermis, subcutaneous tissue and blood vessel walls. Staining with Congo red under polarized light shows characteristic apple-green birefringence. Systemic amyloidosis can be further classified according to the type of protein deposited: (1) systemic AL amyloidosis associated with monoclonal gammopathies or multiple myeloma; (2) systemic amyloid A amyloidosis associated with chronic inflammation or infection; and (3) other rare forms of amyloidosis related to long-term dialysis or hereditary amyloidosis. Systemic AL amyloidosis is the most common type of systemic amyloidosis, accounting for more than 60% of cases.

AL protein is derived from the overproduction of monoclonal immunoglobulin light chains because of plasma cell dyscrasias, such as multiple myeloma. The clinical manifestations depend on where the AL protein is deposited; around 40% of patients with systemic AL amyloidosis have cutaneous findings. Other organs and systems involved include the heart, liver, kidney, and peripheral and central nervous systems.

Mucocutaneous findings may be the first sign of systemic amyloidosis, especially in multiple myeloma. The reported incidence of amyloidosis in multiple myeloma ranges from 10–20%. Waxy papules, nodules or plaques are the most characteristic skin manifestation, and may present on the face, neck, or inguinal and anogenital regions. Other rare but pathognomonic skin findings include periorbital petechiae (raccoon sign) and ecchymoses.

Macroglossia can be marked in multiple myeloma, but was only found in 22% of patients in a Mayo Clinic report. Of patients with AL amyloidosis, 50% have lingual involvement and only 5% have macroglossia. Other differential diagnoses include hypothyroidism, diabetes, sarcoidosis and neoplasms. Macroglossia is caused by amyloid deposition in the tongue, which becomes firm, dry, fissured, and hemorrhagic with lateral scalloping from impingement on the teeth. Nodular macroglossia occasionally presents as papules, nodules or plaques on the oral mucosa and tongue. Although most patients with oral amyloidosis associated with multiple myeloma present with macroglossia, some authors believe that mucosa nodules are more specific to amyloidosis, since tongue enlargement can also occur in the absence of amyloidosis. Oral amyloidosis is sometimes the initial presentation of multiple myeloma. Dermatologists should therefore be alerted to potentially serious underlying problems in patients who present with increased size or nodular formation of the tongue.

The diagnosis of multiple myeloma should meet three criteria: (1) presence of an M-protein in the serum and/or urine; (2) more than 10% of clonal plasma cells in the bone marrow; and (3) evidence of end-organ damage (hypercalcemia, renal insufficiency, anemia or bone lesions).

The prognosis of systemic amyloidosis is poor, regardless of the presence of multiple myeloma. Pardanani et al reported a survival rate of 32 months in patients with systemic amyloidosis alone, compared with 14 months in those with associated multiple myeloma. When systemic amyloidosis is suspected, dermatologists should be prompted to investigate the possibility of an underlying malignancy.

References