Unusual and memorable

Case Number 20: Series editor: Gary D Wright

A 70 year old recent Russian female emigrant was admitted to our ward with a deep venous thrombosis of the left femoral vein. She reported a four year history of seronegative "rheumatoid arthritis" (RA). She also had symmetrical, shiny, smooth, firm, and spherical papules and nodules of waxy colour that coalesced to form band-like lesions on her face (fig 1). These lesions started one year before admission and progressed over time. Her tongue was diffusely enlarged.

Figure 1 Amyloid papules and nodules on the face. (Reproduced with permission of the patient.)

Examination of her joints disclosed synovial swelling and tenderness of both wrists (fig 2), with reduced range of motion and mild ulnar deviation of both hands. There was mild synovial swelling of the knees. A neurological examination was normal.

Figure 2 Amyloid synovial thickening of the wrist

The rest of the physical examination was negative. Blood chemistry, cell count, urine analysis, electrocardiogram, and chest x ray were normal. Total serum protein was 68 g/l (normal 62–88) and albumin 43 g/l (normal

35–52). Antinuclear antibodies and rheumatoid factor were negative. An x ray examination of the hands showed swelling of soft tissue at the wrist, osteopenia and no evidence of erosions. An x ray examination of the knees was negative. Skin biopsy of one of the facial skin lesions showed eosinophilic amorphous deposits in the papillary and reticular dermis (fig 3) and in the wall of large and medium sized vessels in the deep dermis. After Congo red staining these deposits showed green birefringence when viewed by the polarised light microscope. Bone marrow biopsy showed a 50–60% infiltration of premature and mature plasma cells.

Figure 3 Amyloid deposition in the papillary and reticular dermis.

Immunoelectrophoresis showed IgG 4.91 g/l (normal 8–16), IgM 0.44 g/l (0.45–1.5), and IgA 0.53 g/l (normal 0.9–3.2) with a λ chain monoclonal fraction. Bence-Jones protein in the urine was also found: 117 mg of urinary protein was found in a 24 hour collection (λ light chains). An open synovial biopsy from the left wrist showed amyloid infiltration with no evidence of any inflammatory changes.

Computed tomography of the abdomen and an echocardiogram were normal. A skeletal x ray survey showed osteoporosis with no evidence of lytic lesions. The patient was diagnosed with primary systemic amyloidosis.

Although nodules, papules, and plaques are listed among the common skin manifestations of systemic amyloidosis,¹ the extent and facial distribution of these lesions on presentation of our patient is unique. Joint symptoms and signs are common manifestations of AL amyloidosis (primary systemic amyloidosis or myeloma amyloidosis). It affects mainly large joints and may resemble and be misdiagnosed as RA.² "Shoulder pad sign" due to amyloid infiltration of the shoulder joint is characteristic. Arthropathy due to deposition of β_2 microglobulin amyloid (reactive amyloidosis in patients receiving long term haemodialysis) is also not uncommon. These patients may also have tenosynovitis and develop bone erosions. On the other hand, the clinical manifestations of AA amyloidosis (most common type of reactive amyloidosis) are primarily renal disease and amyloid arthropathy is uncommon.

1 Brownstein MH, Helwig EB. The cutaneous amyloidosis. II. Systemic forms. Arch Dermatol 1970;102:20–8.

2 Gordon DA, Pruzanski W, Ogryzlo MA, Little HA. Amyloid arthritis simulating rheumatoid disease in 5 patients with multiple myeloma. Am J Med 1973;55:142–54.

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