Multiple Myeloma with Extramedullary Plasmacytomas

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Many systemic diseases are associated with cutaneous manifestations, and multiple myeloma is no exception. Skin ulcers often present interesting problems, most of them due to varicose veins and their complications or to arterial insufficiency, but a few demand fuller explanation. Occasionally they reflect an underlying blood dyscrasia, collagen disorder, neoplastic condition, or other form of internal disease. In the following case ulceration of skin proved to be a feature of a previously unrecognized multiple myeloma.

CASE REPORT

The patient, a 78-year-old man, was first seen in May 1965. He had been well until 12 months previously, when he had noticed an ulcer on the skin of the right thigh. Starting as a small nodule, it became enlarged to a size of approximately 1 in. (2.5 cm.) in diameter and ulcerated. This healed without any specific treatment, but was quickly followed by similar lesions on the abdomen, left shoulder-blade, and anterior chest.

In addition to the skin lesions he was troubled with severe pains in the back, radiating to the lower abdomen. He was seen in the surgical outpatient department on account of arthritis in both hips, and was found to have an enlarged, hard prostate. As this was thought to be malignant, he was given stilboestrol therapy.

Examination showed a frail, kyphotic old man who had great difficulty in walking. He was moderately anaemic, but there was no lymphadenopathy or hepatosplenomegaly. Blood-pressure was 190/90. The skin showed ulcerated lesions about 2 in. (5 cm.) long near the sternal area, and a smaller ulcer on the left shoulder-blade. These were irregular in shape, shallow, and had clean-cut margins (not unlike those of pyoderma gangrenosum). There was no evidence of infection. Healed oval lesions with slight discoloration were also present on the right thigh and abdomen. Purpura was not noted.

Investigations.—Hb 10.2 g./100 ml. W.B.C. 5,000/c.mm. Film showed occasional metamyelocytes, but no overall shift to the left. Red blood cells normochromic, with a mild degree of macrocytosis. Platelets plentiful. E.S.R. 60 mm. in first hour (Wintrobe-corrected 26 mm.). Serum calcium 9.1 mg./100 ml. Serum alkaline phosphatase 8.3 K.A. units. Serum acid phosphatase 3.4 K.A. units. Blood urea 24 mg./100 ml. Serum total proteins 8.5 g./100 ml. (true albumin 2.8 g., total globulins 4.8 g.), A.G. ratio 0.6/1. Immunoelectrophoresis showed a band in the γ -globulin position strongly suggestive of the presence of myeloma protein. There was no evidence of cryoglobulins. Urine electrophoresis revealed traces of albumin and of β - and γ -globulins. Bence Jones proteins were present. Bone-marrow films showed preponderance of plasma cells consistent with diagnosis of myelomatosis.

X-ray examination: "There are multiple lytic deposits in the skull. They are varied in size and shape. . . . There are multiple deposits in the spine and pelvis, and many of the vertebral bodies have collapsed. All the lesions are lytic. There is extensive arterial calcification."

Skin biopsy: "Paraffin sections showed an ulcer of the skin with granulation tissue. Numerous plasma cells were seen in the granulomatous infiltrate."

In view of his poor general condition and the extent of his disease, symptomatic therapy only was instituted. He was readmitted six months later with extensive skin ulceration and bronchopneumonia,

and died two days later. Necropsy findings confirmed the diagnosis of multiple myeloma with characteristic deposits in skull, sternum, and vertebrae. Cause of death was pulmonary collapse and bronchopneumonia.

COMMENT

Extramedullary plasmacytomas are comparatively rare tumours, and have a predilection for the mucous membranes, especially the upper part of the respiratory tract. Hellwig (1943) collected many cases which were histologically identical with plasmacytoma of the bone-marrow. The first case report was that of Bruno Bloch in 1910. His patient was a 67-year-old man who had lesions on the extremities and chest which began as erythematous macules and then became papules with scales and crusts. Necropsy revealed multiple myeloma with metastatic lesions.

Many authors have reported skin changes in multiple myeloma, though ulceration is infrequent. Hicks (1950) described a non-specific ulcer in multiple myeloma which did not contain plasma cells. Cowan (1961) described one case of skin ulceration in multiple myeloma, but as there were no bony changes, and the abnormal protein was in the β band, this case was possibly one of macroglobulinaemia.

The most important contribution is that of Bluefarb (1955), who divided the skin changes into specific and non-specific groups. The latter consist of complications like amyloid, cryoglobulins, pancytopenia, toxic cutaneous lesions, and, by involvement of internal organs, produce many diverse features. None of the non-specific features appeared in our patient. The specific group consists either of skin lesions which are extensions from tumours of the bone or of extramedullary plasmacytomas. The lesions seen in our patient, even though some were present on the bony surfaces, could not have been due to the extensions from underlying osseous involvement, as the healed lesions were easily movable on the bony surface, and a similar lesion was present on the abdomen.

Impaired blood supply and repeated minor trauma have been suggested as an explanation for ulceration, but this is unsatisfactory in the absence of cryoglobulins and poor peripheral circulation.

It can be reasonably assumed that this patient's ulcers were extramedullary plasmacytomas, especially as plasma cells were found in abundance on skin biopsy. According to Lewis *et al.* (1951) the diagnosis of extramedullary plasmacytoma tumours can be made only by histological examination, because of the tremendous variation in the clinical picture.

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