Bacillary angiomatosis

The first case reported in South Africa

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Abstract **A 28-year-old white man, positive for HIV, who was admitted to hospital for pneumonia, had for 2 months had several fluctuating cutaneous purple nodules on his legs and abdomen. Cultures of two lesions were negative, but light and electron microscopy showed organisms characteristic of bacillary angiomatosis. The patient responded well to therapy with erythromycin. This is the first reported case of bacillary angiomatosis in South Africa.**

S Afr Med J 1993; **83:** 855-856.

Bacillary angiomatosis, first described in the USA in 1983,[1] is an infectious disease that affects immunocompromised hosts. It is characterised by pseudoneoplastic angioproliferative cutaneous lesions and may also involve internal organs.[2] It is caused by a pleomorphic bacillus, probably *Rochalimaea henselae*.[3] We report on the first documented South African patient who presented with skin lesions, unusual for this disease. As the organism is difficult to culture and the disease responds well to treatment, it is important for clinicians to be aware of the condition and to do biopsies to verify the diagnosis.

Case report

The patient was a 28-year-old white homosexual man who had been confirmed as HIV-positive by means of enzyme-linked immunosorbent assay (ELISA) and Western blot in July 1991. He had never travelled overseas. In August 1992 he complained of a cough productive of yellow sputum, and pneumonia was diagnosed. On admission to hospital, 8 cutaneous lesions were noted on both thighs, his left calf and abdominal wall. The lesions had appeared at different times during the preceding 2 months. The lesion on his right thigh resembled a carbuncle; it was dome-shaped, purplish in colour, fluctuant in the centre and draining pus from a central ulceration (Fig. 1). All the other lesions were soft, non-fluctuant, erythematous, purplish subcutaneous nodules only slightly raised above normal skin surface.

Chest radiographs revealed an extensive bilateral patchy infiltrate.

Laboratory investigations

A full blood count was done, the results of which were as follows: haemoglobin 7,4 g/dl, white cell count 3,6 x 10^9/l, platelets 168 x 10^9/l, and CD4 count 21 cells/μl. Blood and sputum cultures and serological tests for *Mycoplasma pneumoniae* were negative. Culture of pus from the ulcerated lesion was negative for bacteria and fungi.

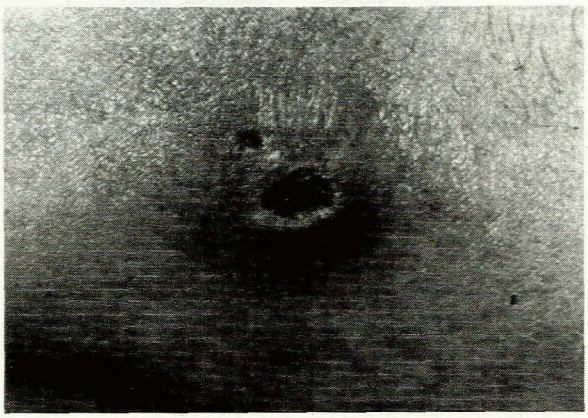

FIG. 1.
Healing ulcerated lesion on right thigh, 4 days after erythromycin was started. Biopsy wound left upper quadrant.

Skin biopsies

The ulcerated nodule on the thigh and a subcutaneous nodule on the abdomen were biopsied 2 days after antibiotic therapy was initiated. The epidermis was ulcerated in the thigh lesion and slightly atrophic in the other. Apart from this, the lesions had similar histological features. Within the dermis there were proliferating vascular channels that showed a lobulated pattern. These vessels were lined with plump endothelial cells which showed moderate cytological atypia, mitoses and small areas of necrosis. Abundant neutrophils showing leucocytoclasis and foamy macrophages were present adjacent to the capillaries. There was abundant extracellular eosinophilic material, also associated with the capillaries. Periodic acid-Schiff, acid-fast and Gram stains failed to stain this material, while Warthin-Starry stains showed it to be composed of clumped, argyrophilic, extracellular interstitial bacteria (Fig. 2). Electron microscopy confirmed the presence of extracellular bacilli with trilaminar cell walls (Fig. 3).

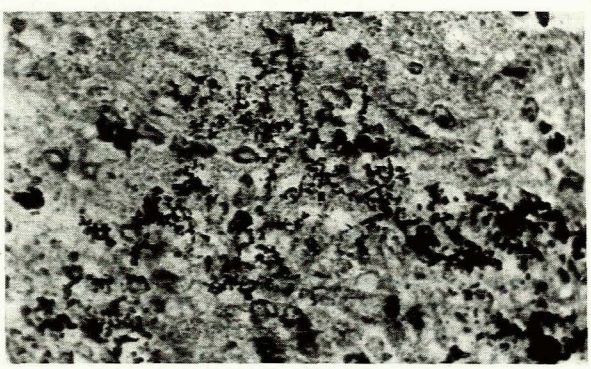

FIG. 2.
Clumped masses of interstitial argyrophilic bacteria (Warthin-Starry x 400).

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Accepted 11 Feb 1993.

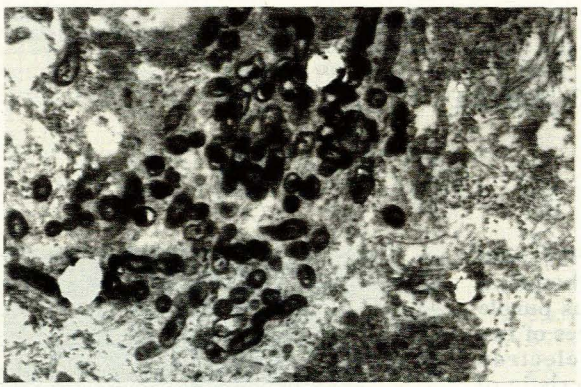

FIG. 3.
Electron micrograph showing pleomorphic bacilli with trilaminar cell walls (x 12 600).

Course

The patient was treated with oral erythromycin, 500 mg 4 times a day. After 12 days of therapy he was apyrexial and no longer coughing; he refused further hospital treatment, but returned 2 days later, at which stage a further course of erythromycin (500 mg 4 times a day for 20 days) was prescribed. All the skin lesions resolved within 4 weeks.

Discussion

In 1983 Stoler *et al*[1] described the sudden appearance of multiple subcutaneous nodules in which abundant clumped organisms were seen in a patient with AIDS. Subsequently bacillary angiomatosis was reported mainly in patients with AIDS[1-5] but several cases have been reported in other immunosuppressed patients[6] and at least 1 case has occurred in an immunocompetent individual.[7]

The cutaneous lesions in bacillary angiomatosis may be solitary, few in number or multiple and may occur in a dermal or subcutaneous location or on the skin surface.[4] The deeper skin lesions are usually skin-coloured while superficial lesions may be skin-coloured, red or purple, dome-shaped or accuminate. The cutaneous lesions usually start as small papules which gradually increase in size to form nodules and tumours — macules and plaques are not usually seen. Individual lesions usually resemble pyogenic granulomas or small angiomas, and the surface may be smooth, ulcerated or crusted.[4] The clinical differential diagnosis includes pyogenic granuloma, haemangioma, Kaposi's sarcoma and various subcutaneous tumours.[4] Oral, anal, conjunctival and gastro-intestinal mucosal surfaces are commonly concomitantly involved.[5] More recently, bacillary angiomatosis involving bone,[8] lymph nodes,[9] bone marrow,[10] soft tissues,[11] liver and spleen[12] has been described.

Histologically, lesions of bacillary angiomatosis are pyogenic granuloma-like, with a lobular capillary proliferation which, in the skin, may be superficial or deep. Endothelial cells are plump and there is a marked inflammatory cell infiltrate with polymorphonuclear cells and leucocytoclasis. Some cases show endothelial cell atypia and mitoses. Granular argyrophilic interstitial material is present, staining positive with the Warthin-Starry stain.[4] Electron microscopy demonstrates the presence of pleomorphic bacillary structures with trilaminal cell walls.[4,13]

The organism that causes bacillary angiomatosis has only recently been determined. Originally it was thought to be related to the organism that causes cat-scratch disease which, clinically, is quite unlike bacillary angiomatosis.[14] Then *Bartonella bacilliformis*, which causes a similar angioproliferative disease in humans, was suspected.[14] Recently Birtles *et al.*[15] demonstrated via an analysis of the 16S ribosomal RNA gene sequence that the causative agent is closely related to, but distinct from, *Bartonella bacilliformis.*

They concluded that together with *Rochalimaea quintana* and *B. bacilliformis* it represents a species from a closely related group of rickettsia-like organisms. Subsequently by using immunocytochemical techniques, Slater *et al.*[3] demonstrated that the causative agent in bacillary angiomatosis is a distinct organism, *R. henselae*, from the order *Rickettsiales.*[2]

Bacillary angiomatosis usually responds well to antibiotic therapy with oral erythromycin. A dose of 250 - 500 mg 4 times a day for 2 weeks to 2 months has given excellent results[4,5] and is considered the treatment of choice. Trimethoprim-sulfamethoxazole, isoniazid and rifampicin have also been used but with variable results.[4]

The clinical appearance of the lesions in our patient was unusual in that they resembled boils or abscesses but the histological features of angiomatous proliferation and the presence of the characteristic organisms were pathognomonic for the disease. Rapid response to erythromycin was also a feature.

It is important to recognise this condition as antibiotic treatment in this potentially life-threatening disease is curative.[4]

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