Multifocal Kaposi's Sarcoma Mimicking Allergic Vasculitis

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Accepted August 18, 2003.

Sir,

Kaposi's sarcoma (KS) is a multifocal neoplastic process occurring predominantly in the dermal vasculature (1, 2). Bluish-red macules, plaques and tumours characterize the neoplasm, which most commonly affects the feet (1–3). The recognized clinical variants of KS include classic KS, endemic African KS, KS in iatrogenically immunocompromised patients and HIV-associated KS (1, 4). The discovery of herpes-virus-like DNA in all variants of KS supports a theory of viral aetiology for this neoplasm (3, 5, 6). The clinical course of KS in iatrogenically immunocompromised patients is similar to that of the classic KS, e.g. it is characterized by slow progression and predominantly benign course, but may be more aggressive in the case of continued immunosuppression (3).

We report the case of a woman with corticosteroid-treated rheumatoid arthritis who presented with bilateral palpable purpura-like eruption on the dorsal feet initially diagnosed as hypersensitivity vasculitis. Her skin biopsy made 2 years later demonstrated typical histological signs of KS.

CASE REPORT

In April 1996 a 74-year-old female patient presented at the dermatology clinic of the University of Tartu with a 5-month history of slightly painful, gradually increasing in number, papular eruptions on the dorsal feet.

The patient had a 15-year previous medical history of rheumatoid arthritis. Additionally, she had suffered from severe coronary artery disease, cholelithiasis, and fungal foot and nail disease for an unknown period. Her past medical history included viral infections, pyelonephritis and appendectomy at the age of 24 years. For many years the patient's rheumatoid arthritis had been controlled with oral triamcinolone (4 mg day^{-1}) and occasionally with NSAIDs. She also used β-blockers, diuretics and cimetidine regularly.

Cutaneous examination demonstrated several discrete, firm, symmetrically distributed violaceous-red dome-shaped non-blanching papules of 3–4 mm in diameter on the dorsum of her feet, the first toes and on the dorsal and lateral aspects of the ankle. Affected skin as well as the skin on the left leg was slightly swollen and tender. Routine laboratory tests (full blood count, urinanalysis, rheumatological markers) were normal.

The patient was diagnosed to have hypersensitivity vasculitis and she was treated with ascorutine 0.5 g t.i.d., ibuprofene 0.2 g t.i.d. and potent glucocorticoid, administered topically. She also received topical antimycotics.

After treatment, swelling on the lower extremities disappeared and some papular elements decreased in size. However, new papules continued to emerge.

In the following 2 years the patient was twice hospitalized in the therapy clinic due to exacerbation of rheumatoid arthritis and vasculitis and treated with high doses of systemic prednisolone. The treatment relieved acute symptoms (pain and swelling of the feet and legs) but did not prevent the progression of papular eruptions. On the contrary, during these 2 years the number and size of papules (now up to 8 mm in diameter) increased and at re-admission to our clinic in April 1998 they involved all toes, both sides of the feet, and legs up to the knees. Besides pea-sized reddish papular skin lesions she had developed small confluent plaques and reddish-brown macules, 1–2 mm in size, along superficial veins in the last 3 months of the follow-up. In September 1998 the patient, now admitted to the oncology clinic so that treatment options could be considered, died as a result of atherosclerotic heart disease.

Examination of the first skin biopsy performed in April 1998 revealed a well-circumscribed encapsulated nodule in the dermis, containing vascular spaces with large ectatic thin walls in peripheries and similar in appearance to a cavernous haemangioma (Fig. 1). The central area revealed a spindle cell component containing cleft-like and sieve-like vascular spaces filled with erythrocytes and surrounded by extravasated erythrocytes and hemosiderin. In that area, eosinophilic

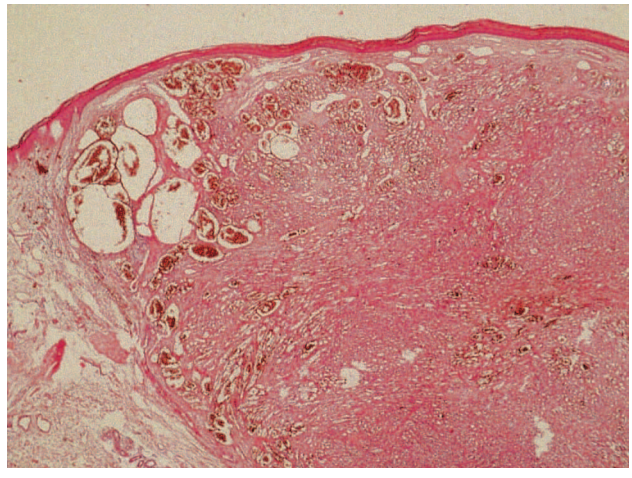

Fig. 1. In April 1998 the first biopsy specimen showing well-circumscribed encapsulated nodule with large ectactic vascular spaces in the periphery and spindle cells with slit-like and sieve-like vascular spaces in central areas (haematoxylin-eosin stain; original magnification; ×100).

DOI: 10.1080/00015550310006077

hyaline globules were easy to detect. Pleomorphism was scarcely expressed and mitotic cells were absent. On initial examination these findings were interpreted as an angiomatous malformation.

The second skin biopsy, obtained 3 months later, demonstrated more definitive signs of KS in the dermis, including an uncircumscribed prominent spindle cell component and cleft-like vascular spaces filled with erythrocytes. The area contained mitotic spindle cells, pleomorphism (Fig. 2) and a few hyaline globules that stained positively by PAS.

DISCUSSION

Immunosuppression is a well-documented risk factor for the development of KS. Iatrogenic, immunosuppressive drug-related KS is seen primarily in organ transplant recipients (5, 7), but is not an exception in patients receiving immunosuppressive therapy for many other reasons (3, 4), including rheumatoid arthritis (6).

Compared to KS, vasculitis is a far more frequent complication of rheumatoid arthritis and may occur in 5–15% of patients (8). It can be the cutaneous manifestation of systemic rheumatoid vasculitis or a hypersensitivity reaction to drugs used for the treatment (9). The presenting clinical symptom of hypersensitivity vasculitis is palpable purpura (10). Considering the absence of systemic symptoms, our patient's case was interpreted as a treatment-related hypersensitivity reaction. The two misleading clinical signs contributing to belated accurate diagnosis were, first, the abrupt onset of eruption characterized by rapid development of numerous small palpable purpura-like papules with a predilection for distal parts of the lower extremities, and, second, the small size of papules, i.e. only 3–4 mm, maximum 8 mm in diameter. Usually in KS papules and plaques are larger, reaching up to 1–3 cm in diameter in later stages (2). Two years later, when it became clear that the patient's skin eruption had lasted without any remissions and progressed, skin biopsy was finally performed. The histological features were characteristic of KS.

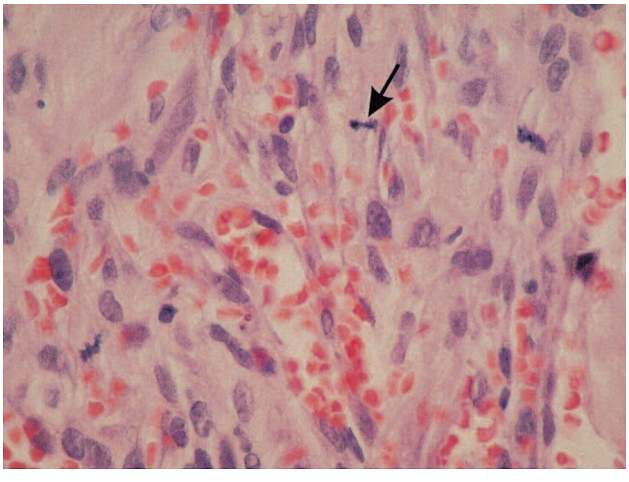

Fig. 2. The biopsy in July 1998 showing characteristic signs of KS: a spindle cell component with mitotic cells (arrow), slit-like vascular spaces and extravasated erythrocytes in the dermis (haematoxylin-eosin stain; original magnification; ×1000).

In iatrogenically immunocompromised patients, KS may resolve upon reduction or withdrawal of immunosuppression (1, 11). In the case of our patient, intensive immunosuppressive therapy continued for 2 more years, now with the additional goal of influencing extremely treatment-resistant hypersensitivity vasculitis. Classic, as well as iatrogenic, variants of KS run a protracted course and most patients die of an unrelated cause (12). Unfortunately this happened in the case of our patient.

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