Case report

CYSTIC KAPOSI'S SARCOMA

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ABSTRACT

A Case report of rare variant of Kaposi's sarcoma - cystic Kaposi's Sarcoma - is presented. Combination of alpha-interferon with retinoid treatment produced nearly complete remission of skin symptoms.

KEY WORDS

cystic Kaposi's sarcoma, interferon therapy

In 1872, Moritz Kaposi first described this entity, subsequently named after him, terming it "sarcoma idiopathicum multiplex haemorrhagicum.

The first description of the cystic manifestation of Kaposi's sarcoma is associated with the name of Pelagatti (1905). Since then, several authors, including Ronchese and Kern (1), Gange and Jones (2), Hunyadi et al (3), Recht et al (4) and Pichler et al (5) have described lymphangioma-like manifestations of Kaposi's sarcoma. Only 20 reports of such cases had appeared in the world literature before 1990.

At present, the following forms of the disease are differentiated:

1. classic Kaposi's sarcoma
2. endemic Kaposi's sarcoma
3. Kaposi's sarcoma associated with HIV infection
4. iatrogenic Kaposi's sarcoma (in conditions of immunosuppression)

The following case report describes the special cystic form of Kaposi's sarcoma in which a great number of cystic lesions are observed in addition to the typical classical clinical picture.

CASE REPORT

In May of 1988, an 85-year-old female patient presented with considerable swelling of her right leg. Livid, flattened nodules appeared first on the extensor, and later on the lateral surface of her right lower leg, as well as in the ankle region. These lesions were soft on palpation and gradually increased in number. Months later, vesicles with serous content developed, joining the previous symptoms.

At the time of hospital admission, non-pitting edematous swelling of the right lower leg was observed. Numerous, 4-10 mm diameter, sharply

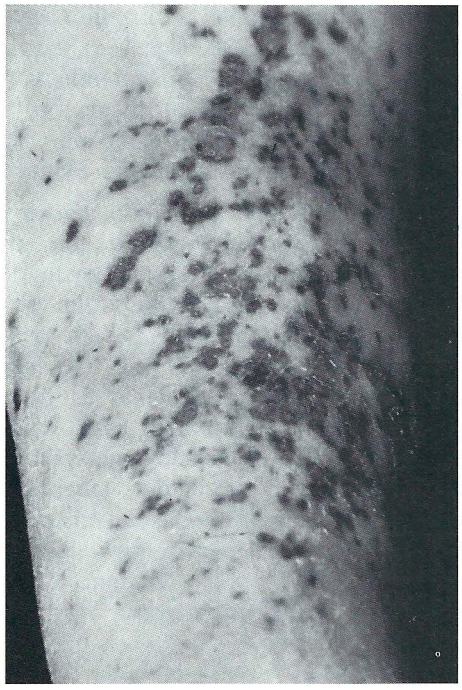

Fig. 1: Innumerable, sharply demarcated, discrete and confluent livid-red macules of 4-10 mm diameter, as well as slightly infiltrated livid bluish papules on the extensor surface of the right lower leg.

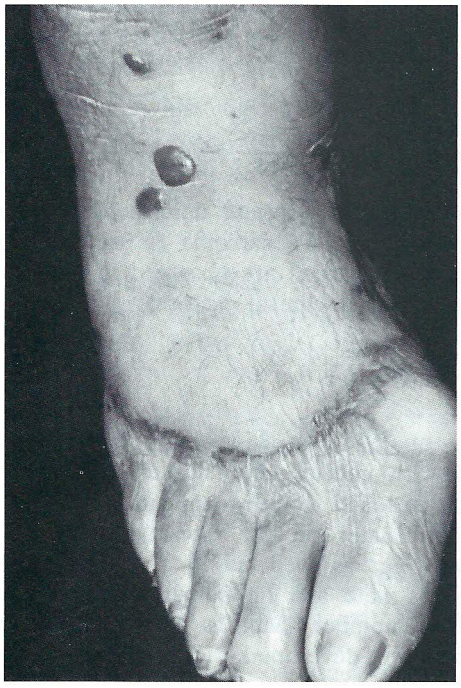

Fig. 2: Compressible cystic lesions 5-20 mm in diameter, with hemorrhagic contents on the right foot.

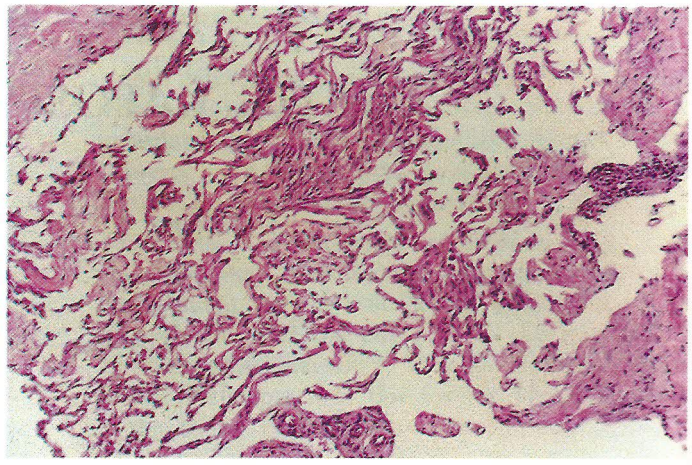

Fig. 3: Photomicrograph of a lymphedematous focus: revealing cystically dilated cavities lined with endothelium, but containing no erythrocytes. (H E stain, 100x)

demarcated, discrete and confluent livid-red macules and slightly infiltrated livid papules were evident. These lesions were distributed over the extensor and lateral surfaces of the right lower leg, in the ankle region, on the dorsum, sole, and toes I-V of the right foot, as well as on the medial and plantar surfaces of the left foot (Fig. 1). Sharply demarcated, compressible cystic structures, 5 to 20 mm in diameter. containing either serous or hemorrhagic fluid were seen on the right foot (Fig. 2). Livid red infiltrations with irregular margins were noted on the right

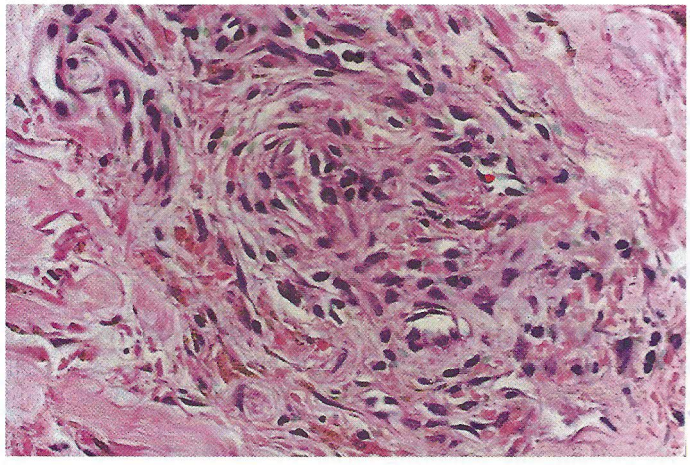

Fig. 4: Photomicrograph of a typical later stage Kaposi lesion. Cavities lined with endothelium may be seen, surrounded by a marked extravasation of erythrocytes. Large numbers of lymphocytes, plasma cells, and elongated fibroblasts are evident in the surrounding areas. (HE stain, 100x)

lower leg, and similar ones with sharp margins on the feet.

No lymphadenomegaly was observed. On the basis of the clinical picture, a provisional diagnosis of cystic Kaposi's sarcoma, which is a rare manifestation of classic Kaposi's sarcoma, was made.

Routine laboratory testing of blood and urine samples yielded values in the normal range, except for a somewhat accelerated blood sedimentation rate and cryofibrinogen positivity. HIV and RPR investigations were negative. Examination of the immunological system revealed a moderate increase in the T-lymphocyte and active T-lymphocyte counts. Skin tests with streptococcal and candidal allergens, as well as purified tuberculin remained anergic. Chest X-ray and abdominal ultrasound examination revealed no disease manifestations in the internal organs. A lymphoscintigraphic study was performed to determine the origin of the cystic structures. The results demonstrated dilated lymph vessels and increased lymph flow in the right lower leg; however, no pathological anastomoses between the lymphatic and venous systems were delineated.

Surgical biopsy specimens were taken from a lymphedematous focus (a.) and Kaposi lesion (b.) of the right leg for histological elaboration.

HISTOLOGY

a. The epithelium was found to be regular, in the dermis, cystically dilated cavities containing no erythrocytes can be seen (Fig. 3).

b. Other areas of the dermis reveal moderate edema, capillaries with swollen endothelium and containing erythrocytes, and an increase in the number of small vessels. Also, elongated fibroblasts are visible, with erythrocytes extravasated into the surrounding area (Fig. 4).

Histochemistry demonstrated coagulation factor VIII in the endothelium of all vessels. In some areas of the surrounding tissue, proliferating spindle-shaped cells also displayed factor VIII positivity.

TREATMENT

Initially, the patient received radiation treatment on each side in a total dose of 3000 R, which resulted in significant flattening of the lesions. Following a period of remission of one year, intense swelling of the lower leg developed, along with a slow spreading of the process.

Etretinate (Tigasone) 50 mg/day was administered orally. Recombinant alpha-interferon (Egiferon/Egis) treatment was instituted with 1 million IU s.c./ initially, and increased to 2 million IU s.c./ every other day.

Following an 8-week course of treatment, the lymphedema disappeared entirely, the cystic lesions became nearly completely flat, and the previously livid red infiltrations faded and became macular.

Gange (2) finds the prognosis of cystic Kaposi's sarcoma to be relatively good with appropriate treatment. Besides interferon treatment, some authors consider accelerated electron beam therapy to be useful.

DISCUSSION

In this case, the clinical, histological, and laboratory findings led to the diagnosis of cystic manifestation of Kaposi's sarcoma. Numerous theories have been advanced as to the origin of the cysts, based upon clinical or modern electronmicroscopic studies, as well as investigations of enzyme, immunohistochemistry, or other special markers. Ronchese propounded the interpretation that congestive lymph cysts developed owing to mechanical obstruction of lymph vessels (1). Tedeschi considered colliquative necrosis inside lymph vessel tumors to be responsible for cyst formation (6). According to Palmer and Dictor, hemodynamic deviations observed in patients with Kaposi's sarcoma may result from aberrant connections between veins and lymph sinuses. These lymphatico-venous anastomoses may already be established during embryonic development.

Electron microscopic studies have demonstrated necrotic endothelial cells, fragmented basal membranes, and a lack of continuity in the capillary endothelium. The latter may explain the extravasation of erythrocytes.

A blood vessel or lymph vessel origin of Kaposi's sarcoma is being discussed to this day. Although many earlier authors contended that Kaposi's sarcoma is of blood vessel origin, the theory of lymphatic endothelial pathogenesis has become increasingly accepted lately (3,4). Using an immunohistochemical technique, Beckstead concluded that the phenotype of the spindle shaped cells in these lesions is more similar to lymph vessel endothelium than to that of blood vessels (2). Studies performed with special vessel markers have demonstrated that lymphatic endothelium is negative for the classic blood vessel endothelial markers ATP-ase, naphthylesterase, ALP,

succinate dehydrogenase, aminopeptidase, anti-laminin, anti-IV, type collagen. However, it appears significant that the lectin from Ulex europaeus binds to both lymph and blood vessel endothelium. Since tumor cells from Kaposi's sarcoma have been shown to react positively with UEA-I (Ulex europaeus lectin), but not with the above mentioned blood vessel endothelial markers, we tend to embrace the lymphatic endothelial origin for this tumor (7). This opinion would appear to gain credence from the observations made in the reported case, namely that lymphedema was the first symptom, followed later by development of cystic structures of a lymphatic nature and typical Kaposi's sarcoma lesions.

CONCLUSION

The purpose of this case report is to draw attention to the rare cystic manifestation of classic Kaposi's sarcoma (it being, to our knowledge, the first such case described in Hungary), with its implications pertaining to the pathogenesis of this disease, and the efficacy of combined interferon-retinol therapy

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