The lesions tend to resolve spontaneously after a mean period of 11 months and do not leave a scar.[3] In general, antibiotic therapy is ineffective, although some cases have shown a good response to oral macrolides or topical metronidazole.[7]

We must include IFAG in the differential diagnosis of acquired facial nodules in children. The medical history and the clinical, microbiologic, and ultrasound findings enable us to make an early diagnosis and to avoid unnecessary aggressive procedures.

Conflicts of Interest

The authors declare that they have no conflicts of interest.

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Lymphangioma-like Kaposi sarcoma☆

Sarcoma de Kaposi de tipo linfangiomatoso

To the Editor:

Lymphangioma-like Kaposi sarcoma (LLKS) is a rare histologic variant of Kaposi sarcoma that can present as any of the 4 known clinical variants. LLKS is a vascular neoplasm that develops secondary to infection by human herpesvirus type 8 (HHV-8), which is also known as the Kaposi sarcoma virus. Clinically, it can present with the usual manifestations, namely, patches, plaques, or nodules. However, in some cases, it presents as blisters that may be confused with bullous skin disease.

Case Description

The patient was an 80-year-old man whose history was unremarkable. He presented with raised erythematous, oval plaques measuring 1-3 cm in diameter that had first appeared 5 years previously. The plaques occasionally coalesced and were found on the upper and lower limbs and lower back. The lesions had gradually increased in number and size, although they were neither painful nor pruriginous. The physical examination revealed flaccid blisters (1 cm) containing serum (Fig. 1). Treatment with various topical options had been unsuccessful.

A complete laboratory workup including complete blood count and biochemistry revealed iron-deficiency anemia. Serology testing for HIV was negative.

Histopathology revealed that the epidermis was conserved and highlighted a proliferation of anastomosed vascular spaces that occupied the complete thickness of the dermis, dissected the collagen bundles, and surrounded cutaneous muscles and adnexa. No blood was identified in these structures. Clusters of lymphocytes and plasma cells were common in the stroma. Closer examination revealed that the vascular channels were lined with a layer of flattened endothelial cells and that there was no atypia or mitosis. Immunohistochemistry showed that tumor proliferation was positive for the endothelial markers CD31 and CD34 and for the lymphatic marker D2-40. It also showed clear nuclear staining for latent nuclear antigen 1 of HHV-8 (Fig. 2).

The patient was diagnosed with LLKS and referred to the medical oncology department. Given the poorly aggressive clinical course and the patient's age, it was decided—after

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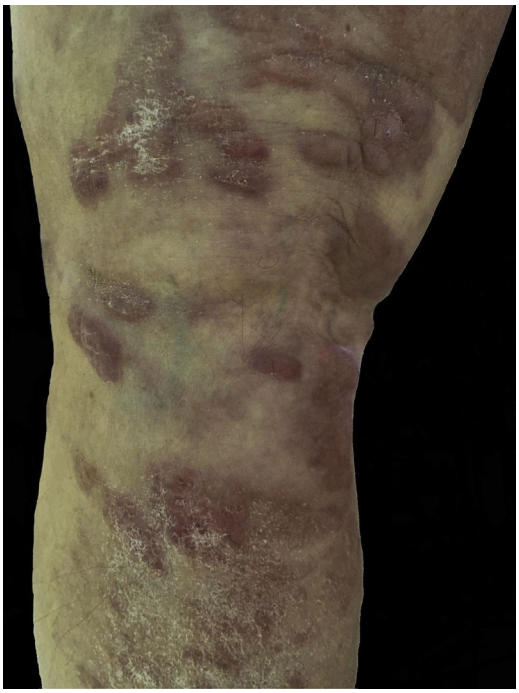

Figure 1 Coalescent erythematous-squamous plaques and blisters measuring 1 to 3 cm on the lower limbs.

evaluation and with the patient's agreement—to adopt a wait-and-see approach.

Discussion

LLKS is a malignant vascular neoplasm with evidence of lymph and blood vessel differentiation. It is a rare histologic variant of Kaposi sarcoma (5% of cases)[1] and can present in any of the 4 existing epidemiologic variants. Etiology and pathogenesis are controversial, as is the question of whether the disease should be classified as reactive or neoplastic.[2] The discovery of HHV-8, which is present in 100% of cases of Kaposi sarcoma, irrespective of the subtype in question, is a major finding in our knowledge of the etiology and pathogenesis of the condition.[3] Similarly, the question of cell differentiation is controversial, since cells express both specific blood and lymph markers.[4,5]

Clinical Manifestations

The most common presentation of LLKS is blisters,[6] although it can also appear in the more classic form of plaques or nodules. LLKS can also present as a mucocutaneous lesion, affecting mainly the legs and arms. The condition is more

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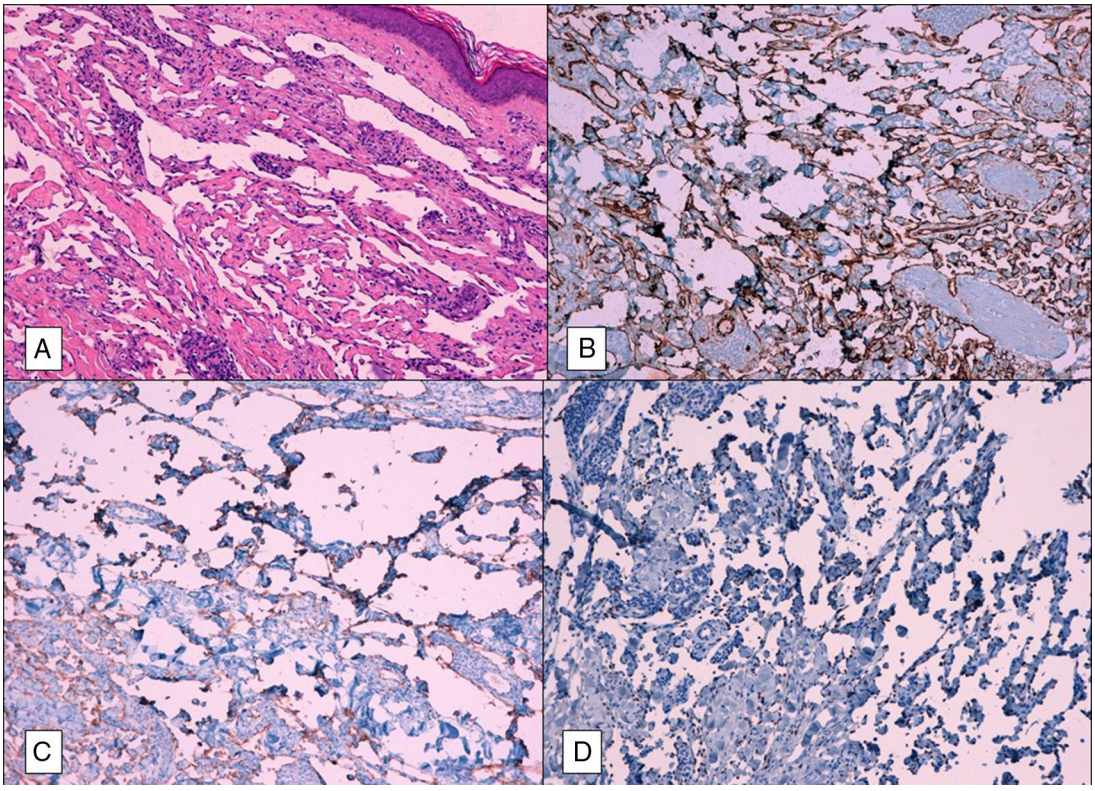

Figure 2 A, Anastomosed, small-caliber vascular channels with no content dissecting collagen bundles (hematoxylin-eosin, original magnification, ×10). B, Expression of CD34 (×10). C, Expression of D2-40 (×10). D, Expression of HHV-8 (×10).

Table 1 Lymphangioma-like Kaposi Sarcoma Affecting the Skin: Cases Published to Date.

Study, y	Clinical Type	Age	Sex	Clinical Lesions	Onset of Kaposi Sarcoma	Distribution of Lesions	Behavior
Gange and Jones, 1979	Classic KS	82	W	Blisters	2 y	Generalized	Slow progression
	Classic KS	55	M	Papules	3 y	Generalized	Slow progression
	Classic KS	72	M	Papules	10 y	Generalized	Slow progression
Leibowitz, 1980	Endemic KS	34	M	Blisters	Unknown	Generalized	Aggressive
Davis and Scott, 2000	KS-AIDS	35	M	Macules	5 y	Generalized	Slow progression
Noel et al., 1997	Endemic KS	42	M	Plaques	Unknown	Localized	Slow progression
Cossu et al., 1997	Classic KS	77	M	Blisters	8 y	Localized	Slow progression
	Classic KS	71	W	Blisters	2 mo	Generalized	Aggressive
	Classic KS	66	W	Blisters	3 y	Generalized	Slow progression
	Classic KS	80	M	Macules	2 mo	Localized	Slow progression
	Classic KS	75	M	Papules	5 y	Localized	Slow progression
	Classic KS	71	M	Papules	3 y	Generalized	Slow progression
	Classic KS	59	M	Macules	1 mo	Generalized	Slow progression
Roncheses and Kern, 1957)	Classic KS	68	M	Blisters	4 y	Generalized	Slow progression
	Classic KS	65	M	Blisters	Unknown	Generalized	Slow progression
Bossuyt et al., 1995)	KS-AIDS	57	M	Nodules	1 y	Generalized	Slow progression
De la Torre et al., 1998)	KS-AIDS	25	M	Nodule	Unknown	Generalized	Died 4 mo later
Ramírez et al., 2005)	Classic KS	83	W	Blisters	3 y	Legs	Slow progression
	KS-AIDS	38	M	Plaques and nodules	2 mo	Neck, chest, right arm	Slow progression
	Classic KS	75	W	Papules	1 y	Legs	Unknown
	KS-AIDS	40	M	Papules and plaques	3 y	Legs	Slow progression
Mohanna et al., 2006)	Classic KS	65	M	Blisters	8 y	Legs and arms	Slow progression
	Classic KS	89	W	Blisters and nodules	1 mo	Legs	Slow progression
	Classic KS	61	M	Plaques and nodules	1 y	Legs	Slow progression
	KS-AIDS	36	W	Plaques and nodules	7 mo	Legs	Died 2 mo later
Friedman et al., 2013)	Classic KS	85	M	Gangrene	2 wk	Right foot	Slow progression
Agustí-Mejías et al., 2011)	Classic KS	75	W	Plaque	8 y	Right leg and arm	Slow progression
Present case	Classic KS	80	M	Plaques and nodules	5 y	Legs and back	Slow progression

Abbreviation: KS; Kaposi sarcoma.

common in males, with a mean age at onset of 45.1 years (Table 1). Despite the low number of cases reported in the literature, data seem to indicate that LLKS is more indolent than the classic variety, although cases with an aggressive course have been reported.[7,8]

Histologically, the disease is characterized by ectatic vascular spaces with a labyrinthine architecture that dissect collagen bundles and dermal adnexa and are accompanied by a nodular lymphoplasmacytic infiltrate. The vascular structures are lined with flattened endothelial cells with no mitosis and no—or very scant—atypia. The cells of Kaposi sarcoma express endothelial cell markers (CD31 and CD34) and lymphatic markers (D2-40), as well as HHV-8.[9] The differential histologic diagnosis should be with other vascular tumors that show an infiltrative growth pattern between the collagen bundles or that comprises spindle cells, as is the case with low-grade angiosarcoma, kaposiform hemangioendothelioma, spindle cell hemangioma, hobnail angioma (targetoid hemosiderotic hemangioma), and benign lymphangioendothelioma.[10] The condition can also present as lymphangiectasia, although it is easy to detect this form with histopathology, which reveals dilation of pre-existing lymph vessels and absence of new formations, as occurs in the lymphangiomatous type.

Treatment

LLKS can be systemic or local, depending on the epidemiologic type, the patient's immune status, and the number of lesions. Local treatment can be carried out with liquid nitrogen–based cryotherapy, radiotherapy, laser, or intralesional injections with chemotherapy agents or interferon. Systemic treatment can be administered with interferon or chemotherapy agents.

Conflicts of Interest

The authors declare that they have no conflicts of interest.

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