Dear Editor,

Although sarcoid vasculitis has been characterized as vasculitis associated with systemic sarcoidosis in nomenclature of vasculitides proposed by Chapel Hill Conference in 2012 (1), sarcoid vasculitis in skin lesions is rarely documented in cases with either cutaneous or systemic sarcoidosis and therefore not specifically mentioned in disorders with cutaneous vasculitis (2). We herein report a case of systemic sarcoidosis, in which granulomatous vasculitis was observed in a biopsied specimen taken from erythema nodosum-like lesion on the thigh.

A 39-year-old man was diagnosed with lung sarcoidosis based on the findings of bilateral hilar lymphadenopathy on plain chest X-ray, small nodular shadows and mediastinal lymphadenopathy by computed tomography, and histopathological features of epithelioid cell granuloma on lymph node biopsy by bronchoscopy two years previously. Neither ophthalmologic nor cardiac involvement was observed. Serum levels of angiotensin-converting enzyme (ACE) began to increase one year previously. He noticed asymptomatic skin lesions on the lower extremities two months previously, and was referred to our department. Physical examination showed a number of infiltrated erythematous plaques with induration on the bilateral lower extremities (Fig. 1a). Histopathological examination revealed multiple non-caseating epithelioid granulomas in the mid-dermis and subcutis (Fig. 1b).

In the mid-dermis, findings of fibrinoid necrosis and destruction of vascular wall with infiltration of histiocytes were observed (Fig. 2a). A small vein at the dermal-subcutaneous junction was infiltrated by a number of histiocytes with fibrinoid necrosis (Fig. 2b). Higher magnification revealed vasculitis with vessel wall fibrinoid necrosis and angiocentric infiltrates of sarcoidal granulomas characterized by collections of CD68-positive histiocytes surrounding and infiltrating into the affected vascular wall (Fig. 2c,d), and Elastica van Gieson staining showed absence of internal elastic lamina of the affected small vein (Fig. 2e). By contrast, the adjacent counterpart

Figure 1. a) Multiple erythematous plaques with induration on the lower leg. b) Histological features showing non-caseating epithelioid cell granulomas with lymphocyte infiltration in the dermis and subcutis (×40).
small artery (arrow in Fig. 2b) remained intact without involvement of the sarcoideal granulomas infiltration. Higher magnification showed fibrinoid necrosis with a predominant infiltrate of mononuclear cells and around the vessel wall (×400). CD68 staining revealed an angiocentric infiltrate of CD68-positive histiocytes in and around the affected vessels (×400). b) Elastica van Gieson staining revealed absence of internal elastic lamina and loss of the elastic lamina of the involved vessel (×400).

Figure 2. a) Sarcoid granuloma with venulitis showing destruction of vascular wall and fibrinoid necrosis in the mid-dermis (×200). Granulomatous vasculitis at the dermal-subcutaneous junction is characterized by an angiocentric infiltrate of histiocytes and multi-nucleated giant cells in and around the affected venous vessel wall (×100). The adjacent counterpart small artery (arrow) remained intact without involvement of the sarcoideal granulomas infiltration. Higher magnification showed fibrinoid necrosis with a predominant infiltrate of mononuclear cells in and around the vessel wall (×400). CD68 staining revealed an angiocentric infiltrate of CD68-positive histiocytes in and around the affected vessels (×400). b) Elastica van Gieson staining revealed absence of internal elastic lamina and loss of the elastic lamina of the involved vessel (×400).

Vasculitis was observed in nearly 30% of patients (12/42), among whom venous involvement was observed in 11 patients (3). In a review by Yazdani Abyaneh et al. (4), granulomatous vasculitis in sarcoidosis is characterized by its association with chronic sarcoidosis, and clinical presentation with ulcers and livedo; while subcutaneous veins and arteries can be involved (4), and dermal venules are affected more often (3). Histopathology of sarcoid vasculitis in dermal or subcutaneous vessels were characterized by dense infiltration of sarcoideal granulomas cuffing around and in the affected vessel walls leading to disruption of vessels. Clinical features of sarcoid vasculitis have been reported presenting with ulcerative sarcoidosis (5), plaque-type sarcoidosis (6), and annular form sarcoidosis (7). This is the first report of sarcoid vasculitis presenting with erythema nodosum-like lesion.

References

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