Livedo Reticularis Associated with Atrial Myxoma

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We describe the case of a 45-year-old woman who had self-regressing livedo reticularis on the lower extremities. Examination of a biopsy specimen from the mottled area revealed myxomatous emboli in the deep dermal arterioles. Echocardiography showed a myxoma in the left atrium. After the tumor was surgically excised, the patient had no further evidence of the disorders during the 3-year period of follow-up. Livedo reticularis caused by an peripheral arterial embolism, which in turn was caused by the tumor fragments in our patient with left atrial myxoma, is considered to be unusual. (Ann Dermatol 10:(3) 159~162, 1998).

Key Words : Livedo reticularis, Atrial myxoma

Atrial myxoma is believed to be the most common primary tumor of the heart[1]. Approximately 75% of myxomas occur in the left atrium, and they are more common in women[1,2]. Signs and symptoms of cardiac dysfunction caused by the tumors almost always can be attributed to the presence of an atrial mass. Although pathologically these tumors are benign, clinically they can be considered malignant, especially when the tumors grow and cause mechanical effects that obstruct intracardiac blood flow, embolic effects, or both[1-3].

We describe a case of livedo reticularis in a patient with a left atrial myxoma.

CASE REPORT

A 45-year-old woman was admitted in March 1994 for investigation of recurrent episodes of developing mottled vascular patterns (livedo reticularis, LR) on the lower extremities. The initial sign/symptom occurred 5 months before hospitalization and the second attack developed 1 week before admission. Each time it seemed to be precipitated by physical exercise. She also experienced some short prodromal symptoms such as headache, shortness of breath, and sudden generalized weakness. However, she did not complain of any symptom relevant to heart or vascular system diseases. The major positive finding on physical examination in this patient was the occurrence of a dull-red mottled discoloration and slight swelling of the feet (Fig. 1), which were recognized just after the short prodrome. This discoloration and swelling occurred mostly at the same anatomical sites each time. She felt slight numbness/ fullness, and some tenderness in the affected parts. The color of this LR did not change by warming or cooling the areas. However it had a tendency of gradual dissolution over a period of 4 months. The patient was in good general health prior to the initial attack, and had no history of drug abuse, congenital heart disorders, or any systemic illness. Her family history was non-contributory.

A biopsy specimen showed that the small arteries in the lower dermis were filled with bluish mucinous material, with some endothelial proliferation, and the mucinous material reacted positively with alcian blue stain (pH 2.5) (Fig. 2). Serial sections revealed stellate-shaped or bipolar spindle stromal cells in myxomatous emboli.

Findings from laboratory studies, including a complete differential blood cell count and urinalysis were within normal limits. The pattern of serum protein electrophoresis, values representing liver/

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thyroid/ adrenal function, blood urea nitrogen and fasting blood sugar were all in normal ranges. Results on tests for antinuclear antibody, cryoglobulin, and latex-fixing rheumatoid factor were negative. However, an echocardiogram demonstrated the presence of a left atrial myxoma (Fig. 3).

With a provisional diagnosis of a left atrial myxoma, open heart surgery was performed; a left atrial tumor (2.0×3.0×1.0cm) with the stalk attached to the intra-atrial septum was excised. The histological appearances of the polypoid gelatinous tumor mass was consistent with a myxoma. It was composed of scattered polygonal stellate or spindle-shaped cells embedded in the abundant myxoma tissue. During a 3-year follow up, she did not have any further episodes of developing LR, and an echocardiogram revealed no evidence of tumor recurrence. The skin lesions of LR have persisted, however, but the color-intensity was somewhat faded.

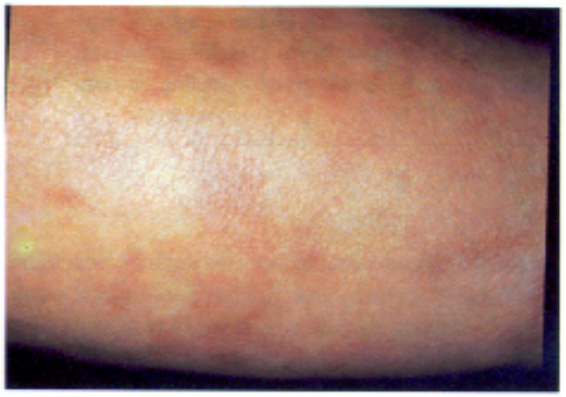

Fig. 1. Livedo reticularis on the lower leg.

DISCUSSION

In our patient we saw peculiar features of livedo reticularis which can be induced by various ways (Table 1). It was caused by arterial emboli in the dermis and consisted of myxomatous material. The diagnosis of left atrial myxoma in our patient was first suggested by the histological findings of the skin lesion and was confirmed by subsequent echocardiography. The concomitant prodromal symptoms were probably induced by a transient cerebral ischemia, pertaining to the mass effect of the tumor obstructing the mitral valvular orifice of the heart.

Peripheral, systemic emboli are frequently the initial manifestations of left atrial myxomas[4], and

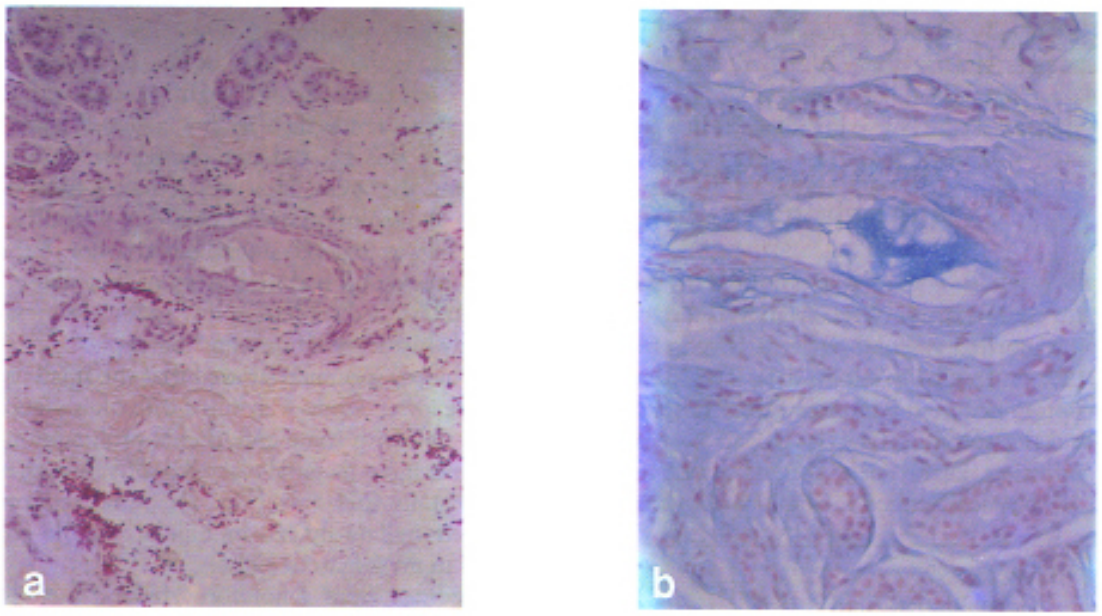

Fig. 2. (a), The biopsy specimen from a dull-red macule reveals a myxomatous embolus in the deep dermal arteriole (H&E, ×100).
(b), The myxoid matrix of the arteriolar embolus reacted positively with an acid mucopolysaccharide stain (Alcian blue pH 2.5, ×100).

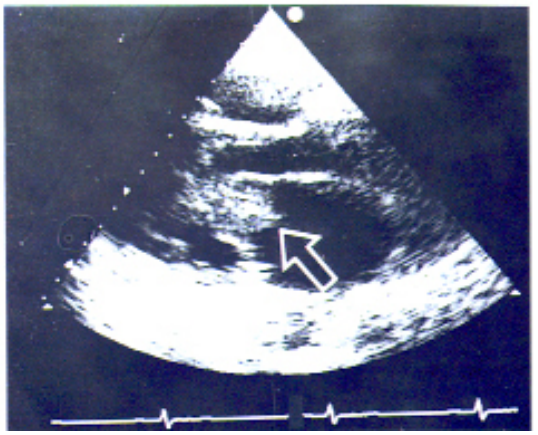

Fig. 3. Left atrial myxoma (echocardiogram; arrow).

have been estimated to occur in about 40% of patients who have left atrial myxoma caused by tumor fragments or surface clots that often obstruct arteries in the brain and extremities[1,5,6,7]. Repetitive trauma and the friable nature of the tumor frequently result in embolization. It is widely appreciated that such tumors may present with nonspecific skin lesions, appearing mostly on the extremities. Various skin lesions that seemed to have been caused by emboli and that have been described previously include erythema on the fingers and toes[8-12], cyanosis on the fingers and toes[8,9], necrosis of the toe[13], splinter hemorrhage[7,10], erythema and petechia on the hands and feet[7,9,11,12,14], Raynaud's phenomenon[13,15], mottling (not well defined) of the legs[14], and subcutaneous tumors[16].

The clinical diagnosis of atrial myxomas remains difficult. It depends on a high index of suspicion that will lead to cardiac evaluation such as echocardiography. Because the histologic benignancy of the tumor contrasts with its malignant clinical behavior concerning systemic embolization, early diagnosis is essential, and surgical excision will reduce mortality and morbidity from dysfunction of the atrioventricular valve and embolic complications to the vital organs. Clinicians will want to include atrial myxoma with peripheral embolization in their differential diagnosis of livedo reticularis, especially in young adults who have some history of atypical cardiovascular symptoms.

Table 1. Diseases associated with livedo reticularis

Connective tissue diseases	
systemic lupus erythematosus	Dermatomyositis
Rheumatoid arthritis	Scleroderma
Anti phospholipid syndrome	
Vasculitis	
Cutaneous angiitis	Livedoid vasculitis
Erythema elevatum diutinum	Polyarteritis nodosa
Cryoglobulinemic venulitis	Temporal arteritis
Drugs	
Drugs with actions on blood vessels	
Drugs causing immune complex vasculitis	
Infections	
Syphilis	Tuberculosis
Rheumatic fever	Meningococcemia
Pneumococcal sepsis	
Metabolic disorders	
Hypercalcemia	Hyperoxaluria
Neoplasms	
Mycosis fungoides	Pheochromocytoma
Atrial myxoma*	
Hematologic diseases	
Thrombocythemia	Polycythemia vera
Thrombotic thrombocytopenic purpura	
paraproteinemias	
Neurologic disorders	
Cerebrovascular accidents	Post traumatic states
Sneddon's syndrome	
Others	
Arteriosclerosis	Pancreatitis
Decompression sickness	Congenital disorders
Idiopathic disorders	

*The present case

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