

Erythematous Macules on the Feet in a Case of Cardiac Myxoma

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Sir,

Although cardiac myxoma is the most frequent benign cardiac tumour, it is still a rare disease: the incidence of the primary tumour is between 0.001% and 0.28% (1). Potential complications include sudden death, heart failure and the formation of emboli. The disease can sometimes be diagnosed based on the appearance of non-specific, cutaneous lesions. We present here a case of left-atrial cardiac myxoma in a 35-year-old woman with a circulating anticoagulant and recurrent painful macules on the soles of her feet.

CASE REPORT

A 35-year-old woman with a 6-month history of recurrent leg pain, face and leg swelling, and sole rash was seen at our unit. Her outbreaks occurred once a month, lasted one week, and left her generally impaired. She complained of fatigue and weight loss (6 kg in 6 months). History included three spontaneous abortions 10 years previously. Physical examination was normal. Laboratory studies showed inflammatory syndrome, with an erythrocyte sedimentation rate (ESR) of 22 mm in the first hour and 47 mm in the second hour, slight anaemia (11.6 g/dl), and an increased activated cephalin time (CAT) of 43 s (normal value 34 sec). There was a circulating anticoagulant that was antiphospholipid dependant but non-anti-cardiolipid, and could not be identified more specifically. No other antibody was found. A previous biopsy of an erythematous macule had revealed an intravascular thrombus (Fig. 1A). In view of her history and clinical symptoms, we suspected that she had an autoimmune disease, but we could not make any more definitive diagnosis. We asked her to come back at her next outbreak of symptoms. Two months later she returned, presenting with skin lesions, leg pain, difficulties in moving, and basithoracic pain that was improved by deep breathing. Physical examination revealed

erythematous to purpuric, painful macules and papules, 1–3 mm in diameter, on her soles (Fig. 1B). The remainder of the cutaneous examination was normal, with no evidence of cutaneous myxoma, and no lentiginous or blue naevi. Cardiac and pulmonary auscultation was normal. Laboratory studies were similar to those previously described. Electrocardiogram and chest radiograph were normal. She was not given any medication. Echocardiography revealed a very large tumour, 46 × 37 mm, in the left atrium. Myocardial computed tomography confirmed the presence of a mobile, non-obstructive, atrial myxoma that extended from the inter-atrial septum. Abdominal computed tomography revealed splenic and renal infarcts, which provided an explanation for the basithoracic pain. She underwent urgent heart surgery, during which a voluminous, polypoid, myxoid tumour was removed. No sign of malignancy was found on histological examination of the tumour. The intravascular thrombus seen on the cutaneous biopsy was retrospectively demonstrated to be an embolus originating from the cardiac myxoma.

Immunohistochemical examination of the emboli originating from the myxoma showed them to be expressing the endothelial marker anti-CD31. After surgery, the patient has remained free of symptoms.

DISCUSSION

Atrial myxoma is the most frequent benign cardiac tumour, representing approximately 50–70% of cardiac primary tumours. The tumour usually manifests itself between the third and the sixth decade of life, and occurs mostly in women. Cardiac myxoma is most commonly found in the left atrium, particularly on the inter-atrial septum, and is derived from embryonic remnants. Histological findings show myxoid tumours that are more or less polypoid, whose stroma is rich in acid mucopolysaccharides and surrounded by poly-

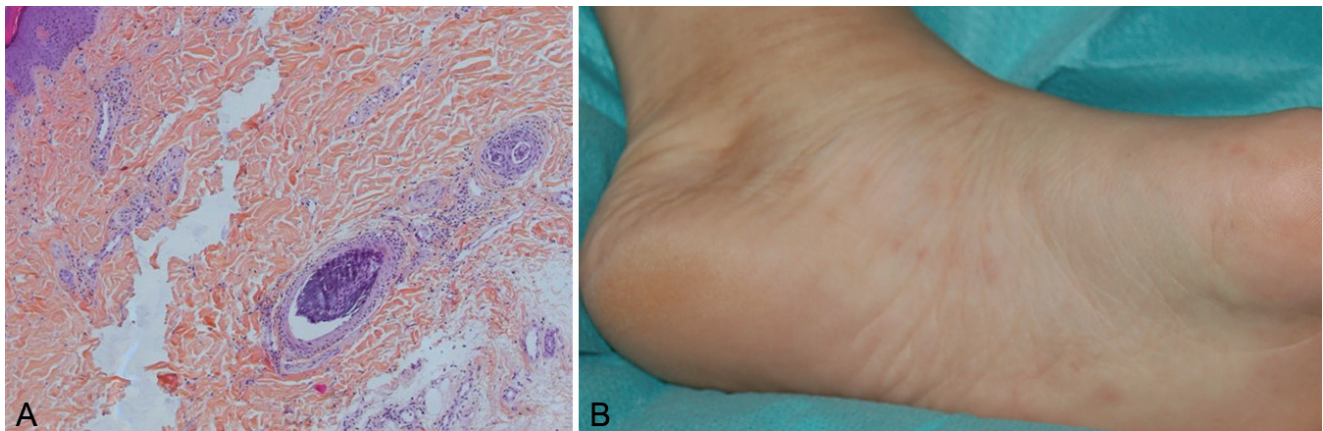


Fig. 1. (A) Histological analysis of a cutaneous biopsy of an erythematous macule of the sole showing an intravascular thrombus in a dermal vessel. (B) Recurrent erythematous plantar macules due to myxoid emboli.

gonal or stellate cells. The stroma often stains positive to Alcian blue, and endothelial markers such as CD31 are widely present (2). The tumour may manifest itself with a myriad of signs and symptoms, such as general disturbances, fatigue, fever, weight loss, or arthralgia. These signs mimic infection, or autoimmune or malignant disease. More specific cardiac symptoms, such as murmur, arrhythmia, endocarditis, or heart failure, can aid the diagnosis. Cutaneous manifestations, although non-specific, are often useful diagnostic clues. They are classified as one of two types (1). First, cutaneous signs can be "due to myxoid emboli": acral erythematous papular eruption (1, 3, 4), rarely on the neck (5) or on the trunk with a necrotic evolution (6), ulceration, or necrosis of the extremities (7), splinter haemorrhage of the nails (7), livedo reticularis (8), digital ischaemia (2). The second classification is "related to auto-immune phenomena": Raynaud's phenomenon (9), malar erythematous eruption (10), or knee eruption (11). These signs not only mimic autoimmune disease, but can also respond to corticosteroids (11, 12), frequently leading to a delay in diagnosis (mean about 9 months, range 1 week to 180 months) (13). Physical examination can disclose specific signs, including blue naevi, lentiginos, or fibromyxoid tumour. These signs, associated with endocrinopathy, are characteristic for the Carney complex, a rare genetic disorder that is inherited as an autosomal dominant trait. It has been mapped to 2p16 (CNC2) and 17q22-24 (CNC1) (14) and was initially referred to by the acronyms NAME or LAMB. Emboli occur in approximately 30–50% of the cases, most often into the brain. These phenomena are more frequent if the tumour is polypoid. Infrequently, the entire tumour can embolize, resulting in acute lower extremity ischaemia. In adults, emboli and constitutional symptoms are most common, while emboli and valvular obstruction predominate in paediatric patients (1). Laboratory studies frequently indicate inflammatory syndrome, production of interleukin-6, or, rarely, autoantibodies. Antinuclear antibodies seem to be the most frequent auto-antibody (9). There appears to have been only a single report of circulating antiphospholipid antibody (6). Each of these anomalies typically disappears after surgical resection of the tumour (15). Skin biopsy is rarely performed. It can be unremarkable or can mimic an autoimmune disease, exhibiting a vasculitis or a positive immunofluorescence staining (positive lupus band test) (6). It can also reveal an embolus within a dermal vessel (2–4). The demonstration of myxomatous emboli in a vessel can be difficult; therefore, repeating the biopsy or examining serial sections may be useful (8). Diagnosis is confirmed by echocardiography, and

emergency surgery is carried out in order to remove the myxoma. Recurrent tumours may appear if surgery is incomplete. In summary, in the case presented here, the presence of erythematous to purpuric macules on the extremities, mimicking vasculitis, along with the history of spontaneous abortions and the presence of circulating anticoagulant, initially delayed the diagnosis. In retrospect, the three spontaneous abortions do not seem to have been related to the primary disease or to the autoantibodies. Dermatologists should therefore be aware of the diagnosis of cardiac myxoma.

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