

## Left Atrial Myxoma Presenting with Migratory Erythematous Maculopapules

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Cardiac myxoma is the most common primary tumor of the heart. Although skin symptoms in patients with cardiac myxoma are common, cutaneous manifestations of cardiac myxoma are usually nonspecific. So only very rarely have the cutaneous signs been clue to the correct diagnosis. We present a 44-year-old-man with a history of episodes of migratory acral erythematous maculopapular lesions on his right hand and foot including the palm and sole, who also had episodes of myalgia in his right side upper and lower extremities. Histological examination revealed an embolus within the vessels in the dermis. Alcian blue staining was positive, consistent with a diagnosis of myxomatous embolus. Two-dimensional echocardiogram revealed a 4 (3 cm-sized mobile mass in the left atrium. A diagnosis of left atrial myxoma was strongly suspected and the tumor was excised by open-heart surgery. No more cutaneous lesions or other symptoms of systemic embolism have appeared to date.

(Ann Dermatol (Seoul) 19(3) 115~118, 2007)

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*Key Words:* Atrial myxoma, Cutaneous embolism

### INTRODUCTION

Cardiac myxoma is the most common primary tumor of the heart<sup>1</sup>. Most cases occur in the left atrium during the third to sixth decade of life. Classically, the presentations of cardiac myxomas include obstructive, embolic, and constitutional symptoms and signs<sup>2</sup>. We report a case of atrial myxoma associated with migratory maculopapules, representing an embolic phenomenon without cardiac symptoms and showing the histological findings of intravascular myxomatous material in the dermal vessels, a diagnostic clue to the presence of atrial myxoma.

To our knowledge, there has not been any repo-

rted case in the Korean dermatologic literature, to describe left atrial myxoma presenting with migratory erythematous maculopapules. The histologic features of such lesions should alert the clinician to the presence of an otherwise silent atrial myxoma.

### CASE REPORT

A 44-year-old man presented with a week history of tender, erythematous maculopapules on his right foot. He had noted three episodes of acral maculopapules in the previous 6 months, mainly on his right hand including the palm, which had resolved within 2-3 weeks without treatment. The most recent cutaneous lesions had appeared 1 month ago and the lesions had resolved spontaneously within 2 weeks. Additionally, the patient had a 4-month history of intermittent episodes of myalgia in the right upper and lower extremities.

Physical examination revealed multiple tender, violaceous, erythematous maculopapules on the sole and side of the right foot (Fig. 1). Routine laboratory analyses, serology for treponematosi (TPHA,

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Received May 9, 2006

Accepted for publication May 18, 2007

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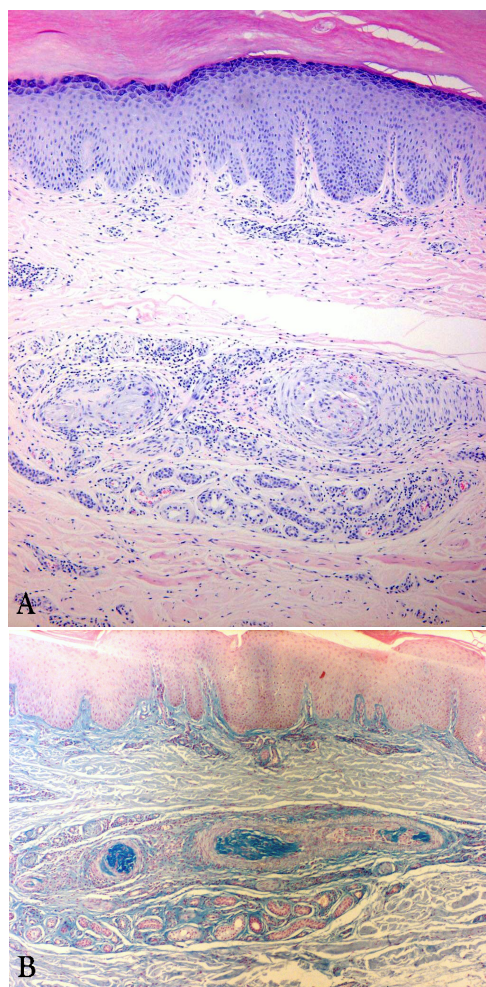
**Fig. 1.** Tender, blanching, erythematous maculopapules on the side and sole of the right foot.

VDRL), and muscle enzymes were within normal limits. He denied any history of fever, chills, arthralgias, weight loss, dyspnea, syncope, palpitation, or cardiac symptoms. A skin biopsy was taken from a tender papule on the sole. Histopathologic examination revealed no abnormality in the epidermis. But in the dermis, there were vessels occluded by hypocellular myxoid material (Fig. 2A). The myxoid intravascular material reacted positively with alcian blue stain at PH 2.5 (Fig. 2B). Based upon these findings, embolization by cardiac myxoma was suspected and an echocardiogram was performed. It demonstrated a 4 × 3 cm-sized mobile mass in the left atrium.

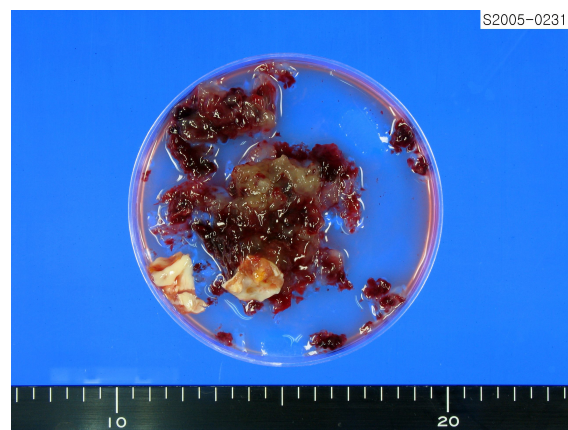
Open-heart surgery was performed for mass excision (Fig. 3). Pathologic examination confirmed a diagnosis of myxoma. Following surgery, the tender erythematous maculopapules and myalgia disappeared rapidly.

## DISCUSSION

Cardiac myxomas are the most prevalent primary cardiac tumors, although they are rare<sup>1</sup>. While they arise in any of the four heart chambers, or rarely on heart valves, about 90% are located in the atria, with a left-to-right ratio of approximately 4 : 1. The common clinical manifestations of cardiac myxomas have included obstructive, embolic and constitutional symptoms and signs such as fever and malaise. Obstruction of intracardiac blood flow by a left atrial myxoma may cause congestive heart failure, chest



**Fig. 2.** (A) Amorphous intravascular material occluding the vessels in the dermis (H & E, × 100). (B) Myxoid intravascular deposit reacted positively with alcian blue stain at PH 2.5 (Alcian blue, × 100).



**Fig. 3.** The cardiac myxoma was removed.

pain, syncope, or a murmur<sup>2</sup>. Constitutional symptoms have been estimated to occur in 90% of patients with cardiac myxoma<sup>2</sup> and were characterized by fever, malaise, arthralgia, weight loss, anemia, elevated ESR, and elevated gamma globulin. Several studies demonstrate a correlation between interleukin-6 production by cardiac myxomas and systemic inflammatory findings<sup>3</sup>. Embolism occurs in 20% to 45% of patients with cardiac myxoma, and approximately half of all myxomatous emboli from the left cardiac myxoma go to the brain<sup>4</sup>. Tumor embolization caused by left atrial myxomas may present the first symptomatic manifestation, which may be multiple and massive. The onset of a neurological deficit may be gradual or sudden. The extremities are the next most common sites of embolic phenomenon. Our patient had migratory maculopapules as the sole manifestation without any cardiac or neurological symptoms.

Cutaneous manifestations of cardiac myxoma are usually nonspecific. They may be due to embolism from the myxoma, but there are also other nonembolic cutaneous signs. An association between nonembolic cutaneous lesions and cardiac myxomas was first reported in a patient with prominent facial lentiginosis and a left atrial myxoma<sup>5</sup>. Subsequent reports have described the NAME syndrome (*nevus, atrial myxoma, myxoid neurofibromas, and ephelides*)<sup>6</sup> and the LAMB syndrome (*lentigines, atrial myxoma, blue nevi*)<sup>7</sup>. Approximately 10% of patients with myxoma have a familial cardiac myxoma syndrome, known as Carney syndrome, characterized by autosomal dominant transmission, multiple cardiac and often extracardiac myxomas, spotty pigmentation, and endocrine overactivity<sup>8</sup>. Our patient had no familial history of cardiac myxoma and had a small number of lentigines on the face, fewer than the number described in reported cases<sup>6-8</sup>, so a diagnosis of LAMB syndrome or Carney syndrome could not be established.

The cutaneous manifestations of emboli from atrial myxomas include erythematous macules and papules mainly acral in location, digital cyanosis, petechiae, splinter hemorrhage, telangiectasia, livedo reticularis, Raynaud's phenomenon, ulcerating lesions, and a reddish-violet malar flush<sup>9</sup>. In our patient, the pattern of the cutaneous findings was similar to that described by Garcia-F-Villalta et al<sup>10</sup>. In their report, the patient had noted five or six episodes of acral papular erythematous lesions mainly on the

lower extremities and ischemic neurological events with disorientation, dysarthria, and right arm weakness. There were some other reports showing erythematous maculopapules with neurologic manifestations in the English literature<sup>9,11,12</sup>. Al-Mateen et al.<sup>13</sup> also described two children with cerebrovascular events caused by emboli from left atrial myxomas. They described transient cutaneous eruptions involving the extremities as 'red spots'. The red spots were noted before the onset of the cerebral ischemic event<sup>13</sup>, therefore cutaneous findings would be an important clue in prompt diagnosis and early intervention.

Because there is a broad spectrum of cutaneous findings in patients with cardiac myxoma and skin lesions may be nonspecific, the diagnosis of cardiac myxoma may be difficult, especially when cardiac symptoms are not present. However, cutaneous findings might be important clues to the diagnosis as in our case, so the dermatologist must first be aware of the multitude of cutaneous findings associated with this tumor.

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