Cutaneous Manifestations Revealing Cardiac Myxoma: A Case Report and Literature Review

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ABSTRACT
The clinical manifestations of cardiac myxoma are often non-specific and may delay diagnosis. We report the case of a 53-year-old man who has been presenting for the past four years a recurrent skin rash on his back associated with muscle pain. He was admitted to the emergency room for sudden right-sided hemiparesis and aphasia due to a subacute ischemic stroke at the frontal-parietal and temporal lobes. Echocardiogram revealed a mass in the left atrium suggestive of myxoma. After surgery, the patient remained free of cutaneous symptoms. Atypical manifestations of cardiac myxoma should be considered in order to not delay adequate management.

Introduction
Cardiac myxomas are non-cancerous primary tumors of the heart, most frequently localized in the left atrium (75%), and constitute about 50% of all primary heart tumors. The average age of onset is the sixth decade of life.

Common clinical findings include obstructive, embolic and constitutional symptoms, and can be uncharacteristic. There are many extra cardiac manifestations associated with atrial myxoma that can bring confusion with other medical conditions, leading to diagnosis and treatment delays, and ultimately to a poor prognosis. Therefore, an accurate diagnosis requires a high index of suspicion and a certain familiarity with the varied modes of presentation.

The diagnostic method of choice is trans-thoracic echocardiography, with a sensitivity of 95% (1, 2, 3).

Patients usually have a favorable prognosis after resection.

This article reports a case of left atrial myxoma presenting initially with cutaneous manifestations leading to a delayed diagnosis of the cardiac tumor at the stage of complications.

Case report
We report the case of a 53-year-old man without modifiable cardiovascular risk factors. His medical history indicated a recurrent skin rash on the back and muscle pain during the last four years, of unknown etiology.

On the cardiovascular level, the patient reported an atypical chest pain with unexplored palpitations of abrupt onset and termination, without the notions of lipothyemia, discomfort or syncope.

The patient was admitted to the emergency room on the 28th of August, 2018 for sudden right-sided hemiparesis and aphasia due to a subacute ischemic stroke at the left frontal-parietal and temporal lobes.

Physical examination on admission revealed a good general condition, an incoherent verbal response, and a residual right-sided weakness (muscle strength rated at 4/5 on the right arm and leg). The blood pressure was 126/73 mmHg with an irregular heart rate of approximately 80 bpm. Heart sounds were clear, moderately loud, without murmurs. Skin examination showed maculopapular rash on the back (fig.1).

Figure 1. Recurrent maculopapular lesions due to myxoid emboli.

ECG recorded a sinus rhythm with supraventricular extrasystoles.

In the framework of establishing the etiology of the stroke, a carotid doppler was performed which did not reveal any abnormality. However, a trans-thoracic ultrasound showed a large, highly mobile, non-obstructive mass of heterogeneous echogenicity, in the left atrium, measuring 3.5 - 4.5 cm in diameter, extending from the inter-atrial septum (confirmed by transesophageal echocardiography) (fig.2,3).

Heart chambers were not dilated. Wall thickness and myocardial contractility of the left ventricle were normal.

Heart valves were without significant changes. There was no fluid in the pericardial cavity.
Cardiac myxoma remains the most common intracardiac tumor. It is an intracavitary, polypoid tumor that grows from a sessile base to the anterior atrial septum, which explains the clear predominance of atrial myxomas. It is characterized by a clinical polymorphism that can be confusing for the clinician, and by a complete and definitive surgical curability at the price of delayed diagnosis which can take months or even years.

Clinical symptoms are determined by the tumor’s location. Myxoma of the left atrium may have cardiac, general, neurological or arterial expressions.

Cardiac manifestations may include signs of paroxysmal mitral stenosis or regurgitation, positional syncope, palpitations, dyspnea, chest pain or heart failure.

It is impossible to mention all the general events reported in the literature that were eventually attributed to myxoma. As for the arterial signs, they are dominated by emboli either of thrombotic material detached from the surface of the tumor or of the myxoma itself in its villous forms or of the myxoma as a whole. Neurological complications considerably worsen the prognosis of myxoma as reported in the Roeltgen series (4), which contrasts with the excellent prognosis of myxoma operated on without neurological complications.

Cutaneous signs can be due to myxoid emboli: acral erythematous papular eruption (5, 6, 7), ulceration, or necrosis of the extremities (8), splinter haemorrhage of the nails (8), livedo reticularis (9), digital ischemia (10). As they may be related to auto-immune phenomena: Raynaud’s phenomenon, malar erythematous eruption, or knee eruption.

The key to diagnosis remains echocardiography, which identifies the size, site, attachment, mobility, and also grossly differentiates the myxoma from a vegetation or a thrombus.

Surgical excision is the treatment of choice which aims at complete and definitive resection, and has excellent outcomes.

In our patient, the non-specific nature of the cutaneous manifestations delayed the diagnosis which was only made at a stage of neurological complications. He underwent urgent heart surgery, with disappearance of the skin lesions.

Conclusion

Skin manifestations are rare in patients with cardiac myxoma. In such situation, the diagnosis can be challenging and the proper treatment delayed. Therefore, dermatologists should be aware of the possibility of cardiac myxoma in these cases, before reaching the stage of complications.

References