Skin Ulceration Due to Metastatic Emboli from Ventricular Myxoma

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We report a case of left ventricular myxoma metastasizing to blood vessels of extremities and causing cutaneous ulceration. Most of left ventricular myxomas are symptomatic (92.7%). Systemic embolism is the most common manifestation (50%) and often leads to death.3 The propensity of cardiac myxomas to embolize is thought to be related to tumor morphology, with friable and gelatinous myxomas being more likely to embolize than firm and fibrous lesions.26, 27 The current case is rare in the sense that the myxomatous material had metastasized to blood vessels of hands and legs, causing arterial insufficiency and hence cutaneous ulceration.

Key words: ........................................

Introduction
Cardiac myxoma is a benign neoplasm that represents the most common primary tumor of the heart. Myxoma typically manifests as a polypoid, intracavitary left atrial mass that arises from the interatrial septum, but it may originate in any cardiac chamber.1, 2 There have been 47 cases of LV myxoma reported in the world literature since 1957. Most of them are symptomatic (92.7%). Systemic embolism is the most common manifestation (50%) and often leads to death.3

Cutaneous manifestations are frequently reported and can correspond to cutaneous manifestations of emboli, symptoms related to autoimmune disorders and specific cutaneous findings that suggest that the lesion is part of more complex syndromes.4 We present here a case of ventricular myxoma, with the patient presenting with cutaneous ulceration secondary to metastatic emboli from ventricular myxoma.

Case Report
A 24-year-old male presenting with dyspnoea and dry cough for one month. Patient also had non-healing ulcers on hands and feet for the same duration. Patient had low grade, intermittent fever for 1 week and weakness and pain of left leg for 1 day. Patient was a known smoker for 5 years. On examination his vital signs were stable. Clubbing was positive. Examination of cardiovascular system revealed gallop rhythm and a systolic murmur. There was non-tender hepatomegaly with liver being 3 cm below the costal margin. Touch and proprioception sensations were absent in left lower limb. Also reflexes and power were decreased in left leg. Local examination revealed non-healing, punched out ulcers in both hands and feet in various stages of healing including some with granulation tissue (Figure: 1). TLC was 16100/μL with 75% neutrophils. ESR was 39. ECG showed right atrial deviation and right bundle branch block. Doppler ultrasound of left lower limb was consistent with thromboembolic process involving arterial system of left lower limb up to the level of junction of common femoral and superficial femoral arteries. Doppler ultrasound study of venous system of left lower limb was normal. No evidence of DVT was found. Echocardiography revealed a myxoma measuring 9.3 cm in left ventricular cavity arising from interventricular septum (Figure: 2). Patient was started cap Ampicillin 500mg, inj heparin 5000U s/c TDS, tab chymotrypsin TDS, tab loprin OD, tab ofloid 400mg BD, tab flagyl 400 mg TDS. Skin biopsy from ulcerated area was performed and on histopathology a diagnosis of epidermal necrosis due to metastatic emboli from ventricular myxoma was rendered. After 8-9 days of hospital stay the echo was again repeated and it showed that the myxoma had reduced in size to a dangling, fragile myxomatous tissu in left ventricular cavity further predisposing the patient to thromboembolism(Figure 3). Currently, patient is symptom free. The ulcers on
the hands and feet are in healing phase.

Pathologic Findings

The specimen submitted for histopathology comprised of a single elliptical fragment of skin measuring 0.8x 0.3x 0.2 cm.

Microscopy revealed skin fragment composed of epidermis and dermis. Epidermis showed hyperkeratosis, acanthosis, elongation of rete ridges and focal ulceration. Dermis revealed dense inflammatory infiltrate. Myxomatous, foamy material was seen within the small blood vessels that in some places was seeping out into the dermis from damaged vessel wall and attracting eosinophils. (Figures: 4, 5, 6, and 7)

Comment

Primary tumors of the heart are rare, with an incidence between 0.0017 and 0.19 percent in unselected patients at autopsy. Three quarters of the tumors are benign. Nearly half the benign heart tumors are myxomas, and the majority of the rest are lipomas, papillary fibroelastomas, and rhabdomyomas. A left atrial myxoma was first described in 1845. Before 1951, the diagnosis of intracardiac tumors was made only at postmortem examination; in that year the diagnosis of an intracavitary left atrial tumor was confirmed by angiocardiography. The first successful excision of a left atrial myxoma was reported in 1955. The introduction of echocardiography has greatly facilitated the antemortem diagnosis of cardiac tumors. Myxomas occur in all age groups but are particularly frequent between the third and sixth decades of life. Women predominate in most series. “Complex” cardiac myxomas are those that occur in families, in combination with two or more of the following conditions: skin myxomas (single or multiple), cutaneous lentiginosis, myxoid fibroadenomas of the breast, pituitary adenomas, primary adrenocortical micronodular dysplasia with Cushing's syndrome, and testicular tumors (characteristically, large-cell calcifying Sertoli-cell tumors). At the time of the diagnosis, patients with familial myxomas are usually considerably younger than those with nonfamilial myxomas. Family studies suggest an autosomal dominant pattern of inheritance with a variable phenotype. Cardiac myxomas usually develop in the atria. About
75 percent originate in the left atrium, and 15 to 20 percent in the right atrium. Most myxomas arise from the interatrial septum at the border of the fossa ovalis, but they can also originate, in descending order of frequency, from the posterior atrial wall, the anterior atrial wall, and the atrial appendage. Only 3 to 4 percent of myxomas are detected in the left ventricle, and only 3 to 4 percent in the right.  

Figure 5: Myxomatous Material within Blood Vessel with Surrounding Eosinophilic Reaction. H & E (X200)  

Figure 6: Myxomatous Material within Damaged Blood Vessel. H & E (X 100).  

Figure 7: Two Eosinophils in the Blood Vessel Wall. H & E (X 400)  

Left atrial and left ventricular tumors can present a variety of signs and symptoms that include, but are not restricted to, fever,
chills, dizziness, dyspnea on exertion, cold sweats during exercise or at night, and nonproductive cough. Since tumors may embolize, they also can lead to seizures, transient ischemic attacks, and cerebrovascular and peripheral-vascular accidents.

Myxomas are neoplasms of endocardial origin. The cells giving rise to the tumor are considered to be multipotential mesenchymal cells that persist as embryonal residues during septation of the heart and differentiate into endothelial cells, smooth-muscle cells, angioblasts, fibroblasts, cartilage cells, and myoblasts. Myxomas are generally polyloid, often pedunculated, rarely sessile, and round or oval, with a smooth or gently lobulated surface. The tumors range from 1 to 15 cm in diameter; most are 5 to 6 cm. Microscopically, myxomas consist of a myxoid matrix composed of an acid-mucopolysaccharide–rich stroma. Polygonal cells with scant eosinophilic cytoplasm are scattered throughout the matrix. The cells are arranged singly or in small clusters and may be multinuclear, but mitoses are not found. These cells also form capillary-like channels that occasionally communicate with the surface of the myxoma. Large blood vessels – arteries and veins – are abundant at the base of the myxoma and are derived from the subendocardium.

With the advent of echocardiography, the diagnosis of cardiac myxoma has become very easy with the technique displaying dense array of wavy tumor echoes. The treatment for cardiac myxoma is urgent surgical resection. The stalk is resected along with the entire zone of attachment. Surgical mortality is very low and the long-term prognosis is excellent.

**Conclusion**

Although cardiac myxomas are histologically benign, they may be lethal because of their strategic position. They can mimic not only every cardiac disease but also infective, immunologic, and malignant processes. Myxomas must therefore be included in the differential diagnosis of valvular heart disease, cardiac insufficiency, cardiomegaly, bacterial endocarditis, disturbances of ventricular and supraventricular rhythm, syncope, and systemic or pulmonary embolism. The symptoms depend on the size, mobility, and location of the tumor. Surgical removal of the tumor should be performed as soon as possible; the long-term prognosis is excellent, and recurrences are rare. In follow-up examinations as well, echocardiography is essential.

**References**

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