



Case report

The sentinel rash and neurocutaneous presentation of atrial myxoma: Case report and literature review

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ABSTRACT

Introduction: Presenting symptoms of atrial myxoma are classically viewed as a triad of cardiac, embolic, and constitutional symptoms. Dermatologic manifestations are viewed as constitutional symptoms and have not received much attention.

Methods: Illustrative case presentation and literature review.

Results: I admitted a 28 y woman transferred to the comprehensive stroke center with an ischemic stroke treated with intravenous thrombolysis. She had a two year history of skin lesions in the hands/fingers and feet/toes/sole. Skin biopsies showed perivascular neutrophilic inflammatory dermatitis without skin vasculitis. Admission physical exam was notable for chronic skin changes on her hand and foot and an acute erythematous petechial rash over the right side of her face in the distribution of the right external carotid artery. MRI brain showed ischemia involving the right internal carotid artery territory. Transthoracic echocardiogram identified a 2.4 cm atrial myxoma with normal mitral valve function. She underwent uncomplicated cardiac surgery with confirmation of a villous myxoma. At her 8 week follow up visit, she had resolution of her neurologic deficits, and her skin rashes had cleared.

Literature review identified 17 additional published cases of sentinel rashes preceding diagnosis of atrial myxoma. Skin changes were often reported in the hand and feet and may involve the digits, palm, and sole. In 8 cases, skin biopsy identified myxomatous emboli in dermal vessels. These cutaneous manifestations share similarities with Osler nodes and Janeway lesions first described in the 1890's, now understood to represent microemboli to the dermis. Seven of these 17 cases (41 %) developed strokes after the initial presentation of cutaneous changes. Our patient had an embolic event primarily affecting the right common carotid artery territory with facial cutaneous changes due to emboli to the external carotid artery territory and cerebral ischemia in the right internal carotid artery territory.

Conclusion: I recommend reclassification of the cutaneous signs of atrial myxoma as embolic (to the dermis microvasculature) rather than a constitutional symptom. As the skin biopsy often fails to show myxomatous emboli, physicians should have a high suspicion for a proximal embolic source for patients with a migratory rash preferentially affecting distal extremities. Earlier diagnosis of atrial myxoma when the sentinel rash first appears provides a valuable window of opportunity for cardiac surgery before the patient experiences stroke or peripheral emboli.

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1. Introduction

Presenting symptoms for patients diagnosed with sporadic atrial myxomas have been carefully studied [1,2]. In the largest case series (n = 112) collected over a 40 year interval (1959–1998), Pinede et al. describe the classic triad of symptoms. Most patients had one or two symptoms from this triad. The most frequent was cardiac symptoms (67 %) caused by prolapse of the atrial myxoma through the mitral valve. This led to syncope, palpitations, pulmonary edema, and congestive heart failure. The second most frequent symptom was cardiac emboli (29 %) causing strokes or limb ischemia. About two-thirds of myxomas have a smooth appearance and one-third are friable and villous. Embolic risk is more than doubled with villous myxomas (45 %) compared to smooth myxomas (19 %). The third symptom (34 %) was termed constitutional signs, and included less specific symptoms such as myalgia, arthralgia, fatigue, and Raynaud syndrome. With the growth of non-invasive cardiac testing such as echocardiography, atrial myxomas are now often diagnosed prior to any clinical symptoms.

I focus on the cutaneous manifestations of atrial myxoma through an illustrative case and literature review. As skin changes may precede stroke in patients diagnosed with atrial myxoma, I emphasize that this is a ‘sentinel rash’ and present data supporting the view that skin changes are due to dermal microemboli. Early diagnosis of atrial myxoma in patients presenting with cutaneous manifestations provides a valuable window of opportunity for cardiac surgery, before the patient suffers stroke or limb ischemia.

2. Case presentation

A 28 y woman (Fig. 1) had a two year history of migratory skin lesions described as pink reticulated patches and macules involving dorsal and palmar hands and fading skin changes from a prior similar eruption in her feet which involved the toes and sole. Skin biopsy on two separate visits showed perivascular neutrophilic inflammatory dermatitis without skin vasculitis. Laboratory testing for autoimmune disease markers was negative. Her rash persisted after trials of colchicine and steroids.

She presented to the emergency department of an outside hospital with sudden onset of left sided weakness involving her face, arm, and leg and a National Institute of Health Stroke Scale score of 5. CT head imaging did not show any acute abnormalities, and intravenous tenecteplase was administered. She was then transferred to our comprehensive stroke center for ongoing stroke treatment. She had no prior symptoms of palpitations, syncope, orthopnea, chest pain, or peripheral edema. She had no additional medical history and no family history of autoimmune disease, early onset stroke or cardiac disease, and no thrombophilias. Vital signs were all within normal limits. Her admission physical exam was notable for chronic skin changes on her right hand and foot and an acute erythematous petechial rash over the right side of her face. No murmur was detected on cardiac auscultation. Lung sounds were clear without crackles and there was no peripheral edema. MRI brain showed scattered areas of restricted diffusion in a linear pattern in the right centrum semiovale, also known as “strand of pearl sign,” with a few additional areas in the cortex of the right frontal and left

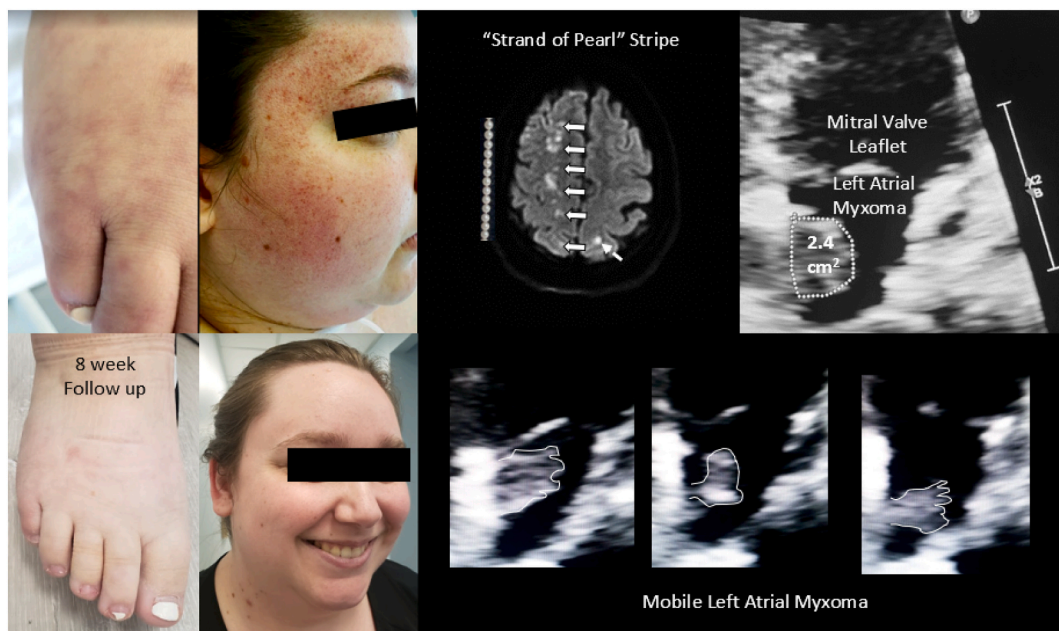


Fig. 1. Neurocutaneous presentation of atrial myxoma and sentinel rash. A 28 y woman with preceding rash (Top row, left) presented acutely with right facial rash (Top row) and ischemic stroke (Top row, MRI DWI sequence). Enclosed arrows show the ‘strand of pearl’ pattern of ischemia in the right corona radiata. An additional small area of ischemia was seen in the left occipital lobe (arrow). Atrial myxoma was found on echocardiography (top row, right). The atrial mass was frond-like and mobile (see bottom row, right). At 8 week follow up, both her stroke deficits and rash had resolved (bottom row, left).

occipital lobes. Transthoracic echocardiogram identified a mobile 2.4 cm atrial mass, normal atrial and ventricular sizes, and normal mitral and aortic valve function. She underwent uncomplicated cardiac surgery with pathologic confirmation of a villous atrial myxoma. At her 8 week follow up visit, her neurologic deficits had resolved, and her rashes had cleared.

2.1. Timeline

2 year history of migratory rash on distal extremities.

Abrupt onset of left sided weakness and right facial rash treated with intravenous tenecteplase.

Table 1

Literature review of published cases of sporadic atrial myxoma with cutaneous manifestations with inclusion of our case. Eight of these 18 patients (44 %) with atrial myxoma presented with acute stroke or TIA.

Reference	Key finding	Dermatologic Pathology	TIA or Stroke
Huston et al., 1978	Transient erythematous papules over dorsum of foot; a second episode in contralateral foot 4 months later; a third episode 7 months of purplish discoloration in right index finger and non-blanching macular rash over right thenar eminence; a fourth episode 11 months later with poplar rash over both feet, cyanosis in third and 4th digits of hand.	Histologic evidence of peripheral myxomatous emboli in gastrocnemius muscle biopsy	No
Feldman and Keeling, 1989	Blanchable, tender, erythematous macules and papules on fingertips, toes, palms, soles, and sides of feet	Noninflammatory destruction of medium-sized vessel in the reticular dermis with focal collections of mucin (myxomatous emboli)	No
Lee CW et al., 1991	Livedo reticularis on hands and forearms	Small arteries in reticular dermis filled with blue mucinous material (Alcian blue stain)	No
Abraham et al., 1995	Tender erythematous papules on fingertips, palms, and soles. Cutaneous vasculitis clinically diagnosed with rapid improvement with oral steroids	NA	No
Navarro et al., 1995	4 months prior to stroke, transient red macules in right hand; 3 months prior to stroke, macular eruption in lower right lower extremity involving the heel, sole, and sides of foot	Myxoid embolus within dermal vessels with positive alcian blue stain	yes
Greeson et al., 1998	Faint erythematous macules on left hypothenar eminence. Blanching erythematous papules (2 mm) on medial aspect of both feet (punch biopsy)	Mild non-specific chronic dermatitis and on occluded arteriole filled with myxomatous material (Alcian stain)	No
Loche et al., 1999	Violaceous papulosis on fingers and toes; 4 months prior had left hemiplegia with CT interpreted initially as hemorrhagic metastases and reinterpreted as embolic myxoma tumor fragments	Thrombosis of dermal vessels	Yes
Garcia-F-Villalta et al., 2002	Transient acral papular erythematous lesions resolving over 4-5 weeks without treatment. (legs/feet/soles). They preceded stroke symptoms by 2 years	Dermal vessels occluded by myxomatous material	Yes
Al-Mateen et al., 2003	an 11 y girl had transient red spots in nail beds of hand 2 months before stroke and similar lesions in soles of feet 2 weeks before stroke; a 10 year boy had 'red spots' involving the toes of left foot a few weeks before stroke	NA	Yes (2 cases)
Patel and Lynn, 2009	Blanching and erythematous rash in hands and feet at initial exam; 1 month later, new erythematous macules localized to feet with nonpalpable purpuric lesions in bilateral feet (punch skin biopsy performed); 6 months later, recurrence of skin changes in palms and soles	Cutaneous medium-vessel vasculitis	No
Paraiso and Prasad, 2009	Papulo-erythematous rash over palm and pallor in the third and fourth digits of the hand	NA	No
Praitano et al., 2010	15 months prior to stroke, transient erythematous macules in right palm and fingertips and soles of feet	NA	Yes
Alonso de Celada et al., 2012	Livedoid macules in palm and fingertips	Myxomatous emboli within Vater-Pacini corpuscles of hand	No
Fuchs et al., 2014	3 months prior to stroke, an intermittent spotted rash on both feet reported by patient; nontender, nonpalpable erythematous lesions on lateral sole and dorsum of foot on exam	NA	Yes
Rodriguez Bandera et al., 2015	Intermittent, violaceous, non-blanching reticulate macules on fingertips (thumb punch biopsy) and ankle	Myxomatoid embolus in dermal vessels	No
Macias et al., 2018	Blanchable, retiform, violaceous patches with a few sites of true purpura in bilateral lower extremities and duskiness of right second toe; biopsy performed on purpura in the dorsum of left foot	Ischemia and focal eccrine gland necrosis without evidence of vasculitis	No
Akins, 2023	pink reticulated patches and macules involving dorsal and palmar hands and fading skin changes from a prior similar eruption in her feet which involved the toes and sole; dermatologic findings preceded stroke by 2 years; simultaneous onset of right facial patchy erythematous macules and focal neurologic deficits from right cerebral ischemia (Fig. 1)	Subtle and sparse perivascular dermatitis with perivascular lymphocytosis	Yes

Stroke work up identified left atrial myxoma by echocardiography.
Cardiac surgery with resection of a villous atrial myxoma.
8-week postoperative visit with resolution of neurologic deficits and clearing of her rashes.

3. Discussion

This is the first published case of atrial myxoma presenting with simultaneous ipsilateral emboli into the internal and external carotid arteries causing acute ischemic stroke and acute facial rash. Our patient had a two year history of migratory skin changes in her distal arms and legs that in retrospect represented a “sentinel rash” of atrial myxoma. Surgical resection of her atrial myxoma lead to resolution of her acute and chronic skin changes.

The initial skin changes seen in my patient showed some features of Osler nodes described by Dr. William Osler in 1893 (tender, purple-to-pink nodules on tips of fingers and toes) and Janeway lesions described by Dr. Edward Janeway in 1899 (non-tender, irregular hemorrhagic papules and macules on the soles and palm) [3]. The differential diagnosis for these cutaneous symptoms includes infectious and non-infectious endocarditis, rheumatologic diseases causing small and medium vessel vasculitis, and systemic infections including Rocky Mountain spotted fever, meningococemia, and disseminated gonococcal diseases.

The “strand of pearls” finding in the corona radiata on her brain MRI (Fig. 1) indicates ischemia in the deep border zone of the cerebral hemisphere supplied by small, penetrating arteries branching off arteries along the cortex. This represents an end zone of cerebral perfusion without collateral blood supply which is vulnerable to emboli and hypoperfusion.

4. Literature review

A literature search was conducted in Pubmed and Google scholar using search terms: atrial myxoma, skin, dermatologic, cutaneous, rash. I identified 17 additional cases of cutaneous manifestations of sporadic atrial myxoma (Table 1) [4–19]. The rash was often seen on distal extremities (hands, feet) including the palm, sole, and digits. In 8 patients, myxomatous emboli were seen in the microvasculature on skin biopsy. Thrombosis of dermal vessels and ischemic skin changes were also observed. In 10 patients, the skin biopsy findings prompted cardiac imaging and the atrial myxoma was diagnosed and treated prior to onset of stroke or limb ischemia. The patient that I present is the first published case where the dermatologic findings involved the face and occurred simultaneously with stroke symptom onset.

A limitation of this case report and literature review is publication bias, as this is a rare symptom of a rare disease. A strength of this case report is the simultaneous appearance of the facial rash and cerebral ischemia in the same vascular territory (right common carotid artery) and rapid resolution after cardiac surgery.

5. Conclusion

Atrial myxoma can present with a sentinel rash. The skin changes should be re-conceptualized as embolic phenomena to the dermal microvasculature rather than constitutional symptoms. The distal appearance of the rash in the extremities, particularly when it involves fingertips and toes, is a common finding in patients with cardiac emboli. Earlier diagnosis of atrial myxoma when the sentinel rash first appears provides a valuable window of opportunity for cardiac surgery and tumor resection, before the patient experiences stroke or peripheral emboli.

Ethics statement

My patient provided written consent for publication of her case including photographs that included her face for the purpose of advancing medical education.

Data availability

I have no additional data to share regarding this case report.

CRedit authorship contribution statement

Paul T. Akins: Writing – review & editing, Writing – original draft, Methodology, Formal analysis, Conceptualization.

Declaration of competing interest

The authors declare the following financial interests/personal relationships which may be considered as potential competing interests: Paul T Akins reports a relationship with KAISER PERMANENTE MEDICAL CENTER - SACRAMENTO that includes: employment. I did not receive any funding for this case report and literature review If there are other authors, they declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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