Top of the Basilar Syndrome with Painful Palmar Reticulated Erythematous Patches Complicating Cardiac Atrial Myxoma: A Case Report

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Abstract

The authors reported an uncommon ischemic stroke in a young patient with an unusual dermatologic manifestation. A 28-year-old Thai woman presented with acute vertigo, ataxia, vertical diplopia, and impaired vertical gazes. Multiple painful, ill defined-border, reticulated, erythematous, and non-blanchable patches were formerly presented in both palms one day before the stroke onset. MRI showed hypersignals in bilateral thalami and left paramedian midbrain region on DWI with corresponding ADC map. Hyperechoic lobulated mass was detected at interatrial septum by transthoracic echocardiography. Top of the basilar syndrome with a rare skin sign were diagnosed and etiology was hypothesized as embolic phenomena caused by cardiac myxoma embolism. The dermatologic sign was exceptionally rare but the lesion can be a diagnostic clue for embolic mechanism caused by atrial myxoma. *(J Thai Stroke Soc 2015; 14 (3): 166-171.)*

Key words: cardiac myxoma, embolism, stroke, erythematous patch

Introduction

Atrial myxoma is a rare cause of stroke, however cardiac myxoma embolism should be considered in a differential diagnosis of ischemic stroke in young adults [1,2]. Top of the basilar syndrome is an uncommon ischemic stroke syndrome with various manifestations and mostly caused by embolism [3,4,5,6]. Skin manifestations in cardiac myxoma can be a part in complex syndromes including LAMB (lentigines, atrial myxomas, and blue nevi) and Carney syndrome which dermatologic lesion is a chronic presentation, however an unusual acute skin sign has occasionally been reported [7,8,9]. The authors presented a young Thai woman suffering acute ischemic stroke of the top of the basilar syndrome and uncommon dermatologic sign of bilateral painful palmar reticulated erythematous patches.

Case presentation

A 28-year-old Thai woman presented with acute vertigo, ataxia, vertical diplopia and impaired vertical gazes approximately 15 hours before admission. One day before the neurological complaints developed, she complained of painful erythematous rashes on both palms without weakness, numbness, dysphagia, facial paresis, or speech abnormality. She was a nonsmoker, and she had no history of diabetes, hypercholesterolemia and hypertension. She refused a family history of thrombo-embolic stroke, migraine or dementia. She previously had a couple of acute cerebral attacks at 3 months and 1 month earlier. In her first episode, she presented with acute diplopia and was diagnosed as brainstem encephalitis. Corticosteroid was given and her symptoms gradually improved within a few weeks. The second attack was a mild neurological complaint of dizziness.

Physical examination revealed blood pressure 103/67 mmHg, heart rates 95 bpm, normal respiratory rates without fever and systemic exams were unremarkable. Neurological examination showed slightly drowsy, limitation of upward and downward gazes, mild dysarthria, and generalized hypereflexia 3+. There was no sensory deficit, weakness or Babinski's sign. Dermatologic signs showed multiple ill defined-border, reticulated, erythematous, and non-blanchable patches on both palms *(figure a and b)*.

She underwent MRI brain and demonstrated hypersignals in the left paramedian area of midbrain and bilateral thalami on DWI with corresponding hyposignals map in ADC *(figure c and d)*. Transthoracic echocardiogram showed slightly reduced left ventricular systolic function with ejection fraction of 55% without regional wall motion abnormality. Hyperechoic lobulated mass 3.4x2.3 cm was detected and it was attached at interatrial septum close to aortic valve cushion and adjacent below anterior mitral valve leaflet. The mass was movable and occasionally moved into the left ventricle during diastolic phase without obstructing blood flow. No intracardiac thrombus was seen. Transcranial Doppler sonography, carotid duplex ultrasound and electrocardiography were normal.

Plasma glucose, blood urea nitrogen, creatinine, cholesterol, triglyceride, electrolyte, and coagulogram were normal. Complete blood count showed normal white blood cells and platelets. Hemoglobin was 12.7 g/dL with MCV 72 fL RDW 15%, 1+ microspherocytosis and ovalocytosis. Anticardiolipin antibody, rheumatoid factor, and antinuclear antibody profile were negative. Erythrocyte sedimentation rate was 23 mm/hr.

Top of the basilar syndrome with a rare dermatologic manifestation caused by atrial myxoma embolism was the diagnosis. Her skin lesions improved within a few days *(figure b)*. She had undergone successful operation for atrial myxoma removal and her neurological deficits were gradually improved.

Discussion

Myxoma is a rare benign cardiac tumor and occurs sporadically or as familial type in combination with two or more of the following conditions: skin myxomas, cutaneous lentiginosis, myxoid fibroadenomas of the breast, pituitary adenomas, primary adrenocortical micronodular dysplasia, and testicular tumors [10]. Classic triads of manifestations are obstructive cardiac symptoms, constitutional symptoms, and embolism which accounts most cases diagnosed with cardiac myxoma. The recurrence rate is low as approximately 5% after treatment [1,2,11]. Myxoma embolism is a rare cause of ischemic stroke and more involved in the anterior circulation [2].

The basilar artery is the most important vessel in the posterior circulation and prognosis of acute occlusion is generally poor. Infarction in rostral segment of the basilar artery (or top of the basilar) results in ischemia of pons, midbrain, thalamus, temporal lobes, occipital lobes and rarely cerebellum. Embolism is the most common cause of the top of the basilar syndrome [3,4,5,6]. Top of the basilar syndrome clinically manifests as numerous combination of abnormalities of alertness, sleep-wake cycles, behavior, oculomotor or pupil functions. Bilateral internuclear ophthalmoplegia, rubral tremor, and daytime somnolence are uncommon manifestations and mild hemiparesis was also reported [3,4,5,6,11].

Dermatologic manifestations can be abnormal pigmentation or myxomas of the skin which is a component of a rare genetic multiple endocrine neoplasia syndrome or Carney complex [7]. Other skin manifestations are lentigines and cutaneous myxomas [8]. Similarly, erythematous macular lesion in palms caused by cardiac myxoma has been reported [11].

The authors described a young 28-year-old woman presented with a limitation of vertical gazes, vertigo, slightly ataxia, and mild vertical diplopia. MRI supported the diagnosis of top of the basilar syndrome with bilateral thalamic infarcts and left medial midbrain ischemia on DWI and ADC *(figure c and d)*. Fascinatingly, the patient developed skin signs of multiple painful, ill defined-border, reticulated, erythematous, and non-blanchable patches on both palms one day before the stroke onset *(figure a and b)*. Skin lesions subsided in a few days after the stroke developed. The reticulated pattern of the dermatologic lesions indicates an association with vascular embolism in nature, for the reason that embolic occlusion in small blood vessels can cause ischemia occurring along reticulate-like blood vessel territories. The embolic occlusion can cause ischemic necrosis of the skin, however the obstruction can spontaneously resolve without skin lesion left in some cases.

Conclusions

The authors presented an uncommon cause of a rare ischemic stroke syndrome with unusual dermatologic signs in a young Thai woman. Cardiac myxoma embolism resulted in top of the basilar syndrome with atypical skin lesions on both palms. Painful reticulated erythematous patches on the palms are benign and can be a diagnostic sign concurrent or preceding with embolic stroke complicating atrial myxoma.

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กลุ่มอาการโรคหลอดเลือดสมองขาดเลือดจากยอดของหลอดเลือดเบสิลาร์ ร่วมกับผื่นแดงที่ฝ่ามือและเนื้องอกหัวใจมิกโซมา:

รายงานผู้ป่วย 1 ราย

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บทคัดย่อ

ผู้นิพนธ์รายงานโรคหลอดเลือดสมองที่พบไม่บ่อยร่วมกับอาการแสดงทางผิวหนังที่พบได้ยากในผู้ป่วยอายุน้อย ผู้ป่วยหญิงไทย อายุ 28 ปีมาโรงพยาบาลด้วยอาการ เวียนศีรษะ เดินเซ เห็นภาพซ้อนในแนวตั้ง และกลอกตาในแนวดิ่งขึ้นบนและลงล่างไม่ได้ โดย 1 วันก่อนอาการทางระบบประสาท ผู้ป่วยมีอาการปวดผื่นแดงขึ้นที่ฝ่ามือทั้งสองข้างลักษณะขอบเขตไม่ชัดเจน เป็นแนวยาวลักษณะเหมือนร่างแหและ เมื่อกดไม่จางลง ตรวจภาพวินิจฉัยด้วยแม่เหล็กพบพยาธิสภาพที่สมองส่วนฐานดอก (ธาลามัส) ทั้งสองข้างและสมองส่วนกลาง (มิดเบรน) ด้านซ้าย ด้วยวิธี ดีดับเบิลยูไอและเอดีซี การตรวจภาพสะท้อนด้วยคลื่นความถี่สูงหัวใจพบ ก้อนลักษณะเป็นปุ่มกลมที่ผนังหัวใจเอเตรียม ผู้ป่วยได้รับการวินิจฉัยเป็นกลุ่มอาการโรคหลอดเลือดสมองขาดเลือดจากยอดของหลอดเลือดเบสิลาร์ ร่วมกับอาการแสดงทางผิวหนังที่ฝ่ามือที่พบได้น้อย อันมีสาเหตุจากลิ่มเนื้องอกมิกโซมาอุดตันในหลอดเลือด อาการแสดงทางผิวหนังนี้พบได้น้อยมากและอาจเป็นอาการแสดงวินิจฉัยเฉพาะของโรคหลอดเลือดอุดตันจากเนื้องอกมิกโซมาได้

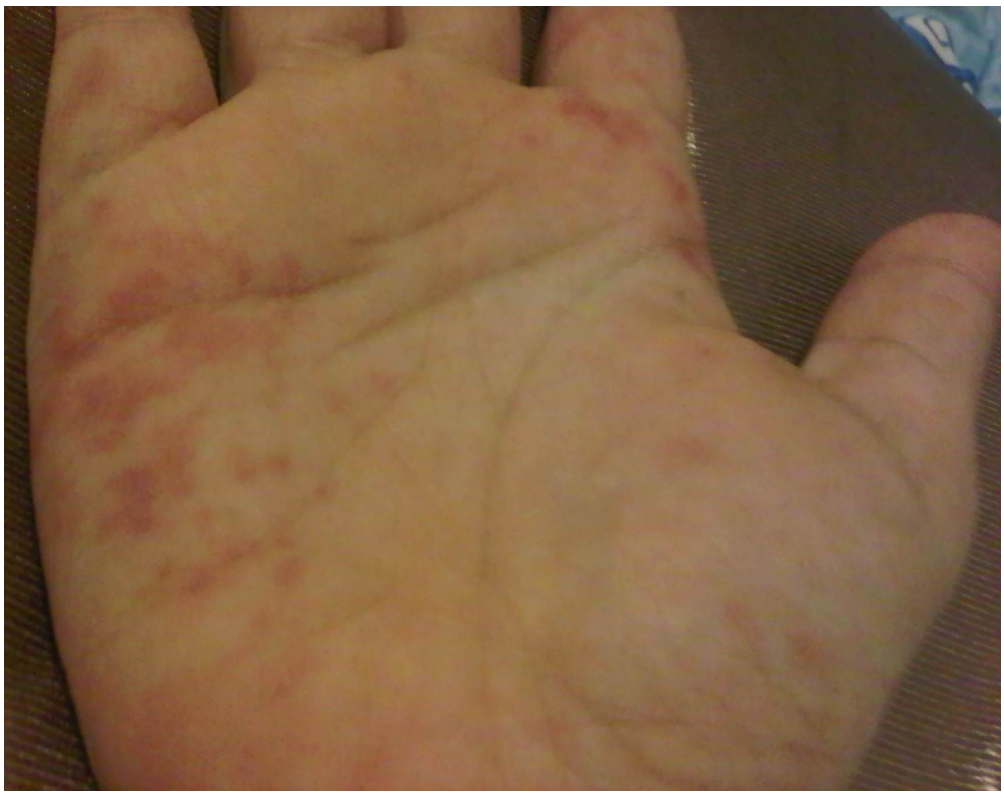

Figure a. Multiple painful, ill defined- border, reticulated, erythematous, and non-blanchable patches on the right palm.

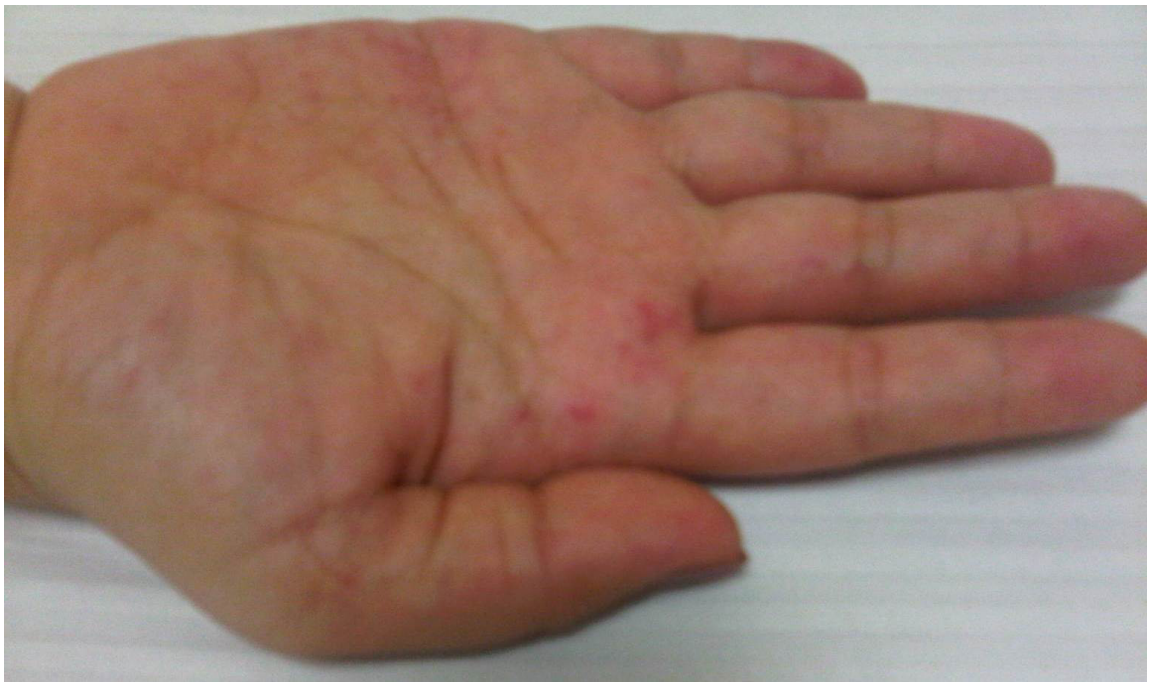

Figure b. Skin lesions gradually improved within a few days.

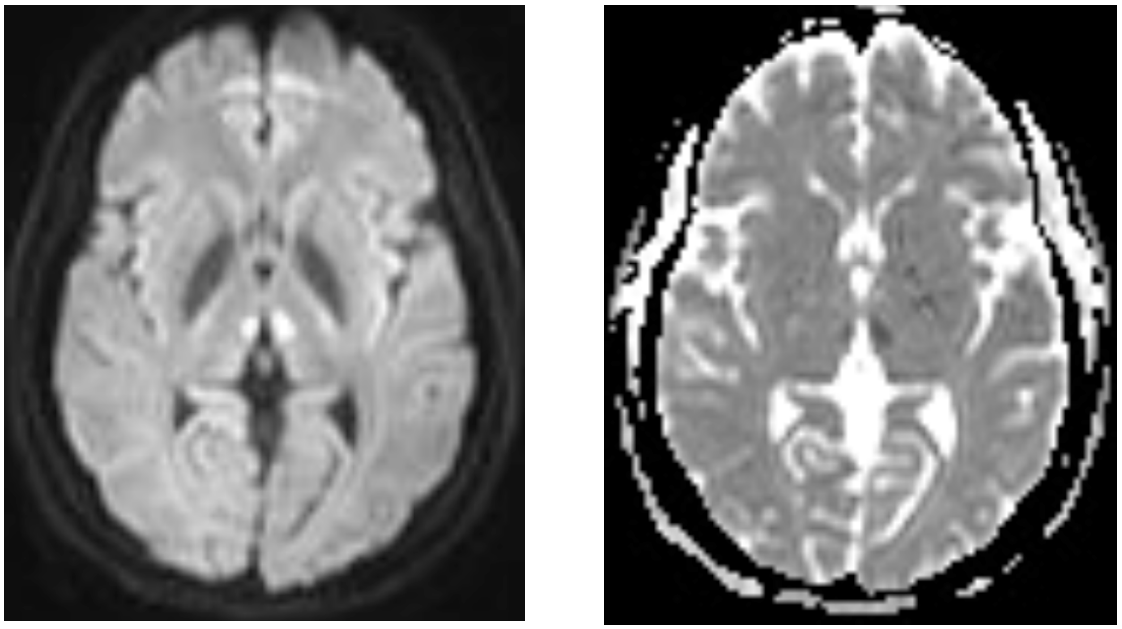

Figure c. Hypersignals in both thalami on DWI and corresponding hyposignals map on ADC.

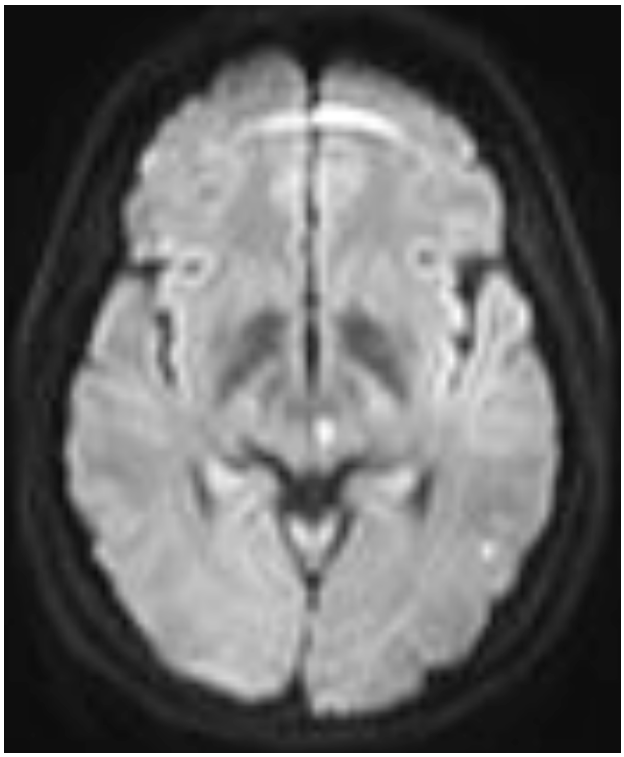

Figure d. Hypersignal at the left paramedian of the midbrain on DWI.