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Recurrent Embolic Stroke Associated with Long-Latency Relapsing Cardiac Myxoma

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Dear Editor,

Cardiac myxoma is the most common benign cardiac tumor. Although it has a very low reported incidence of 0.5−1 per 1,000,000 person-years, it is a potentially devastating cause of cardioembolic stroke. About 12% of cardiac myxoma cases are associated with neurologic disorders, most commonly ischemic stroke.[1] Surgical resection is the treatment of choice, and the recurrence rate was reportedly less than 1% per year, predominantly within 4 years.[2] Transthoracic echocardiographic screening is currently recommended in postoperative settings, but there is little evidence over appropriate duration or periods of follow-up strategies. Here we report a patient who presented with recurrent cardiac myxoma revealed by an embolic stroke event that occurred 12 years after complete resection, which had gone undetected during 9 years of routine echocardiographic follow-ups.

A 50-year-old female patient was referred to a clinic due to sudden-onset dysarthria, vertigo, and left-sided veering. In a neurologic examination the patient showed left lateropulsion and right-beating nystagmus. In addition, the patient presented with small (<1 mm) nontender painless rashes (i.e., Janeway lesions) on the volar side of both hands, which had developed 2 days prior to the symptoms (Fig. 1A). The patient had an infarction in the posterior inferior cerebellar artery territory 12 years previously. Newly detected 3.5-cm sized myxoma was regarded as a causative embolic source. The patient underwent complete resection of tumor, and had followed up with echocardiography annually for 9 years.

MRI performed after the current presentation showed multiple embolic infarctions mainly involving bilateral cerebella and the left frontal cortex (Fig. 1B). No arrhythmia was detected in electrocardiography and Holter monitoring. The patient underwent transthoracic and transesophageal echocardiography, which revealed a *de-novo* 3.5-cm hypermobile mass at the interatrial septum of the left atrium (Fig. 1C). The patient underwent complete resection, and myxoma was pathologically confirmed (Fig. 1D, E). The patient was eventually discharged without further neurologic or functional deterioration.

The surgical outcome of cardiac myxoma resection is generally favorable,[3] with recent data indicating a 10-year reoperation rate of less than 5%. Postoperative echocardiographic screening is recommended, but the need for screening may be questioned due to the low recurrence rate in nonfamilial forms.[4] Evidence is limited for the adequate duration of screening, with some anecdotal reports recommending surveillance for 5−10 years postoperatively.[5] Nonetheless, in the present patient no myxoma recurrence was seen during a thorough echocardiographic follow-up lasting 9 years after the resection, whereas recurrence was detected by a multiple embolic stroke after 12 years postoperatively. These findings indicate that a postoperative echocardiographic evaluation for more than 10 years may be beneficial in some patients with ischemic stroke due to myxoma even after complete resection.

In addition, cardiac myxoma is a causative agent of embolic stroke, but only a small proportion of embolic strokes are attributable to cardiac myxoma. The low recurrence rate could

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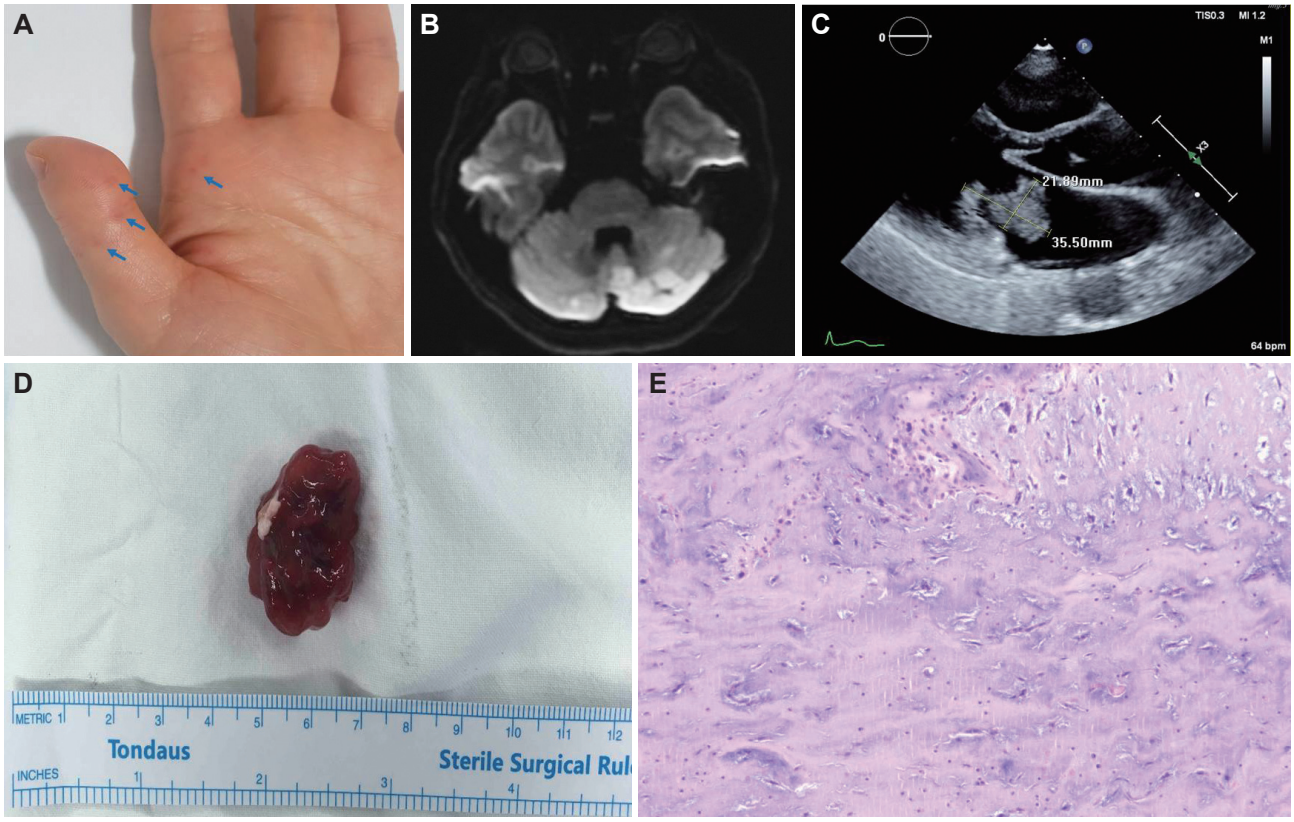

Fig. 1. Characteristic clinical, radiologic and histopathologic findings of the patient. A: Janeway lesion in the volar surface of left hand. There are several non-tender erythematous rash at the volar surface of thumb and palm (arrows). B: Diffusion Weighted Image of the patient. There are multiple diffusion restrictions in both cerebelli, mostly posterior inferior cerebellar artery territory. C: Trans-esophageal echocardiography of the patient, which reveals 3.5×2.1 cm-sized hypermobile echogenic myxoma in the left atrium. D: Surgical specimen of cardiac mass, which is attached to the left atrial septum. E: Microphotograph of myxoma from the left atrium. The tumor was composed of stellate cells with cords or nested structures in abundant myxoid substances (hematoxylin and eosin stain, ×100).

result in a physician facing embolic stroke with a history of myxoma resection overlooking the possibility of recurrence. However, the time interval between the embolic event and surgical resection is known to be the sole contributor of embolism recurrence,[2] and so timely diagnosis and management are important. There are a few reports on recurrent ischemic stroke due to recurrent myxoma, but in all cases the recurrence was detected within 10 years postoperatively.[6,7] Nevertheless, embolic stroke due to myxoma recurrence could occur even with an echocardiographically proven long tumor-free period, and so clinical suspicion is necessary. In the present case, the transient cutaneous manifestation in both hands suggested that Janeway lesion could be a clue for cardioembolic stroke, which has also been reported in myxoma.[8] A long tumor-free interval since myxoma resection should therefore not be used to exclude a cardiac etiology of stroke, and a thorough neurologic evaluation with etiologic assessments are imperative to ensure appropriate diagnosis and management.

This case was subject to a particular limitation. Some cardiac myxomas are genetically associated with known Carney complex, presenting as skin pigmentation, endocrinopathy, and tumors. Familial predisposition with autosomal dominant pattern is seen, and cardiac myxoma could occur in multiple forms with any cardiac chamber in Carney complex. The present patient exhibited recurrent myxoma in the atrium without a familial predisposition nor clinical features compatible with Carney complex, and so the likelihood of Carney-complex-associated myxoma is assumed to be low, resulting in genetic confirmation of mutations in PRKAR1A not being performed. Nonetheless, reports concerning long-latency recurrence or neurologic manifestation of Carney complex are also lacking, and so further case studies are necessary.

Author Contributions

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Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

REFERENCES

1. Lee VH, Connolly HM, Brown RD Jr. Central nervous system manifestations of cardiac myxoma. *Arch Neurol* 2007;64:1115-1120.
2. Stefanou MI, Rath D, Stadler V, Richter H, Hennersdorf F, Lausberg HF, et al. Cardiac myxoma and cerebrovascular events: a retrospective cohort study. *Front Neurol* 2018;9:823.
3. Lee SJ, Kim JH, Na CY, Oh SS. Eleven years' experience with Korean cardiac myxoma patients: focus on embolic complications. *Cerebrovasc Dis* 2012;33:471-479.
4. Vroomen M, Houthuizen P, Khamooshian A, Soliman Hamad MA, Van Straten AH. Long-term follow-up of 82 patients after surgical excision of atrial myxomas. *Interact Cardiovasc Thorac Surg* 2015;21:183-188.
5. Shah IK, Dearani JA, Daly RC, Suri RM, Park SJ, Joyce LD, et al. Cardiac myxomas: a 50-year experience with resection and analysis of risk factors for recurrence. *Ann Thorac Surg* 2015;100:495-500.
6. Ghosh A, Bhattacharyya A, Niyogi P. Recurrent left atrial myxoma with recurrent stroke. *Indian Pediatr* 2001;38:1190-1192.
7. Abu Abeeleh M, Saleh S, Alhaddad E, Alsmady M, Alshehabat M, Bani Ismail Z, et al. Cardiac myxoma: clinical characteristics, surgical intervention, intra-operative challenges and outcome. *Perfusion* 2017;32:686-690.
8. Rodríguez Bandera AI, Stewart NC, Uribe P, Minocha R, Choi JY. Cutaneous embolism of an atrial myxoma. *Australas J Dermatol* 2015;56:218-220.