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Cardiac Papillary Fibroelastoma in a Patient with Diabetic Ketoacidosis: A Case Report

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Abstract

Papillary fibroelastoma, a benign primary cardiac tumor, has potentially life-threatening complications. We report a patient with an initial diagnosis of diabetic ketoacidosis. Magnetic resonance imaging of brain showed a recent infarct at right temporal lobe. Non ST-segment elevation myocardial infarction (NSTEMI) was noted on hospital day 2. Echocardiography revealed normal wall motion and a 1-cm pedunculated mass at the aortic valve without aortic regurgitation. The tumor was thought to be responsible for cerebral infarction and NSTEMI that accelerated diabetic decompensation. The patient underwent surgical excision, and the histological feature was consistent with a papillary fibroelastoma. (J Intern Med Taiwan 2017; 28: 41-46)

Key Words: Papillary fibroelastoma, Diabetic ketoacidosis, Cerebral infarction, Myocardial infarction

Introduction

Primary cardiac tumors are extremely rare, with an incidence rate of 0.02% revealed in autopsy series[1]. While the exact incidence of papillary fibroeslastoma is unknown, it accounts for 5% of primary cardiac tumors based on surgical series[2]. Over 80 percent of papillary fibroelastomas are found on the valve leaflet. They are also the most common tumor of cardiac valves. The etiology of papillary fibroeslastomas remains unknown. Theses tumors usually occur in adulthood, particularly in those aged 40-80 years[2,3]. Histological appearance is characterized by a collection of avascular fronds of dense connective tissue, superficially covered by endothelial cells. Here, we report a case of diabetic ketoacidosis (DKA) accompanied by cerebral infarction and non ST-segment elevation myocardial infarction (NSTEMI), a cardiac papillary fibroelastoma was discovered.

Case report

A 48-year-old man presented to the emergency department with acute unconsciousness and shortness of breath. He was diagnosed with type 2 diabetes two months prior to this presentation. Over the preceding three days, he experienced intermittent convulsions of the left upper limb without an altered

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mental status and visited a local clinic. Each convulsive episode lasted from a few seconds to a few minutes. Repeated episodes affected him to hold a rice bowl and water glass. He could not eat and drink well. He has no previous history of seizure, head trauma or illicit drug use. The family history was negative for neurologic diseases, cardiovascular diseases and neoplasms. A positive family history of diabetes was reported.

On physical examination, his blood pressure was 132/86 mmHg and his pulse was 128 beats/min. The respiration was 32 beats/min. The body temperature was 38.5°C. His Glasgow Coma Scale was E3M5V2 and his chest sounds were clear. He had rapid heart beats with no cardiac murmurs. His abdomen was soft with hypoactive bowel sounds and his limbs were symmetrically weak without convulsive motions. His skin turgor was decreased. Petechiae or subcutaneous nodule were absent.

Laboratory tests revealed a high blood glucose level of 905 mg/dl and ketone body of 5.0 mmol/L (0–0.06mmol/L). Arterial blood gas showed a pH of 7.120, PCO_2 of 15.8 mmHg, PO_2 of 133.7 mmHg and HCO_3^- of 5.2 meq/L on O_2 mask 6 L/min. The sodium, potassium, creatinine, and blood urea nitrogen were 113 mmo/L, 5.5 mmol/L, 2.82 mg/dL and

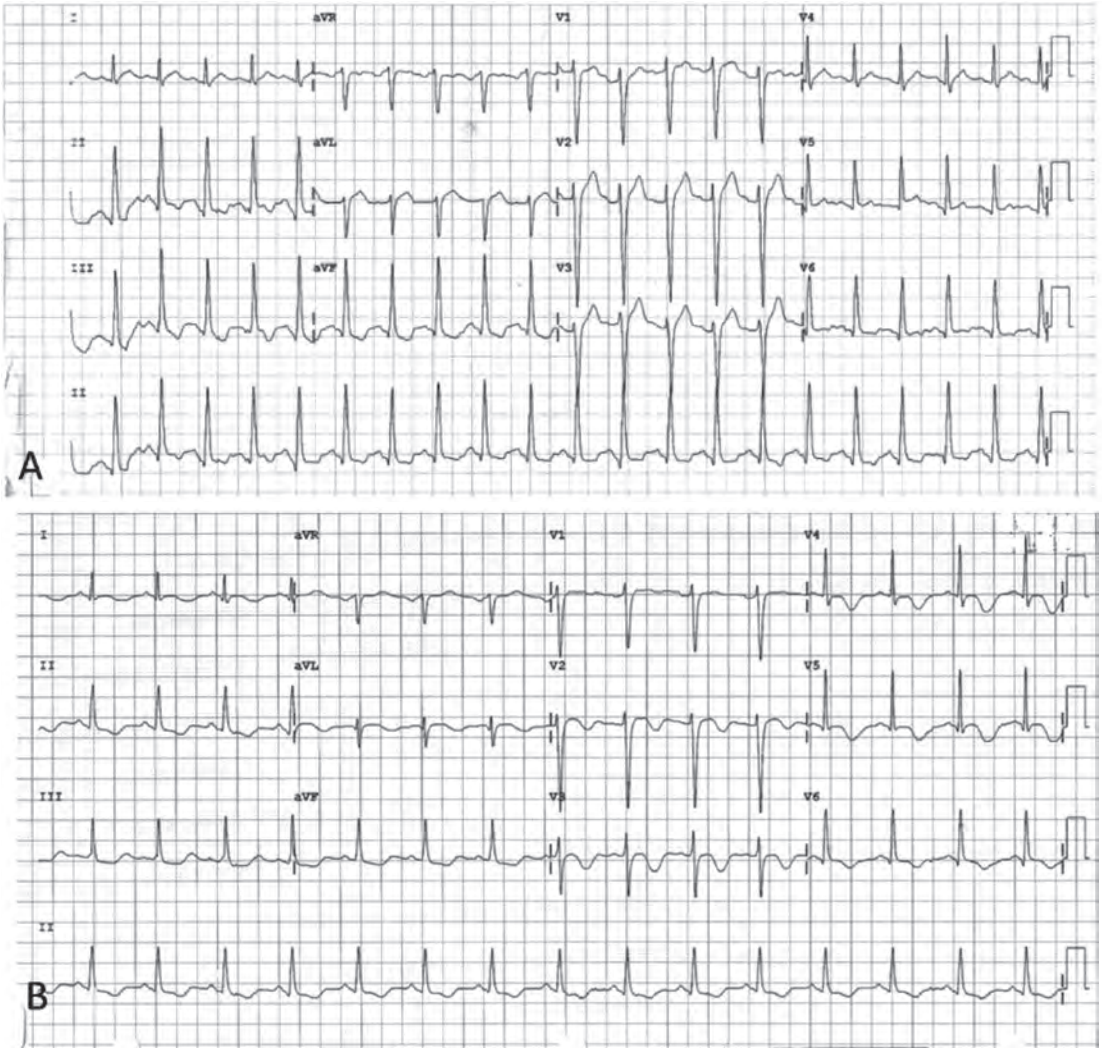

Figure 1. Day 1 ECG : sinus tachycardia, left ventricular hypertrophy and borderline inferior Q wave in II, III, aVF (A). Day 2 ECG : T wave inversion with ST depression in leads II, III, aVF and V2-V6 (B).

60 mg/dL, respectively. The white blood cell count was 25310 cells/uL with 80% of neutrophil. C-reactive protein level was 0.814 mg/dL (0–0.8 mg/dL). Serum lipase was 255 IU/L (22–51 IU/L). Troponin I was 0.111 ng/mL (0–0.04 ng/mL). Creatine phosphokinase (CPK) was 264 IU/L (30–170 IU/L). CPK-MB was 6.2 ng/mL (0.6–6.3 ng/mL). Electrocardiogram (ECG) showed sinus tachycardia, left ventricular hypertrophy and borderline inferior Q wave in II, III, aVF (Figure 1). Chest X-ray and brain computed tomography (CT) were negative. Focal atelectasis and consolidation with air-bronchogram at both lower lung fields were found on CT. However, the CT scan was negative for pancreatitis. Fluid and insulin replacement therapies were immediately started. Empiric antibiotic therapy was administered for suspected pneumonia. A nasogastric tube was placed and some coffee grounds were drained.

For investigating the other causes of the altered mental state, brain magnetic resonance imaging (MRI) was performed. Diffusion-weighted imaging revealed a 1.2-cm hyperintense lesion in the right temporal lobe (Figure 2). On hospital day 2, the patient regained his consciousness. However, follow-up troponin I of 12.886 ng/mL, CPK of 1240 IU/L and CPK-MB of 34.6 ng/mL were obtained. A repeat ECG showed T wave inversion with ST depression in leads II, III, aVF and V2-V6 (Figure 1). Simultaneous hypokalemia (serum potassium, 2.2 mmol/L) required treatment. The diagnosis of NSTEMI was made. Clopidogrel was given. Transthoracic echocardiography (TTE) revealed no regional wall motion abnormality but a shaggy mass over the aortic valve. Infective vegetation could not be excluded. Subsequent transesophageal echocardiography (TEE) showed a pedunculated mobile mass at the aortic valve without aortic regurgitation, highly suggestive of papillary fibroelastoma (Figure 3). Blood cultures, urine culture, antinuclear antibody and anticardiolipid antibody were negative. The patient was transferred to the cardiac surgery department and underwent tumor excision. Histopathology confirmed a papillary fibroelastoma, com-

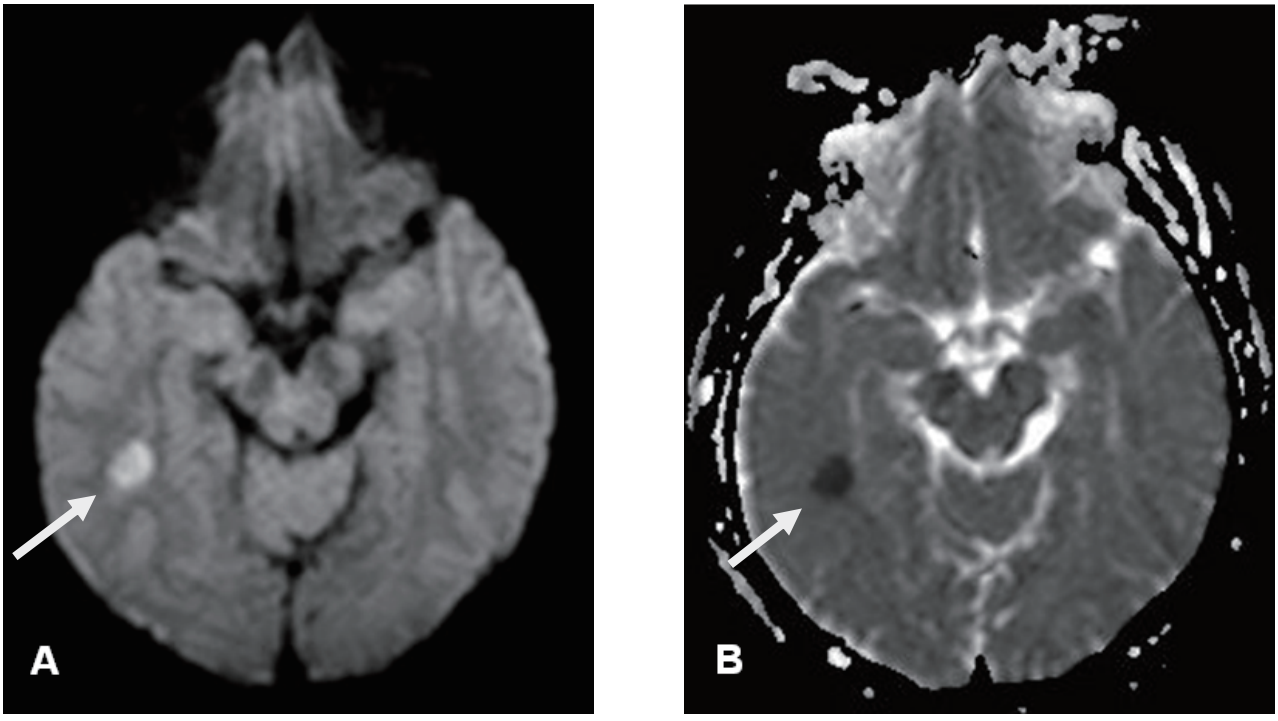

Figure 2. Diffusion-weighted imaging revealed a 1.2-cm hyperintense lesion at the right temporal lobe (A). Apparent diffusion coefficient had a low signal intensity (B).

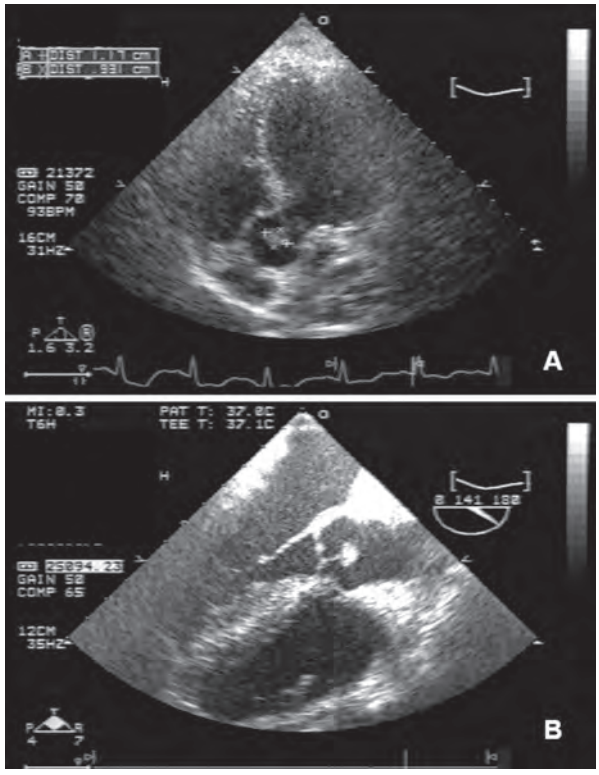

Figure 3. Transthoracic echocardiography showed a 1.17 × 0.93 cm, shaggy mass at the aortic valve (A). Transesophageal echocardiography showed a mobile mass attached to the aortic valve by a thin stalk (B).

posed of papillary lesion with hyaline avascular core and superficial endothelial lining. The intermediate layer contained a few fibroblasts and inflammatory cells (Figure 4). The patient's postoperative recovery was uneventful.

Discussion

DKA is an acute complication of diabetes; its annual incidence rate is estimated to be 4.6–8.0 episodes per 1000 patients with diabetes[4,5]. Most patients with DKA are type 1 diabetes in Europe and North America. In contrast to white people, type 2 diabetes accounts for 54.6%–79.5 % of DKA admissions in Taiwan's medical centers[6,7]. DKA is a state of absolute or relative insulin deficiency coupled with elevated counter-regulatory hormones. Infection is the major precipitating factor followed by non-compliance of medication and

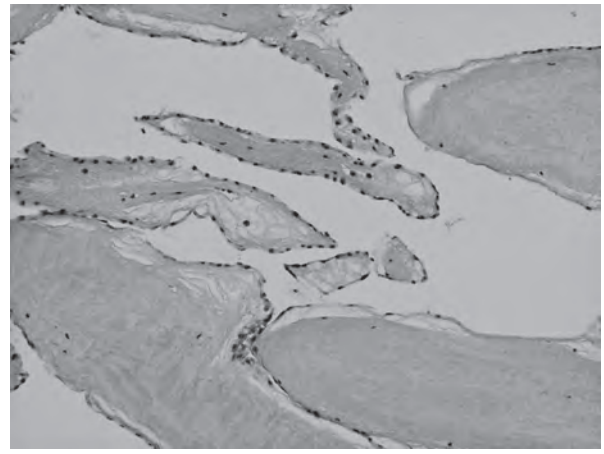

Figure 4. Hematoxylin and Eosin stain, 200×: Papillary fronds. Note the endothelial covering, intermediate proteoglycan-rich matrix, and inner avascular fibrous cores.

new-onset diabetes mellitus. The other acute causes include pancreatitis, trauma, myocardial infarction, cerebrovascular accident, drugs, and psychological problems. Because of advances in the clinical recognition and management, the mortality rates have fallen from 7.96%–12.9% to 0.67%–2.65%. However, the poor prognosis is observed in the elderly patients and anyone with severe comorbidities[7,8,9]. Thus, investigating the possible concurrent illness in all DKA admissions is crucial.

In our patient, unilateral and repetitive seizure-like events were witnessed before admission. Brain imaging should be used to exclude or identify a structure abnormality. Brain MRI could provide more information if a CT scan is negative. In our case, there was no typical hyperintensity involving contralateral basal ganglion on T1-weighted MRI to correlate with hyperglycemia-related involuntary movement. However, a recent ischemic infarction was diagnosed by hyperintensity on DWI and low signal on apparent diffusion coefficient image. This contralateral lesion probably provoked his focal seizures with or without impairment of consciousness, despite lack of electroencephalogram investigation, experienced witnesses and detailed history. The prolonged unconsciousness in our patient might

result from severe DKA or during postictal period.

Papillary fibroelastomas arise from the endocardium, usually on cardiac valves. Aortic valve and mitral valve are most common location. They consist of multiple avascular fronds attached to the endocardium by a thin stalk. The gross appearance looks like a sea anemone when placed in water and saline. Although sizes ranging from 2 to 70 mm have been reported, papillary fibroelastomas are generally small (< 10 mm) and solitary[3]. With advances in technology and better awareness of primary cardiac tumor, a presumptive diagnosis of papillary fibroelastoma is possible through echocardiography, especially TEE. Alternative cardiac CT and MRI may have limitations in detecting small mobile masses. Tamin et al. reported an increased occurrence (from 0.019% in 1980–1995 to 0.089% in 1995–2010) of papillary fibroelastoma in patients referred for echocardiography[10]. The clinical presentations vary from asymptomatic to severe embolic complications. A comprehensive analysis estimated that 30% of patients were asymptomatic and incidentally diagnosed[3]. Symptoms such as transient ischemic attack, stroke, visual loss, angina, myocardial infarction, heart failure, mesenteric ischemia, renal infarction, limb ischemia, pulmonary embolism, syncope and sudden death have been reported. The most symptoms are attributed to embolism from the fronds of the tumor itself or thrombi on the tumor surface. Some complications result from a direct occlusion of coronary artery ostium or narrowing of the ventricular outflow tract.

Unlike papillary fibroelastomas, myxomas are highly vascular tumors and commonly located in the atria. Lambl's excrescences are smaller filiform fronds occurring at the sites of valve closure. In contrast to vegetations, papillary fibroelastomas are located on the downstream of the valves and do not destruct them. Most patients do not have a history of valvular disease, and papillary fibroelastomas can be safely excised with native valve preservation and low recurrence rate. At follow-up, patients without surgical treatment were more predisposed to stroke and death than those with surgical treatment[10]. However, no data are present explaining whether medical management with antiplatelet or anticoagulation improves outcomes.

In our case, DKA was initially diagnosed. The presence of stroke was identified by MRI. Because of high cardiac enzyme levels, myocardial infarction was noted. A shaggy mass on the aortic valve was found by transthoracic echocardiography (TTE). Transesophageal echocardiography (TEE) is much helpful to recognize papillary fibroelastoma. Coronary angiography was not intervened due to the risk of tumor or surface thrombi dislodge. In our patient, it was speculated that myocardial injury was due to the occlusion of coronary artery by tumor or its emboli, although the possibility of preexisted coronary atherosclerosis could not be excluded. A similar situation has been reported in a case of hyperglycemic hyperosmolar state, not DKA. Khair et al. reported a case of aortic valve papillary fibroelastoma proved by autopsy, who was just discharged after receiving treatment for hyperglycemic hyperosmolar state, pneumonia, and previous cerebral vascular accident, with negative results on TTE. Five days later, the patient was readmitted due to nausea, vomiting, and shortness of breath, and died at the next day[11]. Based on our patient and the previous case, primary cardiac tumor should be considered as possible diagnosis in patient with unexplained simultaneous cardiac and neurological event. A successful treatment requires a prompt and careful investigation of such a precipitating cause.

Conclusion

According to our review of relevant literature, this is the first report of DKA as a novel presentation in a patient with papillary fibroelastoma complicated by NSTEMI and stroke. The tumor was successfully excised. Papillary fibroelastoma should be

considered as a possible precipitant of DKA if unexplained vascular events are identified.

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糖尿病酮酸中毒合併心臟乳突纖維彈性瘤一病例報告

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摘 要

乳突纖維彈性瘤是罕見的疾病，雖屬於良性原發性心臟腫瘤，但它的併發症可能潛藏致命風險。我們分享一個案例起初診斷糖尿病酮酸中毒，核磁共振確認腦部出現小血管阻塞，次日發現非ST波段上升心肌梗塞存在，心臟超音波顯示心肌收縮正常，但在主動脈瓣上看到一個有梗腫塊，大小為一公分。考慮這腫瘤應病人中風與心肌梗塞的元兇，且間接引起糖尿病失控。於是安排手術切除。病理組織證實為乳突纖維彈性瘤，病人手術癒後良好。