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Brief Report

Congenital left atrial appendage aneurysm associated with a systemic embolism

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Abstract A 20 year-old woman presented with systemic embolisation. On subsequent investigation, she was diagnosed with a congenital left atrial appendage aneurysm. Few case reports are reported in the literature. This cardiac malformation presents a diagnostic challenge in patients with cardiomegaly.

Keywords: Aneurysm; arrhythmia; embolism

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Isolated aneurysmal dilation of either the left or right atrium is rare as a congenital abnormality, being first described by Semans and Taussig in 1938.[1] Such congenital aneurysm of the left atrial appendage is extremely rare because, since first described by Diamond and colleagues in 1960,[2] only 50 additional cases have been reported. It presents a diagnostic challenge in patients presenting with cardiomegaly. Most cases are ultimately recognised when the patient develops a supraventricular arrhythmia and/or systemic embolisation. With successful surgical resection, the prognosis is excellent. We review here a patient diagnosed with such an aneurysm, along with the pertinent literature.

Case report

A 20-year-old woman presented with a 1-year history of intermittent high-grade fevers, chills, and headaches. The febrile episodes typically were intermittent with a frequency of 1–2 episodes per day lasting for 8–10 days alternating with 8–10 days, during which she was afebrile. She denied a history of cough, symptoms of an upper respiratory tract infection, dysuria, or diarrhea. Her past history was negative for diabetes, hypertension, and intravenous drug use. When her chest radiograph demonstrated cardiomegaly, she was referred to our hospital for further evaluation. Her physical examination was entirely normal. The chest radiograph demonstrated a convexity of the left heart border compatible with a dilated left atrium (Fig 1a). The electrocardiogram revealed normal sinus rhythm, normal axis, and a heart rate of 86/minute with no significant abnormality. The 24-hour holter monitor uncovered no significant arrhythmia. The transthoracic echocardiogram was consistent with a giant aneurysm of the left atrial appendage. This aneurysm measured 88 mm in the longitudinal axis in the subcostal window, with a transverse diameter of 42 mm in the apical window. The neck of the appendage, communicating with the left atrial cavity, was 18 mm in diameter. A large echo-dense shadow, consistent with thrombus, was seen in the left atrial appendage and measured 16 by 22 mm (Fig 1b). The tricuspid regurgitation jet predicted mild pulmonary hypertension. The left ventricular diameter was within normal limits and there was mild eccentric hypertrophy of the left ventricular walls. The left ventricular systolic function was well preserved and its wall motion was normal. The interatrial septum and heart valves were all normal. The visceral layer of the pericardium appeared to be intact. The complete blood count revealed a microcytic hypochromic anemia with normal white blood cell and platelet counts. The erythrocyte sedimentation rate was 30 mm/hour, C-reactive protein was 28 mg/L, and the blood culture was negative. Thyroid function tests were within normal limits. Because the patient refused a

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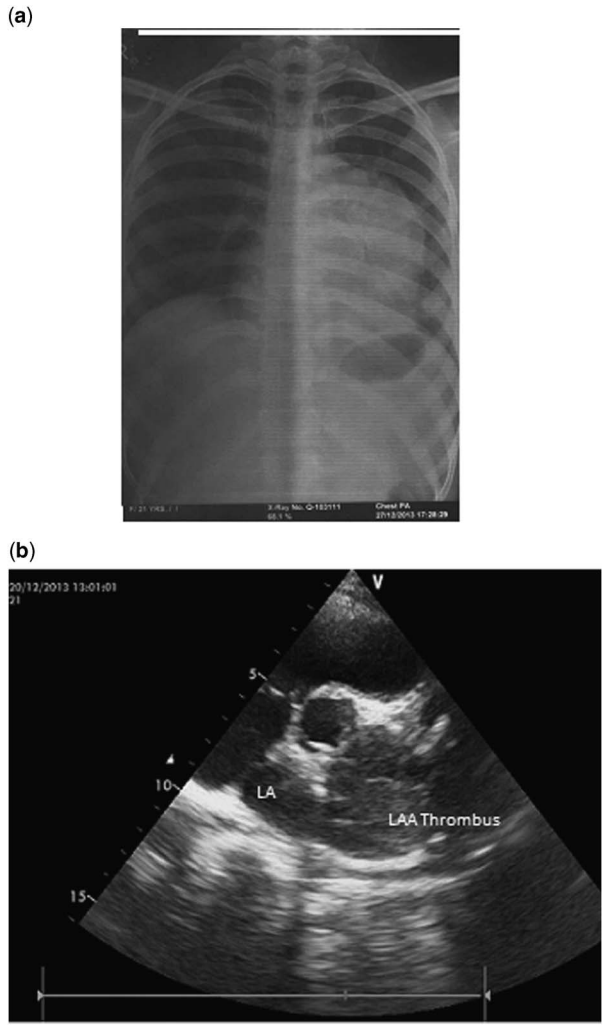

Figure 1.
*(**a**) Chest radiography reveals prominent left heart border. (**b**) Two-dimensional echocardiogram showing thrombus containing left atrial appendage aneurysm communicating with the left atrium.*

transoesophageal echocardiogram, cardiac magnetic resonance imaging was obtained and confirmed a giant left atrial appendage aneurysm containing thrombus with communication to the left atrium (Fig 2). For 4 days subsequently, the patient developed a right femoral artery embolism for which embolectomy was performed. The patient was advised to undergo surgical resection of the left atrial appendage aneurysm, but she refused. She was discharged on oral anticoagulation, warfarin 5 mg, and was subsequently lost to follow-up.

Discussion

Aneurysm of the left atrial appendage is a rare anomaly; with about 50 published case reports.[3] Some authors describe it as a "dangerous fifth chamber".[4] The majority of these cases are congenital, although there are also reports of aneurysms acquired because of an elevation of the left atrial pressure.[5] It generally presents as an isolated cardiac malformation and is related to dysplasia of the pectinate muscles in the left atrial appendage and of the bands of atrial muscle from which they arise.[6] They are rarely diagnosed during childhood and generally become manifest during the second or third decades of life.[7] Our patient presented with intermittent episodes of fever, chills, and headaches followed, after the diagnosis of the left atrial appendage aneurysm, by the development of a femoral artery embolism, despite the absence of an atrial arrhythmia such as atrial fibrillation. Although most patients with this cardiac malformation are reported to be asymptomatic, some can present with palpitations related to atrial arrhythmias, dyspnea, angina related to compression of the left coronary artery, and cerebrovascular incidents due to systemic embolisation. Atrial thrombi

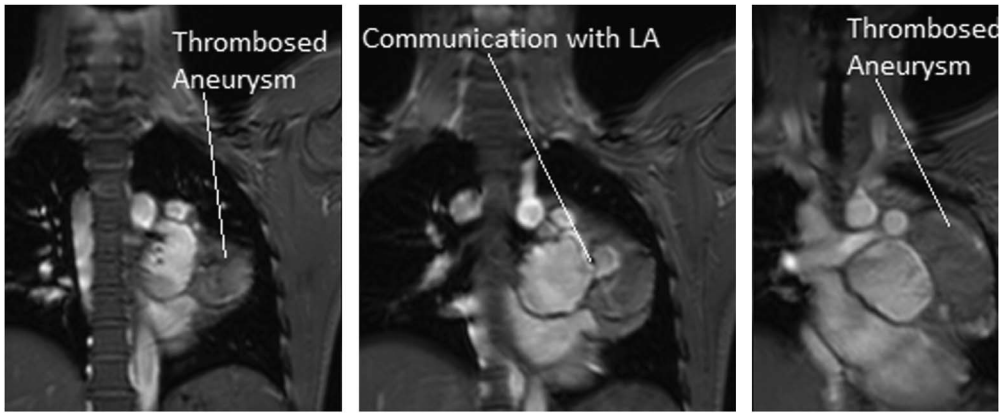

Figure 2.
Cardiac magnetic resonance imaging shows thrombus containing left atrial appendage aneurysm.

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occur, especially in patients with atrial arrhythmias, and also in the absence of arrhythmias. Thrombosis with subsequent systemic embolisation has been reported, similar to our case. Stasis of blood within the aneurysm may be the predisposing factor for thrombus formation.[8] Radiographically, a left atrial appendage aneurysm must be differentiated from other pathologies such as a mediastinal mass, pericardial cyst, cardiac tumour, and a pericardial or extracardiac fluid collection. They are not linked to any other pathologies and should be distinguished from other diagnoses such as congenital deficit of the pericardium with atrial herniation, mitral valve pathology causing atrial dilation, or juxtaposition of the atrial appendages. Suggested criteria for the diagnosis of a congenital left atrial appendage aneurysm include (1) the absence of any concomitant cardiac pathology that could cause atrial dilation, (2) the presence of a left atrium and appendage of normal morphological characteristics, (3) a direct continuity of blood flow between the left atrium and the appendage, and (4) the absence of pericardial defects.[9] The diagnosis is usually made by transthoracic and/or transoesophageal echocardiography. Cardiac magnetic resonance imaging can also play an important role in diagnosis. Once the diagnosis has been established, aneurysms of the left atrial appendage must be treated because of their potential morbidity and mortality. The recommended treatment is resection, generally using cardiopulmonary bypass. However, good surgical results have also been reported through a left thoracotomy incision without cardiopulmonary bypass.[7] Surgery has been proven to be effective both in the prevention of new atrial arrhythmias and of systemic emboli. There are case reports in the literature of patients who are free of atrial arrhythmias 8 years after aneurysmectomy.[10,6] Although resection alone is usually adequate, it is sometimes combined with the Cox-Maze procedure for ablation of the atrial fibrillation pathways, especially if there is clinically important dilation of the left atrium or if a previous electrophysiological study has shown induction of atrial fibrillation by atrial foci.[10] As demonstrated by our case, it seems judicious to offer elective aneurysmectomy to an asymptomatic patient with a congenital aneurysm of the left atrial appendage as a prophylactic measure to minimise the chances of the future development of systemic embolisation or atrial arrhythmias.

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Conflicts of Interest

None.

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