



### Multiple Embolisations in Rheumatic Heart Disease: A Case Report

*Plusieurs embolisations in Rheumatic Heart Disease: A case report*

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#### ABSTRACT

**BACKGROUND:** Though systemic thromboembolism is not an infrequent complication of rheumatic valvular disease, multiple embolic phenomena are rare.

**OBJECTIVE:** To present a patient with rheumatic heart disease associated with multiple embolic complications.

**CASE REPORT:** A 44-year-old lady with rheumatic valvular disease and atrial fibrillation defaulted anticoagulant medication, and subsequently presented with acute chest pain, acute left ventricular failure, focal neurological deficit and gangrenous lower limb extremities. Electrocardiography showed atrial fibrillation and an old anteroseptal myocardial infarction. Echocardiography showed multiple valvular lesions and multiple thrombi in the left atrium. Computed tomogram scan demonstrated a large infarct involving the region supplied by the right middle cerebral artery. Bilateral above knee amputation was performed. Recovery from neurological deficit was complete. She had, during a 4-year follow-up and anticoagulation remained free of new clinically evident embolic complications. Serial echocardiography however showed that the atrial clots had persisted and presumably fibrosed.

**CONCLUSION:** Multiple systemic thromboembolisms are serious complication of atrial fibrillation of valvular aetiology, and their prevention requires continuous anticoagulation. *WAJM* 2009; 28(4): 274–276.

**Key words:** Valvular lesions, atrial fibrillation, Embolisms, Anticoagulation.

#### RÉSUMÉ

**CONTEXTE:** Bien que thrombo-embolie systémique n'est pas une complication rare de valvulopathie rhumatismale, plusieurs phénomènes emboliques sont rares.

**OBJECTIF:** Présenter un patient présentant une cardiopathie rhumatismale associée à de multiples complications emboliques.

**CASE REPORT:** A 44-year-vieille dame avec une valvulopathie rhumatismale et de fibrillation auriculaire par défaut médicament anticoagulant, et par la suite présenté une douleur thoracique aiguë, insuffisance ventriculaire gauche, un déficit neurologique focal et gangrénée extrémités des membres inférieurs. Electrocardiographie montrée la fibrillation auriculaire et d'un infarctus du myocarde antéroseptal vieux. L'échocardiographie a révélé de multiples lésions valvulaires et thromboses multiples dans l'oreillette gauche. Tomographie par ordinateur analyse a démontré un infarctus de grande envergure impliquant la région alimentée par l'artère cérébrale moyenne droite. Bilatéraux amputation au-dessus du genou a été réalisée. Récupération d'un déficit neurologique était complète. Elle avait, au cours d'une 4-year follow-up et anticoagulation restées indemnes de nouvelles complications emboliques cliniques évidents. Échocardiographie de série a toutefois montré que les caillots auriculaire avait persisté et, vraisemblablement, fibreux.

**CONCLUSION:** Plusieurs accidents thromboemboliques systémiques sont complication grave de la fibrillation auriculaire, d'étiologie valvulaire, et leur prévention exige anticoagulation continue. *WAJM* 2009; 28(4): 274–276.

**Mots clés:** lésions valvulaires, la fibrillation auriculaire, embolies, l'anticoagulation.

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Abbreviations: RHD, Rheumatic heart disease.

## INTRODUCTION

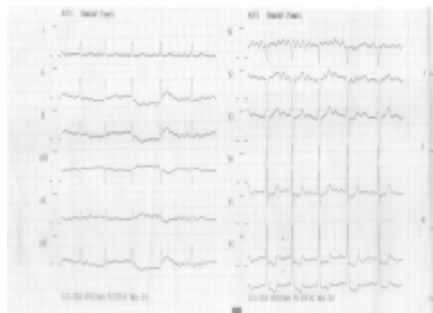
Rheumatic heart disease (RHD) is a chronic sequel of rheumatic fever, a type II hypersensitivity reaction to pharyngeal infection with beta haemolytic Lancefield Group A organism. It occurs frequently among children and young adults in low income nations because of poverty, overcrowding and poor hygiene. In East Africa and India, for example, about 6.4 per 1000 and 6.8 per 1000 children of school age, respectively have rheumatic heart disease.<sup>1,2</sup> It accounts for about 13.3% of adult cardiovascular diseases in Nigeria.<sup>3</sup> Though thromboembolism is not infrequent,<sup>2,3</sup> concomitant embolic complications are rare in RHD. This report describes a patient with RHD complicated by embolisation to the peripheral, cerebral and coronary arteries that resulted in lower limb gangrene, ischaemic stroke and myocardial infarction, respectively.

## CASE REPORT

A 44-year old indigenous Nigerian woman with RHD and atrial fibrillation defaulted anticoagulation and presented eight weeks later at the medical emergency



**Fig. 1: Gangrenous feet and legs: The affected limbs are dry, hairless and hyperpigmented with dystrophic nails.**

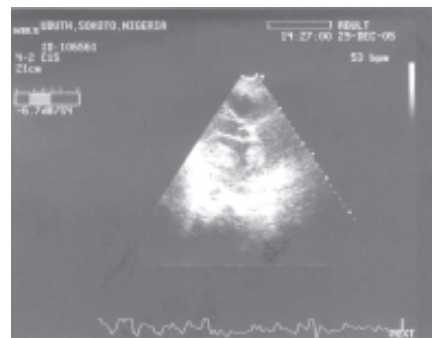


**Fig. 2: Electrocardiography showing atrial fibrillation and anteroseptal myocardial infarction. Note the flutter (“saw tooth”) waves and QS morphology in leads V<sub>1</sub> and V<sub>6</sub> and absence of P wave.**

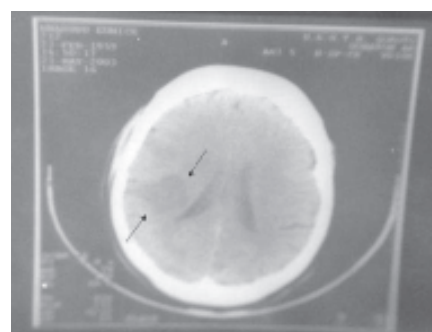
unit of Usmanu Danfodiyo University Teaching Hospital, Sokoto, Nigeria with sudden onset of chest pain, acute left ventricular failure, transient loss of consciousness, left hemiparesis, and progressively worsening numbness and pain of the lower limb extremities. She had no risk factors for coronary artery disease. She was a petty trader and had five children while her husband was a commercial vehicle driver. The distal quarters of her lower limbs were gangrenous (Figure 1) and cold with loss of sensation to touch. The dorsalis pedis arteries were impalpable. Blood glucose and lipid levels were normal.

Electrocardiography showed atrial fibrillation/flutter and old anteroseptal myocardial infarcts (Figure 2). The chest x-ray showed evidence of pulmonary congestion. Echocardiography confirmed an enlarged left atrium measuring 42 cm<sup>2</sup> in area. It contained two discrete and mobile thrombi (Figure 3). Mitral stenosis was evidenced by commissural fusion, thickened mitral valve leaflet, diminished valve excursion, and parallel motion of the anterior and posterior valve leaflets in diastole. Colour flow doppler demonstrated mitral as well as aortic regurgitant jet, and turbulence flow across the mitral valve. Left ventricular ejection fraction was 44%. Computed tomogram scan of the brain showed an irregular hypodense area compatible with an infarct in the region supplied by the right middle cerebral artery (Figure 4).

A diagnosis of rheumatic valvular heart disease complicated by multiple embolic lesions was made. Anticoagulant and antiplatelet medications were initiated with heparin and acetyl salicylic acid, respectively. Heparin was later replaced with warfarin as a long-term anti-coagulant. The ventricular rate was controlled using digoxin. Heart failure responded to diuresis and angiotensin converting enzyme inhibition. Orthopaedic surgical review was obtained and bilateral above knee amputation was done after intense counseling. Infective endocarditis and rheumatic fever prophylaxis were given using vancomycin and benzathine penicillin, respectively. Neurological recovery was complete. Facilities for coronary angiogram, embolectomy and valvular heart surgery



**Figure 3: 2-D parasternal long axis Echocardiography. The thrombi are located in the body of the left atrium. They have defined edges and are presumably fibrosed.**



**Figure 4: Computed tomogram brain scan showing hypodense area which is compatible with an infarct in the region supplied by the middle cerebral artery.**

Serial echocardiography showed that the atrial clots were presumably fibrosed and had persisted during the last four years of follow up and anticoagulation. She has, however, remained free of clinically evident embolic complication.

## DISCUSSION

It is estimated that progression to chronic valve deformity occurs in 9–39% of patients with acute rheumatic carditis. The mitral and aortic valves lesions are the most frequently encountered, accounting for 13.5% and 18.4%–36.2% of cases respectively.<sup>2,3</sup> Mitral valve disease, particularly, mitral stenosis causes left atrial enlargement which, in turn, predisposes to atrial fibrillation, stasis, increased coagulation activities and embolic phenomena. Atrial fibrillation occurs in 5.9% of patients with RHD, the prevalence being markedly higher (45.8%) in patients with mitral stenosis.<sup>4</sup> Embolic complications are more frequent in valvular than non-valvular atrial fibrillation.<sup>5</sup>

Although emboli from the heart tend to be evenly distributed, about 80% of clinically evident cardio-embolism occurs in the brain and predominantly involves the anterior circulation.<sup>6</sup> Compared to the patients in sinus rhythm, those with atrial fibrillation are at increased risk of embolic stroke. Plasma endothelin-1 has been found to be elevated in patients with rheumatic mitral stenosis complicated by cerebral embolism. Its contribution to embolic process is however not known.<sup>7</sup> Left atrial thrombus, as in the reported case, is a strong predictor of ischaemic stroke in a setting of atrial fibrillation.<sup>8</sup>

Embolic complications also frequently involve the peripheral vessels. Unlike Caucasians in whom atherosclerosis underlies peripheral embolic vascular disease,<sup>9</sup> intracardiac thrombosis resulting from atrial fibrillation of valvular aetiology, and severe systolic dysfunction are the frequent causes of acute peripheral embolic arterial occlusive lesions in the native African population.<sup>10</sup> The frequently involved sites of occlusion include the femoral, popliteal, brachial and cubital arteries. Peripheral artery embolic occlusion in the lower limb may progress rapidly to ischaemia and gangrene because of inadequate collateral blood supply.

In contrast to cerebral and peripheral arteries, coronary artery embolism is rare in RHD, the prevalence rate being between 7.5% and 12.2%.<sup>11</sup> Its frequency varies according to the valve involved: mitral valve disease (13.5%), aortic valve disease (13.5%) and combined mitral and aortic valve lesions (9%).<sup>11</sup> On the whole, valvular heart disease accounts for about 40% of coronary artery embolism and, left atrial thrombus, as in the current case, is

a frequent substrate.<sup>12</sup> Coronary artery embolism is probably under-diagnosed because of its distal location and tendency to cause infarct without detectable coronary artery abnormality on angiography.<sup>13</sup> In addition, chest pain in this setting may be absent or atypical.<sup>11-13</sup>

Late presentation and lack of diagnostic and treatment facilities make the management of embolic arterial disease very challenging in low income nations. Facilities for cardiac surgery, embolectomy, and pharmacological dissolution of clots are rarely affordable or available. Furthermore, in spite of the efficacy of anticoagulation in reducing embolic complications, compliance with anticoagulant prophylaxis is poor, even in the developed nations.<sup>14</sup> In conclusion, systemic thromboembolism is a serious complication of atrial fibrillation of valvular aetiology. Its prevention requires continuous anticoagulation. Measures aimed at primary prevention of rheumatic fever are advocated.

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