# Mycotic Abdominal Aortic Aneurysm in a Patient with Rheumatic Heart Disease: A Case Report*

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## ABSTRACT

Rheumatic heart disease is one of the most common acquired heart diseases with valvular heart damage as its hallmark. It is preventable, and the prognosis is good when caught early on. However, serious complications could arise if early detection and prompt intervention are not done. One of its dreaded complication is mycotic abdominal aortic aneurysm, an uncommon sequela that could arise from infective endocarditis. In such cases, surgical intervention is warranted due to its high risk of rupture. The case presented is a 9-year-old female with rheumatic heart disease and infective endocarditis who later developed mycotic abdominal aortic aneurysm. Manifestations were fever, joint pains, left-sided body weakness and abdominal mass. Echocardiography revealed mild aortic insufficiency with vegetations. Abdominal CT scan and CT angiogram showed a mass in the left para-aortic region and a saccular abdominal aneurysm, respectively, which confirmed the diagnosis. Antibiotics and cardiac support were given, and eventually underwent repair of aneurysm with mitral valve replacement. The aim of presenting this report is to highlight one of the rarest complications of rheumatic heart disease and infective endocarditis – mycotic abdominal aortic aneurysm.

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## INTRODUCTION

Rheumatic heart disease (RHD) is a complication of a streptococcal throat infection which causes heart valve damage and progressive heart failure if not treated. According to the 2005 summary report by World Health Organization (WHO), the estimated overall burden of RHD was 15.6 million prevalent cases with 282,000 new cases and over 233,000 deaths annually. RHD commonly affects school age children with a prevalence of 5.7 cases per 1000 school children.[1] According to the Philippine Pediatric Society (PPS-ICD 10), there are 5,874 cases of RHD from May 2016 to August 2018.[2] According to Pediatric Infectious Disease Society of the Philippines (PIDSP), the prevalence of rheumatic heart disease in school children from the year 1981-1990 is 0.6/1000 school-aged children. Complications may arise from rheumatic heart disease if not treated. According to a study done by Okello, complications occurred in 49% of newly diagnosed rheumatic heart disease cases, with heart failure (46.9%) as the most common complication, followed by pulmonary arterial hypertension (32.7%), atrial fibrillation (13.9%), recurrence of acute rheumatic fever (11.4%), infective endocarditis (4.5%) and stroke (1.3%).[3] In recent years, the incidence of pediatric stroke has reached up to 8 per 100,000 children per year.[4] One of the most frequent causes for stroke in the childhood is acquired heart disease. In the National Children's Hospital registry, only 2 cases of stroke secondary to RHD was found among the 758 cases from the year 2006-2016. Infective endocarditis occurs less often in children than in adults and accounts for 1 in 1280 pediatric admissions per year. In the National Children's Hospital registry from the year 2006-2016, 33 had infective endocarditis among 99 cases with rheumatic heart disease. Infective endocarditis is one of the predisposing factors in developing mycotic aneurysm.

Mycotic aortic aneurysm, most commonly known as infected aortic aneurysm, is an aortic aneurysm caused by an infection. The term "mycotic," although a misnomer, was coined by Sir William Osler in his *Gulstonian Lectures* in 1885 where he described a patient with valve vegetations and 4 aortic aneurysms with morphological fungal resemblance. The disease is rare with an estimated incidence of 0.6% of all aortic aneurysm in Western countries. A search of the MEDLINE database found only 41 cases of abdominal aortic aneurysm in children reported from 1975 to 2008.[5] No case was documented at National Children's Hospital from the year 2006-2016 except for the patient to be presented who was admitted in 2017. In most cases, aortic aneurysms are atherosclerotic in nature and occur most commonly in the elderly. Most of these aneurysms are of the thoracic aorta, whereas the abdominal aorta is infrequently affected by aneurysms. Although abdominal aortic aneurysm is a rare disease in children, it can be easily diagnosed by imaging studies such as CT angiogram, magnetic resonance angiography and ultrasonography. Aneurysm of the abdominal aorta is associated with high morbidity and mortality. Trauma, connective tissue diseases, granulomatous inflammation and bacterial invasion of the aortic wall accounts for a few of these aneurysms.

The aim of this report is to highlight the rare complication of mycotic abdominal aortic aneurysm which stems from the fatal sequelae of rheumatic heart disease and the importance of early prevention and treatment.

## CASE REPORT

This is a case of a 9-year-old Filipino female who came in due to fast breathing. History started 3 months prior to admission when patient had intermittent fever for 3 days with maximum temperature of $41^0$C. No other accompanying symptoms were reported. Consult was done but diagnosis was undisclosed. She was sent home with Paracetamol which afforded relief of fever. Two months prior to admission, there was recurrence of fever now accompanied with joint pains. Persistence of fever and joint pains prompted consult at a private hospital where Cefixime was given for 7 days but no improvement was noted. About 23 days prior to admission, still with intermittent fever and joint pain, the patient had abdominal pain, fast breathing and pallor. Consult was done at Baler Hospital where he was admitted for 4 days. She was transfused with blood, and unrecalled antibiotics were given. Patient was discharged still with fast breathing. Six days prior to admission, she developed weakness of the left upper and lower extremities. Persistence of weakness on the left side of the body and fast breathing prompted consult at Casiguran District Hospital. He was transferred to our institution for further evaluation.

Review of systems showed weight loss, body malaise and constipation. Patient had no history of trauma or surgery. There was family history of heart disease on maternal side. Claimed to have complete immunization.

At the emergency room, patient was awake, pale-looking and in cardiorespiratory distress. Patient was tachycardic at 136 beats per minute, tachypneic at 64 cycles per minute, afebrile and with oxygen saturation of 90% at room air. When hooked to oxygen support via nasal cannula at 2 liters per minute, oxygen saturation increased to 98%. Patient weighed 14 kg with a z score <-3 (severely underweight), height was 118 cm with a z score of <-2 (wasted). Other pertinent physical findings were pale palpebral conjunctivae, multiple dental caries, palpable cervical lymph nodes, distended neck veins, harsh breath sounds, dynamic precordium, tachycardia with Grade 4/6 murmur, direct tenderness on the right abdominal lower quadrant area, liver edge palpable 3 cm below the right subcostal margin, clubbing of finger nails, and with grade 1 non-pitting edema on the lower extremities. Neurologic examination showed a patient who was conscious, coherent, with left facial asymmetry and left tongue deviation. Motor examination of the left extremities showed 0/5 or no movement with decrease in bulk and tone. All extremities were equally sensitive to light touch and pressure with normal deep tendon reflex.

Patient was diagnosed as Rheumatic Heart Disease in failure, to consider Stroke. Complete blood count *(Table 1)* showed persistent anemia and leukocytosis with neutrophil predominance. C-reactive protein (12 mg/L) and lactate dehydrogenase (521 U/L) levels were elevated. Bleeding parameters, serum electrolytes, liver enzymes, urinalysis, fecalysis and anti-streptolysin O titer were within normal range. Blood culture on two sites showed no growth. Arterial blood gas done showed respiratory alkalosis with pH 7.552, $pCO_2$ 51, pO2 51, $HCO_3$ 18.9, base excess -3.5. Electrocardiography (ECG) showed sinus tachycardia with right atrial deviation, biatrial and biventricular enlargement.

**Table 1. Complete Blood Count Series**

| Parameters | Results | | | | | | |
|---|---|---|---|---|---|---|---|
| | 01-17-17 | 01-19-17 | 01-26-17 | 02-01-17 | 02-13-17 | 02-24-17 | 02-28-17 |
| Hemoglobin | 85 | 97 | 88 | 103 | 99 | 91 | 119 |
| Hematocrit | 0.28 | 0.32 | 0.28 | 0.32 | 0.31 | 0.29 | 0.36 |
| White Blood Cells | 23.3 | 15.8 | 10.5 | 13.1 | 11.6 | 11.1 | 12.4 |
| Segmenters | 0.81 | 0.82 | 0.93 | 0.60 | 0.74 | 0.63 | 0.66 |
| Lymphocytes | 0.19 | 0.13 | 0.05 | 0.32 | 0.20 | 0.25 | 0.26 |
| Eosinophils | - | - | - | 0.02 | 0.01 | 0.07 | 0.04 |
| Monocytes | - | 0.05 | 0.02 | 0.06 | 0.05 | 0.05 | 0.04 |
| Platelet | 300 | 268 | 262 | 393 | 350 | 300 | 314 |

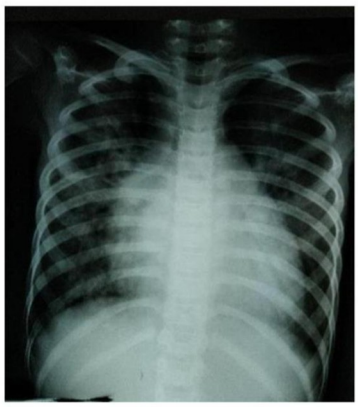

*Figure 1.* Chest Radiograph showed markedly enlarged heart with pulmonary congestion and/ or pneumonia. Diaphragm and sinuses were intact

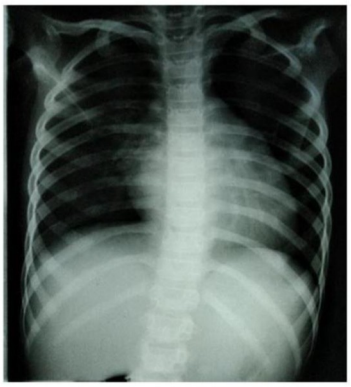

*Figure 2.* Repeat Chest radiograph revealed *slight decrease in the transverse diameter and the heart with marked resolution of the pulmonary congestion and/ or pneumonia*

Chest radiograph *(Figure 1)* showed markedly enlarged heart with pulmonary congestion and or pneumonia. Repeat chest radiograph *(Figure 2)* after 10 days revealed slight decrease in the transverse diameter of the heart with marked resolution of the pulmonary congestion and/ or pneumonia.

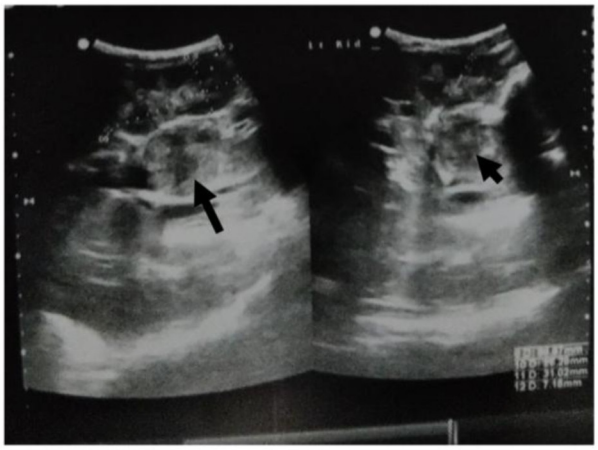

*Figure 3* Whole abdominal ultrasound showed 6.5 x 3.3 x 4.3 cm heterogenous isoechoic mass lesion *(black arrow)* seen in upper abdomen (aortocaval region) with no demonstrable intralesional vascularities to consider large aortocaval lymph node, neoplastic process cannot be ruled out. Normal sonogram of liver, bile ducts, gallbladder, abdominal aorta, pancreas, spleen, kidneys and urinary bladder.

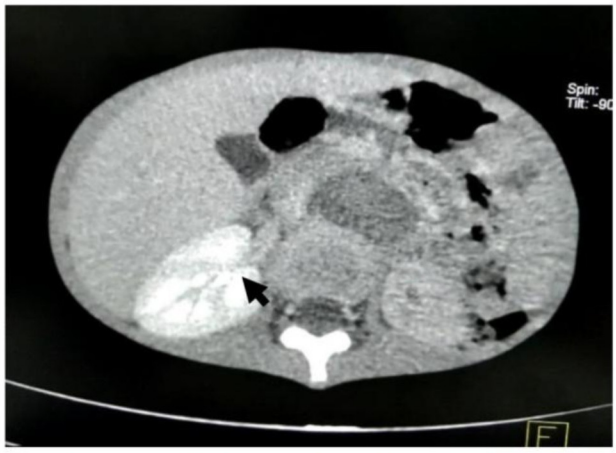


*Figure 4.* Abdominal CT Scan: oval-shaped mass *(black arrow)* in the left upper para-aortic region measuring 2.6 x 4.2 x 4.2 cm closely related to the abdominal aorta consistent with a thrombosed abdominal saccular aneurysm probably mycotic seen immediately below the superior mesenteric artery rather than abscess. Hypertrophied right kidney with minimal pelvocaliectasis and a small area of infarct or inflammation. Minimal to moderate hepatomegaly.

Whole abdominal ultrasound (WAB) *(Figure 3)* was done which showed 6.5 x 3.3 x 4.3 cm heterogenous isoechoic mass lesion in the upper abdomen to consider large aortocaval lymph node; neoplastic process cannot be ruled out. It was also during this time that on physical examination a palpable irregular mass at the epigastric area was noted.

2d echo revealed rheumatic heart disease; thickened mitral valve leaflets; mild aortic insufficiency and trivial pulmonic insufficiency; good systolic function with ejection fraction of 75% with 3 vegetations at the mitral with in and out motion across the valve, one at the sub-valvar apparatus of the mitral valve and one at the papillary muscle area; with sub valvar membrane noted at the septum below the aortic valve with bridging connection to the papillary mitral stenosis. There was no pericardial effusion. Pulmonary arterial pressure was normal. Abdominal CT scan (*Figure 4*) showed oval-shaped mass in the left upper para-aortic region measuring 2.6x4.2x4.2cm.

The opacified aorta appears encased by the mass which may represent an abdominal thrombosed saccular aneurysm probably mycotic seen immediately below the superior mesenteric artery. Cranial CT scan showed multiple small areas of slightly hyperdense change with contrast enhancement in the right basal ganglia, right parasylvian area and left splenium corpus callosum consistent with subacute infarcts with disrupted blood brain barrier and lucency perfusion changes. This is secondary to cardio-embolic phenomenon related to rheumatic heart disease. Small area of chronic and ischemic changes. Irregularities of the right midcerebral artery and branches. Minimal ventricular and sulcal dilatation secondary to cerebral atrophy.

CT angiogram of the abdominal aorta done later showed saccular abdominal aneurysm seen just below the level of the superior mesenteric artery extending down the level just before the inferior mesenteric artery, approximately measuring 5.3 x 6.6 x 6.5 cm. There is associated thrombus formation with maximum thickness of 1.6cm and effective luminal diameter of 4.4cm.

Patient was hydrated at BSA 500cc/day. She was given Dobutamine drip at 10mcg/kg/min, Penicillin G (200,000mkd), Gentamicin (7.5 mkd), Captopril (0.5mkd), Furosemide (1 mkdose) and Citicoline 200mg once a day. In the ward, she had 1 episode of melena and later had icterisia. She also continued to be afebrile with occasional abdominal pain radiating to the back and weakness on the left side of the body. Patient was referred to Infectious Service because of mycotic abdominal aneurysm and fluconazole was started. On the 25$^{th}$ hospital day, aspirin was also given for subacute infarcts as seen in the cranial CT scan. Patient completed Gentamicin and Penicillin for 42 days and was transferred to Philippine Heart Center (PHC) for further evaluation of the aortic aneurysm.

At the Philippine Heart Center, patient underwent repair of abdominal aortic aneurysm with mitral valve replacement and was discharge improved. Patient was lost to follow-up and last seen at the PHC OPD on July 2017.

## CASE DISCUSSION

Based on the two months history of fever and joint pains accompanied with weight loss, body malaise, fast breathing and physical examination of multiple dental caries, tachycardia, distended neck veins, dynamic precordium, grade 4/6 murmur, hepatomegaly and bilateral non-pitting edema of lower extremities, rheumatic heart disease was highly considered. This was supported by the findings seen on 2d echo. Based on the 2015 Jones criteria, the patient was diagnosed to have RHD in the presence of 1 major (carditis) plus 2 minor (fever, elevated CRP and monoarthralgia) criteria.[6] The presence of one-week history of sudden left-sided weakness of the body points to a diagnosis of a stroke. Aside from the physical and neurologic examination, this was supported by the findings on Cranial CT scan which showed subacute infarcts secondary to cardio-embolic phenomenon. Multiple risk factors may cause stroke in children. These are arteriopathies, heart conditions, hematologic disorders or inborn error of metabolism. In our case, the presence of rheumatic heart disease and infective endocarditis poses higher risk of developing stroke. History of fever in a rheumatic heart disease with 2d echo findings of vegetations supports the diagnosis of infective endocarditis. According to Duke Criteria, for a diagnosis of endocarditis, 1 major (presence of vegetations in 2d echo) and 3 minor criteria (fever, rheumatic heart disease, vascular phenomenon: aneurysm and intracranial bleed) are needed which were fulfilled in the patient's case.

Rheumatic heart disease is one of the most common and preventable acquired heart diseases with valvular heart damage as the hallmark. It results from an autoimmune inflammatory reaction to throat infection caused by Group A streptococci. It commonly occurs in childhood and can lead to death and life-long disability. The best treatment is to prevent rheumatic fever from occurring. Antibiotic is given to treat the strep infection and patients need to take penicillin possibly for life, to prevent recurrences and lower the risk of heart damage.

Infective endocarditis usually presents with prolonged fever, weight loss, diaphoresis, and myalgias which are secondary to ongoing

bacteremia, valvulitis, embolic and immunological phenomenon. Vegetations may embolize and produce symptoms of ischemia or hemorrhage of the organ involved. In this case report, embolization occurred in the abdominal aorta.

Cardioembolic stroke is diagnosed when sudden focal neurologic signs, maximal at onset, developed in patients with peripheral systemic embolism and recent myocardial infarction or rheumatic mitral stenosis.[7] There are three basic groups of causes of cardioembolic strokes: (1) cardiac wall and chamber abnormalities (2) valve disorders and (3) arrhythmias. According to Leary & Caplan, among patients with rheumatic mitral valve disease, brain embolism is seen in 7% of patients with mitral insufficiency. Strokes can have a wide range of different clinical manifestations such as sudden hemiparesis, hemisensory loss, confusion, trouble speaking, difficulty understanding, visual loss, diplopia, ataxia, vertigo or even sudden severe headache with no known cause.[8] There is high incidence of acute midcerebral artery blockage in patients with sudden onset hemispheric strokes. There is limited evidence regarding the efficacy of secondary prevention strategies in childhood stroke however, aspirin is widely used.

In 1885, Osler coined the term mycotic aneurysm to describe infected aneurysm. The term "mycotic" is a misnomer and did not imply fungal etiology. Its classical description is infective aneurysms secondary to rheumatic endocarditis. Most of infected aneurysms were due to infective endocarditis due to streptococcal infection.[9] Pathology of aortic infection can be explained by several theories. Microemboli may lodge in the vasa vasorum causing occlusion and damage to the aortic wall leading to degeneration and aneurysm formation. It can also be due to direct inoculation into the arterial wall.

According to Sorelius & Summa, for the past 15 years, various combination of the following 4 criteria had been used to diagnose mycotic aortic aneurysm: (1) clinical presentation such as pain, fever, concomitant infection, elderly patient with cardiovascular disease and or immunosuppressive state; (2) laboratory findings of increase inflammatory parameters such as C-reactive protein, leukocytosis and positive culture; (3)radiological findings on computed tomography or magnetic resonance imaging either saccular, eccentric, multilobular aneurysm, periaortic mass, periaortic gas and rapid aortic expansion, and (4) intraoperative findings. Presence of one of the criteria is not sufficient to make a diagnosis. The patient presented with clinical manifestation of abdominal pain, fever with concomitant infection, leukocytosis, elevated CRP and radiological finding on CT scan of abdominal saccular aneurysm. Once abdominal aneurysm is diagnosed, surgical treatment is required because of the risk of rupture.[10]

Abdominal aortic aneurysm is rare in children and various causes have been identified such as connective tissue disorders, arteritis, congenital, iatrogenic, post traumatic and mycotic disorders. Most common causes are infectious-inflammatory, associated with known genetic disease, idiopathic and following trauma, all of which are life-threatening. Treatment of abdominal aortic aneurysm centers around control of hypertension and resection of aneurysm with reconstruction.

Eradication of infection and safe establishment of arterial flow are the goals of management of mycotic aortic aneurysm. The gold standard for treatment of mycotic aortic aneurysm is resection, debridement of infected tissues and graft replacement.[11] A minimally invasive option is endovascular therapy. The main goal of surgical treatment is considered after completion of the antibiotics for 4-6 weeks. Often the surrounding tissue is infected with bacteria that it is necessary to do extensive resection. Unfortunately, mortality rate is 40% after open surgical repair.

## CONCLUSION

Early diagnosis and prompt treatment can prevent serious life-threatening sequelae of Rheumatic Heart Disease. Diagnostic criteria such as the Duke and Jones criteria with the aid of echocardiography play a major role in the initial diagnosis of these conditions. Prolonged treatment and appropriate prophylaxis are needed to ensure cure and prevent recurrence. Mycotic abdominal aortic aneurysm is a rare complication of infective endocarditis. Early diagnosis is important and

surgical intervention is often needed for effective treatment due to its high incidence of rupture and death. Better understanding of the disease epidemiology, case detection, pathogenic mechanism, primary and secondary prevention may control and reduce incidence of rheumatic heart disease.

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