invasive pulmonary aspergillosis, septic embolisation or direct extension from the lungs [2]. Ascending aorta or the aortic arch are the most likely locations [1]. *Aspergillus fumigatus* is isolated more frequently, mainly affecting patients on immuno-suppressant therapy [4].

Clinical features of a mycotic aortic aneurysm are non-specific and can vary from septicaemia to manifestations secondary to distal embolisation. The diagnosis requires a high clinical suspicion, given its rarity and the presence of vague symptoms [5].

When clinically apparent, infected aneurysms are often at an advanced stage of development or are associated with complications such as rupture. Non-treatment or delayed treatment of infected aneurysms has a poor outcome, due to fulminant sepsis or haemorrhage. Multi-detector computed tomography and magnetic resonance imaging have replaced conventional angiography as minimally invasive techniques for detection of infected aneurysms [6]. The treatment of a mycotic aneurysm is wide resection of the infected aorta and grafting followed by long term antifungal treatment. However, hospital mortality rate following surgery approaches 40% [7]. Endovascular stent-graft repair can be performed in selected cases. In our patient oral prednisolone given to treat a suspected vasculitis probably aggravated the undetected fungal infection, leading to rupture of the aneurysm. Why our patient became immuno-compromised to develop disseminated Aspergillosis remains unclear.

**References**


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**A young woman with hypergammaglobulinemia, distal renal tubular acidosis and some clinical features of polymyositis**

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(Index words: renal tubular acidosis, polymyositis, hypokalaemia)

**Introduction**

Renal tubular acidosis (RTA) is a disorder of renal acidification of urine due to defective functioning of nephrons. It can be associated with several disease entities including some autoimmune syndromes. We report here a case in which distal RTA is associated with biopsy confirmed polymyositis (PM). So far there is only one case report regarding this association.

**Case report**

A 19-year old female was admitted to North Colombo Teaching Hospital with muscle pain and weakness for 1 week. She had no eye involvement or difficulty in swallowing. There was tenderness over the proximal muscles of thighs and arms on both sides. She had proximal muscle weakness with the power of 3/5 in each limb and in neck. Rest of the physical examination was

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unremarkable. There were no other features of connective tissue disorders.

Serum potassium was found to be persistently low (1.5 mmol/l, 1.8 mmol/l, 2 mmol/l) and serum sodium was 135 mmol/l and chloride 116 mmol/l. Arterial blood gas analysis revealed pH of 7.154, pCO₂ of 3.17 kPa, pO₂ of 14.8 kPa, HCO₃ of 8.5 mmol/l and O₂ saturation of 98%, suggesting normal anion gap metabolic acidosis. Urine pH was 8 with no glycosuria. Urinary pH of more than 5.5 in the presence of severe acidosis suggested RTA.

Investigations showed normal blood counts, blood urea, fasting blood sugar, serum creatinine and serum bilirubin. Serum creatine phosphokinase (CPK) was initially elevated (8723 iu/l). Aspartate aminotransaminase (>150 iu/l) and alanine aminotransaminase (183 iu/l) were elevated. EMG on right quadriceps was compatible with polymyositis. Muscle biopsy showed focal lymphocyte infiltration with muscle fibre destruction and atrophy. Perivascular lymphocyte infiltration was apparent and there was evidence of pericapsular muscle atrophy (Figure 1). These histological features were consistent with polymyositis.

![Figure 1. Muscle biopsy showing focal and perivascular lymphocyte infiltration with muscle fibre destruction and atrophy.](image)

Erythrocyte sedimentation rate (ESR) was around 100 in the 1st hour on several occasions. Albumin/globulin ratio was reversed (34/45 g/l) and on electrophoresis, globulin was of polyclonal origin. Serum autoantibody studies rendered no positive result (anti-nuclear factor, double stranded DNA, rheumatoid factor, anti-smooth muscle antibodies and anti-Jo-1). Chest X-ray, X-ray KUB, Mantoux test, Coomb’s test, echocardiogram and the ultrasound scan of the abdomen did not reveal any abnormal results.

The patient improved markedly after intravenous potassium chloride (oral potassium chloride later) and oral sodium bicarbonate. Steroids or other immunosuppressives were not given. However CPK level became normal in 7 days. ESR remained high months after the clinical recovery. Oral bicarbonate and potassium replacement were continued and titrated according to blood gas analysis and serum potassium level.

At the beginning we did not consider treating her with immunosuppressive drugs for polymyositis as her condition improved with treatment for RTA. Our plan was to follow up with regard to high ESR, increased gamma globulin and to repeat the muscle biopsy to see whether the muscle inflammation continued. However, the patient stopped attending for follow up after a few months.

**Discussion**

RTA is a disorder of renal acidification of urine. In distal RTA there is a secretory defect or a permeability defect of the distal nephron. This defect results in inability to lower the urine pH to less than 5.5 despite metabolic acidosis. There is associated hypokalaemia. Distal RTA may be a primary disease or associated with autoimmune disorders such as hypergammaglobulinemia, cryoglobulinemia, Sjögren syndrome, thyroiditis, pulmonary fibrosis, chronic active hepatitis, primary biliary cirrhosis, systemic lupus erythematosus, and vasculitis [2].

PM is an idiopathic, non-infectious, inflammatory myopathy. Our patient met all criteria to diagnose PM. Bohan and Peter criteria of polymyositis include symmetrical proximal muscular weakness, elevated serum muscle enzyme levels, electromyographic evidence of myopathic abnormalities and characteristic findings at muscle biopsy. Severe hypokalaemia can produce muscle paralysis, similar histological features and rhabdomyolysis, causing elevation of muscle enzymes, but markedly elevated PCK level along with typical muscle biopsy findings confirm PM. However improvement of muscle weakness and CPK levels in the absence of immunosuppressive therapy is not explainable.

Distal RTA is rare as an isolated entity, and more commonly it is associated with systemic autoimmune or connective tissue disease. Association of PM with systemic lupus erythematosus, rheumatoid arthritis, Sjögren syndrome, systemic sclerosis and mixed connective tissue disease is well documented. But association of RTA and PM is not described. Persistent high ESR with polyclonal increase in gamma globulin suggests an underlying inflammatory process that is still ill-defined. Taken separately, the patient satisfied the diagnostic criteria of both polymyositis and distal RTA as the cause of proximal myopathy.

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Congenital pulmonary arteriovenous malformation: A rare cause of cyanosis in childhood

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(Index words: pulmonary arteriovenous malformation, cyanosis, right to left shunting)

Introduction

Pulmonary arteriovenous malformations (PAVMs) are caused by abnormal communications between pulmonary arteries and pulmonary veins, which are most commonly congenital in nature. Although these lesions are uncommon, they are an important part of the differential diagnosis of common pulmonary problems such as hypoxaemia, pulmonary nodules and cyanosis.

PAVM is a rare disorder with an incidence of 2-3 per 1,000,000 population [1]. It occurs twice as often in women as in men, but there is a male predominance in newborns. Around 10% cases of PAVM are identified in infancy or childhood, followed by a gradual increase in the incidence through the fifth and sixth decades. Approximately 70% of the cases of PAVM are associated with hereditary haemorrhagic telangiectasia (HHT). Conversely, about 15 to 35% of patients with HHT have PAVM. PAVMs may cause hypoxaemia and dyspnoea due to right to left shunting, but frequently remain undiagnosed. This intrapulmonary malformation is described in two patients who presented with severe cyanosis.

Case reports

Patient 1

A 6-month-old male infant was found to have a cardiac murmur and cyanosis of one day. He was born at term after a normal pregnancy and without prenatal complications. His family history was unremarkable and specifically negative for cardiopulmonary disorders. He had a dusky colour, but was relatively well and thriving. His mother related a history of orthodeoxia with worsening cyanosis when the infant was embraced, while the cyanosis improved when lying back. Cardiovascular examination showed bounding peripheral pulses in both upper and lower limbs, normal heart sounds and a continuous machinery murmur. The murmur was best heard at the left upper sternal border suggesting a patent ductus arteriosus (PDA). There was normal vesicular breathing. The examination of abdomen, central nervous system, skin and mucosa did not reveal any abnormality.

The arterial blood gas analysis (room air) showed a pH of 7.23, PCO₂ of 5kPa, HCO₃⁻ of 17 mmol/l, and O₂ saturation of 61%. Chest X-ray and ECG were normal. The haemoglobin was 11.8g/dl with normal methaemoglobin of 1.2%.

Echocardiography confirmed the presence of a PDA, but it could not account for the cyanosis. Video assisted thoracoscopic surgery (VATS) for closure of PDA was done. As expected the PDA closure did not resolve the cyanosis and early clubbing. Therefore contrast echocardiographic study was performed. It suggested the presence of a PAVM with significant right to left shunt, as evidenced by rapid filling of the left atrium with dissolved bubbles. Cardiac catheterisation showed the pulmonary arteriovenous malformation in postero-basal segment of lower zone of the left lung (Figure1), with normal pulmonary artery pressure of 15/5mmHg. Oxygen saturation of the aorta was 64%, which made him a candidate for catheter-based intervention.

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