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CASE REPORT

ATYPICAL MANIFESTATIONS OF ACUTE POSTSTREPTOCOCCAL GLOMERULONEPHRITIS A REPORT OF TWO CASES

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ABSTRACT

Acute Post streptococcal Glomerulonephritis (APSGN) is an important renal disease occurring in children. Typical clinical manifestations include rapid onset of gross haematuria, edema and hypertension resulting from a prior group A - beta hemolytic streptococcal infection. However, patients with APSGN sometimes present with atypical clinical manifestations often leading to delayed diagnosis or misdiagnosis. Recognition of these unusual manifestations is important so that patient receives adequate treatment with decreased morbidity. We are reporting two cases of APSGN who presented with atypical manifestations. One case presented with generalized edema, respiratory distress and pulmonary edema with congestive heart failure and another with fever, headache, seizures and altered sensorium. Both patients had hypertension. Urinalysis and other investigations led to diagnosis of APSGN with pulmonary edema and heart failure in first case and APSGN with Posterior Reversible Encephalopathy syndrome in second.

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INTRODUCTION

Acute Post streptococcal Glomerulonephritis (APSGN) is an important renal disease occurring in children resulting from a prior infection with group A β-hemolytic streptoccus (GAS) (Ahn, 2008). APSGN mainly affects children between 5 to 12 years. Pathogenesis of APSGN involves glomerular deposition of nephritogenic streptococcal antigen and subsequent formation of immune complexes *in situ* and/or the deposition of circulating antigen-antibody complexes. Potential nephritogenic antigens include nephritis-associated plasmin receptor (NAPlr) and streptococcal pyogenic exotoxin B (SPEB) (Bagga, 2016). Typical clinical manifestations include an acute onset with gross hematuria, edema and hypertension and moderate proteinuria (acute nephritic syndrome) occurring 1 to 2 weeks after an antecedent streptococcal pharyngitis or 3 to 6 weeks after a streptococcal pyodrema (Ahn, 2008; Rodriguez-Iturbe, 2009). However patients with PSGN sometimes present with unusual clinical symptoms often leading to delayed diagnosis or misdiagnosis resulting in delay in appropriate treatment and thus increased morbidity.

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Delay in diagnosis is more common in children who may not give history of antecedent GAS infection and have microscopic hematuria (Kim, 2016).

Case Reports

Case 1 – A 7 year old female child presented with 3 days history of swelling over face progressing to abdomen and lower limbs and difficulty in breathing. On examination – Child was conscious, oriented, edematous with PR-100 bpm, RR- 30/min, BP – 140/98 mm of Hg (>99th percentile +5mm). Systemic examination showed bilateral basal crepts and hepatomegaly. Provisional diagnosis of congestive cardiac failure probably due to myocarditis was made. However, there was no h/o any preceding viral or bacterial illness. Chest X ray showed bilateral proximal lung field opacities (Bat Wing Opacites) s/o pulmonary edema (Fig.1A). 2D Echo was normal. In view of pulmonary edema and hypertension, urinalysis done which showed dark yellow colour with protein 2+, Blood 3+. Microscopy showed many RBCs with 12-14 WBCs. Further investigations showed antistreptolysin-O(ASO) titre >200 IU/ml, Complement C3 levels- 30.7mg/dl (normal 90-120mg/dl). Complete blood counts, renal and liver function tests CPK-MB, ESR and CRP were normal. She was diagnosed to have APSGN with pulmonary edema and heart failure.

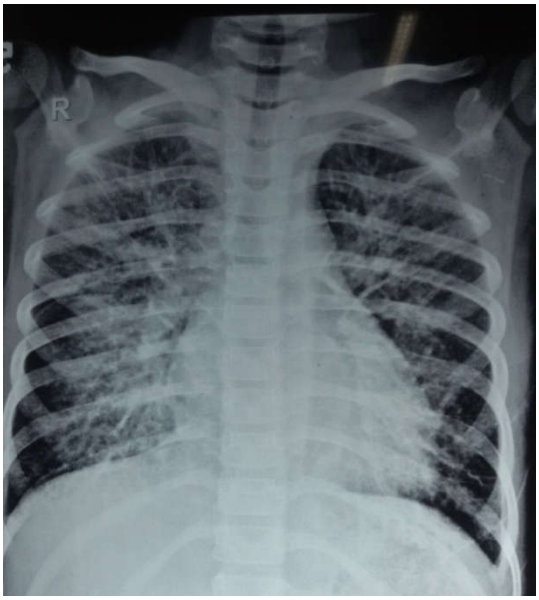

Fig.1A. Chest X-ray showing pulmonary edema in case1

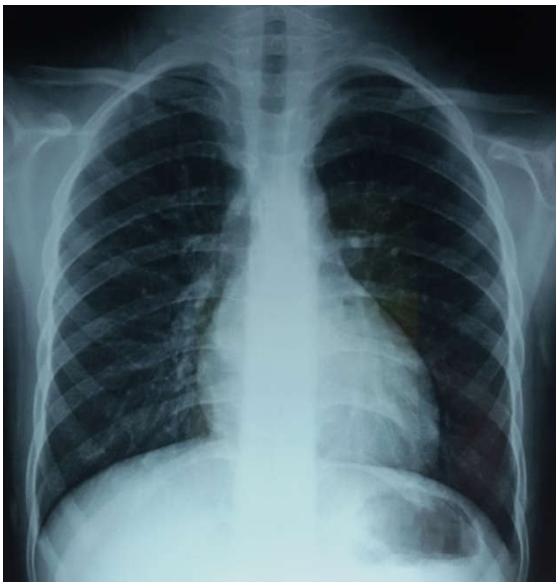

Fig.1B. Repeat chest x-ray after 6 days showing clearance in case 1

She was managed with diuretics and antihypertensive. Repeat chest x-ray done after 6 days showed no pulmonary edema (Fig.1B). Case 2 – 11 year old male child presented with complaints of fever and headache for 7 days and vomitings and seizure (GTCS type) on admission day. On Examination- Child had altered sensorium with glassgow coma score E2V2M4 = 8/15 and BP- 170/110 mm of Hg (>99th percentile +5mm) with no cranial or focal neurological deficit. Motor examination showed decreased tone with normal reflexes and there were no signs of meningeal irritation. Possibility of acute encephalitis syndrome with inceased intracranial tension kept and management started as per protocol. After stabilization, CSF exam done which was normal. Despite treatment hypertension persisted so urinalysis done which showed light yellw colour with 1+ proteins and 20-30RBCs/hpf with 2-3 WBCs/hpf.

Further investigations showed ASO titre >200 IU/ml, C3 levels- 17.3mg/dl. MRI Brain revealed edema in subcortical white matter of bilateral upper fronto-parieto-occipital region and cerebellar white matter consistent with diagnosis of Posterior Reversible Encephalopathy Syndrome (PRES) (Fig.2).

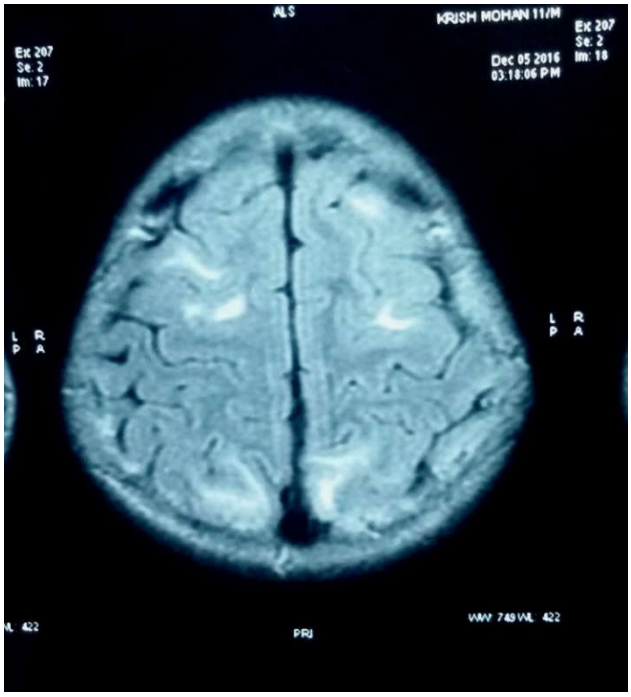

Fig.2. MRI scan showing white matter edema in bilateral fronto-parieto-occipital regions in case 2

Initial CBC showed leukocytosis with neutrophilia which normalized later. Serum transaminases were mildly elevated however renal function tests and lipid profile were normal. Investigations for enteric fever and malaria were negative. Diagnosis of APSGN with PRES was made. He was managed with antihypertensives only and other thearapies withdrawn. In both cases, urine and blood cultures were sterile and renal ultrasound and doppler studies were normal. Both patients were reviewed after 3 weeks, were asymptomatic with well controlled blood pressures on enalapril.

DISCUSSION

Although most patients with PSGN present with classical triad of gross hematuria, edema and hypertension, sometimes patient may present with atypical manifestations. Delay in diagnosis is common especially if visible hematuria is not a presenting complaint (Pais *et al*., 2008). Both of our cases did not give history of gross hematuria. PSGN presenting with symptomatic pulmonary edema and respiratory distress is uncommon. Heart failure and pulmonary edema are thought to result from hypertension or hypervolemia. Patient usually presents with dyspnea and restlessness. Chest X ray shows mild cardiomegaly with features of pulmonary edema. Often the condition is misdiagnosed as bronchopneumonia or myocarditis (Bagga *et al*., 2016; Chiu *et al*., 2004) Posterior Reversible Encephalopathy syndrome (PRES), also known as reversible posterior leukoencephalopathy syndrome (RPLS) is a clinicoradiological entity first described in 1996 by Hinchey *et al*. (1996) Clinical featuers include headache, nausea/vomiting, impaired consciousness, seizures and visual loss including cortical blindness. MRI shows white matter edema mostly in posterior parieto-temporal-occipital regions of the brain. Occasionally edema has been described in frontal lobes, basal ganglia, cerebellum, brainstem in posterior fossa

and cortical grey matter. PRES occurs in 5% to 10% of children hospitalized with acute glomerulonephritis of all etiologies, but exact prevalence of PRES associated with APSGN is unknown.

Although the underlying pathophysiology of PRES remains elusive, three theories have been proposed: 1) hypertension-induced breakdown in cerebral auto regulation; 2) cerebrovascular endothelial dysfunction and; 3) vasoconstriction and hypoperfusion with subsequent ischemia and vasogenic edema (Fugate *et al*., 2010). The preferential involvement of the posterior brain in PRES may be caused by its relative paucity of sympathetic innervation in comparison to the anterior circulation. The outcome of PRES is generally favorable, but delay in initiating the appropriate treatment may result in permanent damage to the brain (Fux *et al*., 2006). Other atypical manifestations of APSGN described in various case reports include hemolytic uremic syndrome (HUS), thrombotic microangiopathy (TMA), nephrotic syndrome, APSGN with minimal urinary abnormalities. Other immune mediated diseases described to occur rarely in concurrence with PSGN include acute rheumatic fever (ARF), post streptococcal reactive arthritis (PSRA), vasculitis (cutaneous vasculitis mimicking henoch –schonlein purpura, cerebral vasculitis, polyarteritis nodosa), immune thrombocytopenic purpura (ITP), autoimmune hemolytin anemia (AIHA) and diffuse alveolar haemorrhage (Toru Watanabe, 2011)

Conclusion

Thus we conclude that in children presenting with dyspnea and edema with chest radiograph showing features of pulmonary edema, proper evaluation including blood pressure recording and urinalysis should be performed immediately. Also possibility of Acute glomerulonephritis and PRES should be primarily considered in differential diagnosis of children with headache, seizures and impaired consciousness with hypertension. Prompt diagnosis and early antihypertensive therapy may avoid unnecessary therapeutic intervention and prevent morbidity.

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