Case Report

An Atypical Manifestation of Post Streptococcal Glomerulonephritis

Uddalak Chakraborty[1], Tarun Kumar Paria[2], Purbasha Biswas[2], Tanuka Mandal[3], Atanu Chandra[4]

Acute post streptococcal glomerulonephritis is a relatively common entity encountered by the internist in day to day practice. Aside the relatively common presentation of hematuria, oliguria and facial puffiness, one may present with convulsions and hypertensive encephalopathy. A 19 year old male patient was admitted with generalized tonic clonic seizure and elevated blood pressure, with some papular rash in different stages of healing in both lower limbs, MRI brain revealed bilateral symmetrical hyperintensities in frontal and parieto-occipital areas which resembled posterior reversible encephalopathy syndrome (PRES) like changes. The evaluation of hypertension in the young boy revealed microscopic hematuria and nephritic range proteinuria with decreased serum complements suggestive of acute glomerulonephritis. Symptomatic management ensured a complete recovery without any neurological deficit and resolution of MRI findings over a week. PRES is a rare clinico-radiological entity and may be a presenting symptom in patients with post streptococcal glomerulonephritis.

[*J Indian Med Assoc* 2020; **118(6):** 58-9]

***Key words*: Post-streptococcal glomerulonephritis; PRES; hypertension; convulsions.**

Acute post streptococcal glomerulonephritis (PSGN) is a nephritic syndrome which may present commonly with oliguria, cola colored urine, puffiness of face and eyelids, pedal edema, and hypertension. Apart from these classical symptoms, PSGN may present with atypical manifestations like myocardial dysfunction, acute renal failure, etc. Reversible encephalopathy due to dysfunction of cerebral autoregulation has been reported in children and juvenile patients with PSGN[1]. An adult patient presenting with reversible encephalopathy and posterior reversible encephalopathy syndrome like changes in MRI brain due to PSGN is relatively rare.

Case Report

A 19 year old male patient presented to us in September, 2019 with his first episode of generalized tonic clonic seizure preceded by headache and nausea. He had a blood pressure of 160/100mmHg and complained of persisting headache and nausea, followed by which he again suffered another episode of generalized tonic clonic convulsion. Clinical examination revealed only a papular rash in both lower limbs in different stages of healing, which was accompanied by some pustular discharge from the rash 2 weeks prior to the current episode. Systemic examination did not reveal any abnormality.

A subsequent MRI brain revealed bilateral symmetrical hyperintensities in FLAIR (fluid attenuation inversion recovery) sequence in frontal and parieto-occipital areas (Figs 1,2), which predominantly involved the white matter. Further evaluation of hypertension in such a young male patient revealed microscopic hematuria with 8-9 RBC/ hpf with few dysmorphic RBC and pus cells 2-3/hpf with trace albuminuria in routine urinalysis, with negative cultures from urine. Urine output was around 700ml in 24 hours. An urinary albumin-creatinine ratio revealed a nephritic range proteinuria with 1685 mg/mcg (n=30-300mg/mcg). Serum complements revealed a diminished C3 level around 26mg/dl(n= 66-185mg/dl)with a C4 level in the normal range. Ultrasonographic imaging of kidneys did not reveal any abnormality. ASO titre was found to be raised around 600 units/ml (n<200 units/ml) and Anti DNAse B levels were raised as well around 300 units/ml (n<85 units/ml). Renal function tests were unaltered. All other routine investigations were non-contributory.

The patient was admitted with a hypertensive emergency and characteristic signal changes in brain imaging. On an

Editor's Comment :

- PSGN may present with hypertensive encephalopathy apart from the common symptoms of nephritic syndrome.
- PRES is a characteristic finding in brain imaging usually associated with hypertensive emergencies due to failure of cerebral autoregulation.
- PRES as a presentation of PSGN though seen commonly in children, may also be seen in young adults as in our case.

[1]MBBS, Postgraduate trainee, Department of General Medicine, RG Kar Medical College and Hospital, Kolkata and Corresponding Author

[2]MBBS, Post graduate trainee, Department of General Medicine, RG Kar Medical College and Hospital, Kolkata

[3]MD, MACP, Senior Resident, Dept of Medicine, RG Kar Medical College Hospital, Kolkata

[4]MBBS, Assistant Professor, Department of General Medicine, RG Kar Medical College and Hospital, Kolkata

Received on : 05/06/2020

Accepted on : 15/06/2020

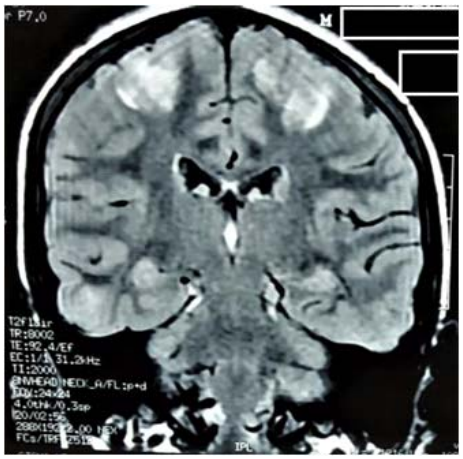

Fig 1 — A T2 Flair MRI showing bilateral frontal and occipital hyper intensities

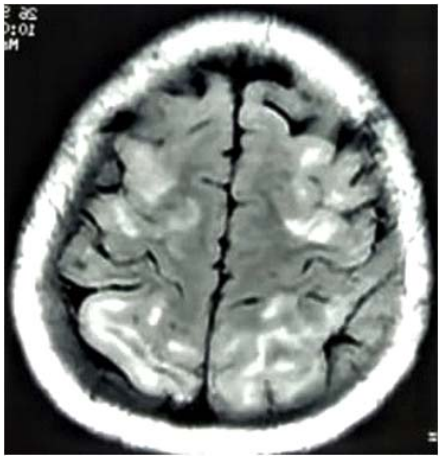

Fig 2 — A T2 Flair MRI showing bilateral parietal and occipital hyper intensities

attempt to work out the cause of hypertension in the young individual, urinalysis revealed microscopic hematuria with dysmorphic RBC and microalbuminuria with low urine output. Decreased C3 levels led along with the supportive findings led us to the diagnosis of Acute glomerulonephritis in the background of healing skin lesions, probably a post streptococcal sequale as evidenced by raised ASO and Anti DNAse levels.

The patient was diagnosed as a case of acute post streptococcal glomerulonephritis with PRES as presenting manifestation. He started on enalapril and levetiracetam. He improved remarkably well with increased urine output over the next 7 days. The proteinuria subsided and repeat serum complement levels were in normal range by 2 weeks. A repeat MRI brain did not reveal any abnormality suggestive of transient abnormalities during hypertensive encephalopathy.

Discussion

PRES has been mainly attributed to vasogenic oedema, predominantly involving the white matter in the parieto-occipital areas and has been classically described in ecclampsia and immunosuppressive therapy[2].

The pathophysiology of PRES is complex and highly debated. It is often attributed to autoregulatory failure of cerebral blood vessels leading to vasogenic oedema as seen in severe hypertension. The increased predilection of vasogenic oedema for the posterior cerebrum may be explained by the decreased density of sympathetic neurons in the posterior cerebral artery territory[3]. At the onset of symptoms, the blood pressure may be normal or minimally elevated. The clinical manifestations of PRES are usually headache, vomiting, seizures, visual disturbances with altered sensorium among which seizures are the most consistent manifestation. PRES is most commonly seen in pregnancy induced hypertension, however it may be seen in hypertensive encephalopathies as in our case but PRES as a first presentation of PSGN is relatively rare.

PSGN usually occurs after infection of throat or skin by nephritogenic strains of group-A beta hemolytic streptococci. PSGN is an immune-complex-mediated disease with decline in the serum complement (C3) levels. Hypertension is found in majority of patients with PSGN and may also be associated with hypertensive encephalopathy in some cases. Toxic effects of streptococcus on the central nervous system may also lead to encephalopathy. Zaki *et al.* reported PRES as an unusual manifestation of PSGN in a 8 year old child[4]. Wirrell et al reported a series of 4 similar cases in children, however such presentation in a young adult is relatively rare[5].

However, in this case, the patient presented with convulsions and hypertension without any features of oedema, oliguria or frank hematuria. Furthermore, the constellation of symptoms were not suggestive of glomerulonephritis and the lesions on the skin with elevated Anti DNAse B levels, microscopic haematuria anddecreased levels of serum complement clinched the diagnosis. Renal biopsy was not attempted in our case as the symptoms resolved gradually and complement levels normalized in due course of time.

T2 weighted MRI images showing diffuse, symmetrical reversible hyperintensities involving the white matter with relative sparing of the grey matter is suggestive of PRES.Cerebral white matter is more susceptible to vasogenic oedema. PRES with hypertensive encephalopathy should be managed aggressively in order to avoid catastrophic complications and permanent residual sequalae. Anti hypertensive therapy with anticonvulsants for symptomatic management is extremely beneficial.

References

1 Fux CA, Bianchetti MG, Jakob SM, Remonda L — Reversible encephalopathy complicating post-streptococcal glomerulonephritis. *Pediatr Infect Dis J* 2006;**25:**85-7. [PubMed] [Google Scholar]

2 Hinchey J, Chaves C, Appignani B, Breen J — A posterior reversible leucoencephalopathy syndrome. *N Engl J Med* 1996; **334:** 494-500. [PubMed] [Google Scholar]

3 Bartynski WS — Posterior Reversible Encephalopathy Syndrome, Part 2: controversies surrounding pathophysiology of vasogenicedema. *Am J Neuroradiol* 2008; **29:** 1043-49. [PubMed] [Google Scholar]

4 Zaki, Syed Ahmed, and Preeti Shanbag — "Unusual presentation of poststreptococcal glomerulonephritis as posterior reversible encephalopathy syndrome." Journal of pediatric neurosciences vol. 9,1 (2014): 42-4. doi:10.4103/1817-1745.131484

5 Wirrell E, Hamiwka L, Hamiwka L, Grisaru S, Wei X — (2007). Acute Glomerulonephritis Presenting With PRES: A Report of 4 Cases. Canadian Journal of Neurological Sciences / Journal Canadien Des Sciences Neurologiques, **34(3):** 316-21. doi:10.1017/S0317167100006740