

Case report:

Posterior reversible encephalopathy syndrome following post-streptococcal glomerulonephritis

Alves GRT¹, Cordenonsi ICO², Silva RVA³, Haygert CJP⁴

Abstract: Posterior reversible encephalopathy syndrome (PRES) comprises a unique pattern of brain vasogenic edema that is seen in the setting of a neurotoxic status. Besides many etiologies have been already associated with PRES development, such as chronic renal disease, use of chemotherapy agents and inflammatory conditions, the imaging features are very suggestive and helpful for an appropriate diagnosis. We report here a case of PRES secondary to post-streptococcal glomerulonephritis (PSGN), which evolved successfully after clinical management. An 11-year-old boy was admitted with a typical history and findings of PSGN, associated with sensory alterations, headache and recent tonic-clonic seizure. Computed tomography (CT) scan of the head has revealed bilateral and symmetric hypodense areas, remarkably located at posterior cerebral regions, indicating PRES. Patient received support therapy with diuretics, and antibiotics prescription after discharge. At ambulatory follow-ups, the patient remains asymptomatic, with complete clinical and radiological improvement.

Key words: posterior reversible encephalopathy syndrome, computed tomography, glomerulonephritis, brain swelling

Introduction:

Posterior reversible encephalopathy syndrome (PRES) is defined as the occurrence of diffuse brain swelling, most often at posterior cerebral areas, in response to a neurotoxic status that may be secondary to various disorders¹. As the term suggests, the alterations of PRES are potentially reversible after the background cause management. However, if recognition of PRES is delayed, or base condition is mistreated, neurological sequelae or even death may occur. If the etiologies are many, imaging findings are typical and consist in a valuable tool for correct diagnosis². The authors report hereby a successfully conducted case of post-streptococcal glomerulonephritis (PSGN), which presented PRES as a life-threatening complication.

Case presentation:

An 11-year-old boy has been admitted at the pediatric care unit presenting progressive sensory alterations for a week, culminating with a tonic-clonic seizure of recent onset. His parents reported that approximately three weeks ago he was treated for an upper airway infection with antibiotics, and remained asymptomatic until last week. Complaints of mild abdominal pain, nausea, blurred vision and moderate occipital headache were also reported. At physical examination, an arterial pressure of 210 x 120 mmHg was noted. Additionally, significant facial and lower limbs edema (+++/4+) was present. The urinary debit was found to be reduced, and urine demonstrated a dark red aspect. No signs of fever and focal neurological changes were detected.

Laboratorial tests performances have consistently

1. Giordano Rafael Tronco Alves. Clinical Medicine Department from the Federal University of Santa Maria, Santa Maria, Brazil
2. Isadora Cristina Olesiak Cordenonsi. Clinical Medicine Department from the Federal University of Santa Maria, Santa Maria, Brazil.
3. Régis Vinícius de Andrade Silva. Radiology Division from University Hospital of Santa Maria, Santa Maria, Brazil.
4. Carlos Jesus Pereira Haygert. Radiology Division from University Hospital of Santa Maria, Santa Maria, Brazil.

Corresponds to: Giordano Rafael Tronco Alves, M.D. Clinical Medicine Department, Federal University of Santa Maria. Roraima Avenue, 1000. 97105-900. Santa Maria, Rio Grande do Sul State, Brazil. **E-mail:** grtalves@gmail.com.

confirmed the suspicion of a nephrotic syndrome. No anemia, suggesting chronic renal disease, was present. White cells count showed a mild left-shifted neutrophilic leukocytosis, and serum C3 and CH50 complement rates were decreased. The investigation of antistreptolysin O titer was also positive, while anti-DNAse B antibodies were absent. Finally, urinary analyses indicated the presence of proteinuria, hematuria, associated with erythrocyte dysmorphism. Imaging examination was subsequently sought to assess neurological symptoms.

A head computed tomography (CT) scan (Fig. 1) revealed extensive posterior hypodensities, and loss of physiologic cerebral sulci, suggesting the occurrence of vasogenic edema in the parietal and occipital lobes. Considering the clinical scenario, such abnormality was consistent with the diagnosis of PRES in a typical case of PSGN. Supportive therapy with furosemide was instituted, and amoxicillin treatment was prescribed at patient discharge. The boy evolved well neurologically, oliguria and edema resolved in two and nine days, respectively. Periodical ambulatory follow-ups have shown uneventful since then.

predisposing factors are known, including hypertensive status, immunosuppressant drug use, infections, pre-eclampsia, and patients with inflammatory or autoimmune conditions¹.

In children, PRES is even more rare. It is usually associated with renal and adrenal diseases, and also with chemotherapeutic agents intake. The most common symptoms include seizures, headache and mental status alteration, often followed by nausea, vomiting and vision blurring²⁻³. In this context, the early recognition of characteristic radiological features is key to diagnosis performance, as clinical findings may be non-specific or mimic other neurological illnesses.

As we report here, acute glomerulonephritis (usually post-streptococcal) can course with nephrotic syndrome followed by PRES. While the clinical findings of PSGN may be markedly variable, the presented case was not challenging for PSGN diagnosis, once symptoms, history and complementary examinations were typical. Therefore, the early identification and therapy of PRES was possible. For PSGN, the absences of a recent upper airway (pharyngitis) or pyodermitis history can difficult diagnosis establishment⁴. In these situations, the

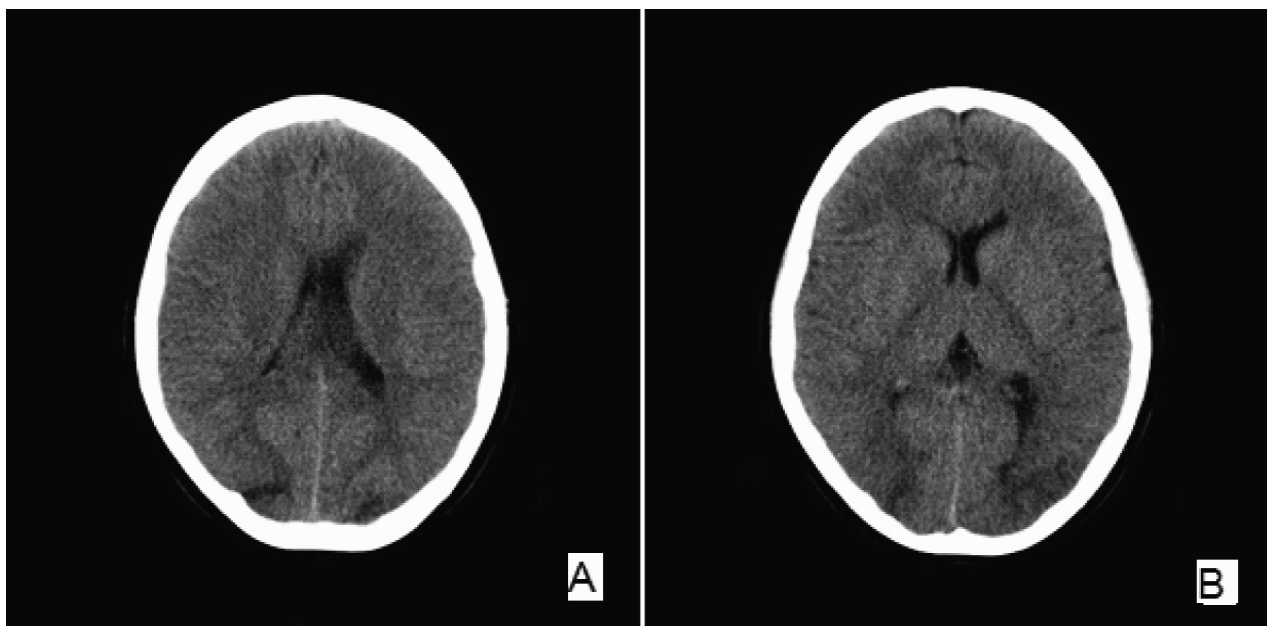


Fig. 1 (A and B) Axial sections of non-contrasted computed tomography (CT) study revealed bilateral parieto-occipital hypodensities of the white matter, representing the occurrence of vasogenic edema. Radiological, laboratorial and clinical findings have corroborated the diagnosis of posterior reversible encephalopathy syndrome (PRES) during post-streptococcal glomerulonephritis (PSGN).

Discussion:

PRES is an infrequent entity, which is characterized by a diffuse cerebral edema secondary to neurotoxicity. However rare, classic etiologies and

combined results of laboratory and imaging examinations may consist in an excellent tool. Additionally, no matter the background condition,

the risk of delayed diagnosis is distinctively high when the child presents previous neurological abnormalities, and the arterial pressure elevation – which is present in around 80% of PRES cases – is overlooked in the setting of agitation⁵.

The precise mechanism behind the vasogenic edema development, and CT or magnetic resonance imaging (MRI) imaging appearance of PRES is still unknown⁶⁻⁸. Two hypotheses are commonly cited, but the issue is very controversial. Most authors accept that severe hypertension exceeds the limits of cerebral autoregulation, leading to breakthrough edema (hyperperfusion hypothesis). However, a more recent theory suggests that the abrupt pressure variance leads to a reflex cerebral vasoconstriction, ischemia, and therefore a late subsequent brain edema would occur. The occurrence of PRES in normotensive patients and an apparent “protection” of brain edema developing in severely hypertensive subjects description support the last one. We may note, however, that they are not truly opposing theories, as the doubt only remains in the first event of PRES setting.

The term PRES was originated from imaging modalities perceives. On CT or MRI studies, the

edema is often widespread, but predominates in the parietal and occipital regions, what suggest the “posterior” description. While absent in the current report, the involvement of the frontal and temporal areas, and also cerebellum is quite common in PRES, along with the occasional presence of lesions in brain stem, basal ganglia, deep white matter, and splenium. As this case demonstrates, abnormalities are habitually bilateral and symmetric, and the edema usually resolves completely after proper clinical management. On the other hand, when atypical presentations occur, MRI diffusion-weighted (DW) sequence study may be recommended to corroborate the vasogenic mechanism of brain swelling².

PRES has become synonymous with a unique pattern of brain vasogenic edema seen in the setting of neurotoxicity, and should be recognized as an infrequent complication of PSGN cases. Its early identification is essential to sequelae avoidance. When a prior history of streptococcal infection is not clear enough, or symptoms and laboratorial findings are questionable, the described imaging features may potentially help in early diagnosis performance.

References:

1. Fugate JE, Claassen DO, Cloft HJ, Kallmes DF, Kozak OS, Rabinstein AA. Posterior Reversible Encephalopathy Syndrome: Associated Clinical and Radiologic Findings. *Mayo Clin Proc* 2010; **85**(5): 427–32. <http://dx.doi.org/10.4065/mcp.2009.0590> PMID:20435835 PMCID:2861971
2. Bartynski WS, Boardman JF. Distinct Imaging Patterns and Lesion Distribution in Posterior Reversible Encephalopathy Syndrome. *AJNR Am J Neuroradiol* 2007; **28**(7): 1320-7. <http://dx.doi.org/10.3174/ajnr.A0549> PMID:17698535
3. Gamie Z, Rizwan A, Balen FG, Clarke M, Hassoon MM. Posterior reversible encephalopathy syndrome in a child with cyclical vomiting and hypertension: a case report. *Journal of Medical Case Reports* 2011; **5**: 137. <http://dx.doi.org/10.1186/1752-1947-5-137> PMID:21470405 PMCID:3080322
4. Ishikura K, Ikeda M, Hamasaki Y, Hataya H, Nishimura G, Hiramoto R, et al. Nephrotic state as a risk factor for developing posterior reversible encephalopathy syndrome in paediatric patients with nephrotic syndrome. *Nephrol Dial Transplant* 2008; **23**(8): 2531-6. <http://dx.doi.org/10.1093/ndt/gfn013> PMID:18258739
5. Froehlich T, Sandifer S, Varma PK, Testa FM. Two cases of hypertension-induced reversible posterior leukoencephalopathy syndrome secondary to glomerulonephritis. *Curr Opin Pediatr* 1999; **11**(6): 512-8. <http://dx.doi.org/10.1097/00008480-199912000-00007> PMID:10590909
6. A Hannan, G Shahjahan, MN Islam. Clinical Profile and Outcome of Acute Post-Streptococcal Glomerulonephritis in Children. *Bangladesh Journal of Medical Science* 1994; **1**(1): 22-27. DOI: <http://dx.doi.org/10.3329/bjms.v1i1.7848>
7. MA Khoybar, S Sultana, AK Shaha, MH Anwar, MI Ashraful, AS Ruhul. Immediate outcome of acute glomerulonephritis in children - experience in a tertiary level hospital. *BJMS* 2011; **10** (4): 269-274. DOI: <http://dx.doi.org/10.3329/bjms.v10i4.9499>
8. Bartynski WS. Posterior reversible encephalopathy syndrome, part 2: controversies surrounding pathophysiology of vasogenic edema. *AJNR Am J Neuroradiol* 2008; **29**(6): 1043-9. <http://dx.doi.org/10.3174/ajnr.A0929> PMID:18403560