

## Posterior reversible encephalopathy syndrome (PRES) associated with acute pancreatitis.

Enrico Pinotti, MD\*; Rubjona Khani, MD; Simone Famuaro, MD; Marta Sandini, MD; Mauro Montuori, MD; Lorenzo Fumagalli, MD; Carlo Ferrarese, MD; Fabio Uggeri, MD, Luca Nespoli, MD; Luca Gianotti MD, PhD

\***Enrico Pinotti, MD**

Department of Surgery, Ospedale San Gerardo, via Pergolesi 33, 20900 Monza (MB), Italy.  
Phone: +39-039-233-3651; Email: enricopinotti@hotmail.it

### Abstract

Posterior Reversible Encephalopathy Syndrome (PRES) is a clinic-neuroradiologic disease characterized by variable associations of neurological symptoms. The brain imaging abnormalities are often symmetric and predominate in the posterior white matter. PRES can develop in association with a vast array of conditions including hypertensive encephalopathy, eclampsia, and the use of cytotoxic and immunosuppressant drugs. Only few cases in the literature show an association between PRES and acute pancreatitis. We report a case of a 61 yo woman presenting PRES after Endoscopic Retrograde CholangioPancreatography related pancreatitis.

### Keywords

posterior reversible encephalopathy syndrome (PRES); pancreatitis; ERCP; endoscopic retrograde CholangioPancreatography

### Introduction

Posterior Reversible Encephalopathy Syndrome (PRES) is a clinic-neuroradiologic disease that was first described by Hinchey et al. in 1996 [1]. It is increasingly recognized and reported in case reports but the real incidence is not known [2]. Patients of all ages appear susceptible and case series suggest that it's more common in women [3,4]. PRES is characterized by variable associations of seizure activity, consciousness impairment, headaches, visual abnormalities, and focal neurological signs [5]. Typical findings in magnetic resonance imaging (MRI) are symmetrical white matter edema in the posterior hemispheres, particularly the parieto-occipital regions [6-9]. Both clinical and radiological features are typically reversible once the cause is removed. PRES occurs most often in the setting of hypertensive crisis, eclampsia, cytotoxic or immunosuppressant therapy and renal disease [10-13]. Regardless the underlying cause, the main abnormality is cerebral vasogenic edema [14]. Only few cases in the literature show an association between PRES and acute pancreatitis [15-23].

### Case Report

A 61 years old caucasian woman, come to our attention from the emergency ward, complaining of abdominal pain lasting for 5 days. She referred to has been submitted in the past to cholecystectomy and

appendectomy, she denied history of hypertension. The remaining medical history was unremarkable. At blood samples, it was discovered an increase of total bilirubin (2.1mg/dl), Gamma-Glutamyl-Transpeptidase (1105U/l) and Alkaline-Phosphatase (615U/l); inflammatory indexes, amylase and lipase serum levels were in the normal range. An abdomen-US was performed: a mild dilatation of main bile duct without evidence of stones was found; a dilatation of intrahepatic biliary ducts was also present. Thus, the patient was admitted to the HPB unit of our hospital where she underwent ERCP (Endoscopic Retrograde CholangioPancreatography). This examination revealed an inflamed papilla, conditioning a sub-stenosis of 1cm. Oddi's sphincterotomy and brushing of the inflammatory lesion was performed (the cytology was negative for malignant tumor cell). A 10Fr biliary plastic stent was placed. After 3 hours from the procedure, the patient developed a post-ERCP pancreatitis, with an elevation of serum amylase (3262U/l) and lipase (3963U/l); WBC was 12, 6mg/dl and C reactive protein was 5 mg/l. She also complained abdominal pain rebound, responsive to pain relief with NSAID. The morning after the procedure the patient was in good general condition with less abdominal pain. In the afternoon the patient experienced the sudden appearance of complete bilateral blindness. Vital parameters were normal except of mild hypertension (160/80). At the Neurological examination the pupillary reflex was intact and no other focal signs were revealed. A vascular and encephalic CT-scan was performed and show no evidences of organic diseases. After oculist examination, it was excluded either a ophthalmological disease. Also EEG was normal. Then the patient was examined with encephalic contrast enhanced MRI that showed in both parietal and occipital lobes the presence of symmetrical areas of increased signal in the T2 FLAIR sequence (Figure 1). The radiological picture was compatible with PRES. An abdomen TC-scan was performed after 48 hours and confirmed a mild inflammation of pancreatic head. The patient was treated conservatively with oral anti-hypertensive therapy and parenteral hydration. After 36 hours from the outset, her vision slightly return normal, but for 3 days she experienced visual hallucinations. Bilateral normal visus was reached after 6 days. At the same time, serum amylase and lipase levels decrease to normal range and the patient was discharged. A MRI was performed two weeks later and documented the complete recovery of the brain signal intensity in the areas previously affected (Figure. 2).

## Discussion

Posterior reversible Encephalopathy occurs in association with various medical condition that include hypertension, sepsis, shock, eclampsia, cytotoxic/immunosuppressive therapy, uremia, porphyria, connective tissue disease, alcohol, intoxication, hematological, renal or autoimmune disorders [10-13]. PRES evolves over a matter of hours, with the most common presenting symptoms being seizures, consciousness impairment, visual abnormalities and headache.

The severity of clinical symptoms is variable. Seizures and status epilepticus are common [24]. Consciousness impairment may range in severity from confusion, somnolence, and lethargy to encephalopathy or coma [25]. The visual abnormalities can manifest as blurred vision, homonymous hemianopsia, cortical blindness, and visual hallucinations [25]. Other less commonly symptoms include nausea, vomiting, and brainstem deficits [1]. Hypertension is often observed in PRES, but its level is not correlated to the severity of PRES.

Neuroimaging is essential to the diagnosis of posterior reversible encephalopathy syndrome. Typical findings are bilateral and symmetric regions of white matter edema and predominating in the parietal and occipital lobes [26].

The pathophysiology of PRES remains unclear, but it appears to be related to disorder of cerebral autoregulation and endothelial dysfunction that lead to breakdown of blood brain barrier which may then trigger vasogenic cerebral edema [14].

In our case, patient developed cortical blindness, with typical neurological findings of PRES in course of acute pancreatitis. She also developed a moderate hypertension. The symptoms and MRI alteration regressed with the normalization of amylase and lipase levels.

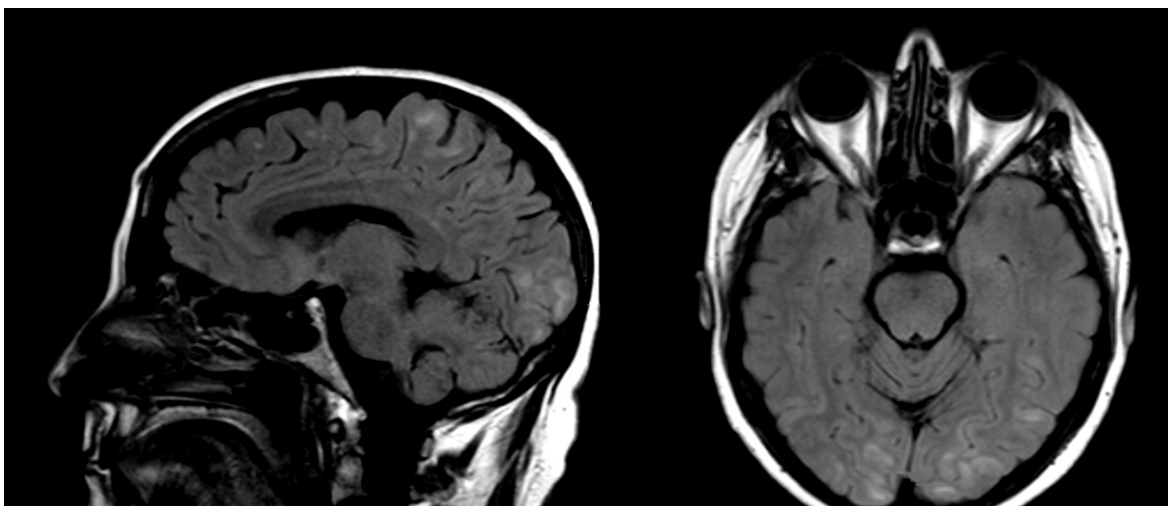
Only a few cases of PRES in the setting of acute pancreatitis, with or without other possible triggers for PRES, have been reported [15-23] (Table 1). The few reported cases of pancreatitis associated with PRES concern both genders and variable age (18-61 years). Various causes of pancreatitis are reported in association with PRES (alcohol, traumatic pancreatitis, nephrotic syndrome, porphyria, steroids and antiphospholipid syndrome).

In acute pancreatitis, the serum levels of pro-inflammatory cytokines, lipase, and amylase are elevated which could play a major role in the development of extra-pancreatic complications [27-28]. Experimental models in rats suggested that during acute pancreatitis the pro-inflammatory cytokines, such as IL-6 and TNF, contribute to vasogenic brain edema through an alteration of the blood brain barrier permeability.

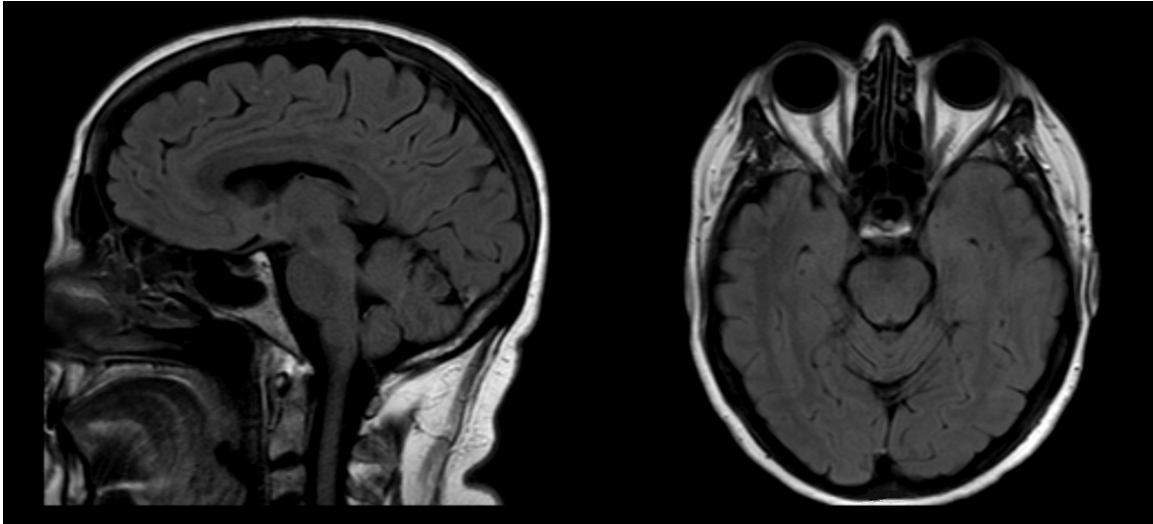
## Conclusion

In conclusion PRES is a rare disease with multiple possible risk factors. Pancreatitis is associated with a systemic inflammatory state that could influence the permeability of hematoencephalic barrier and promote the development of PRES. In literature there are few reports about PRES in acute pancreatitis. Clinicians should know this possible and rare complication of acute pancreatitis in order to recognize it early and focus the diagnostic pathway.

## Figures



**Figure 1:** Magnetic resonance imaging scan showing symmetrical areas of increased signal in the occipital, parietal, and frontal lobes (T2 and FLAIR sequences).



**Figure 2:** Following magnetic resonance imaging scan showing the complete recovery of brain signal intensity in the areas before affected (T2 and FLAIR sequences).

## Tables

**Table 1:** Cases of PRES associated with acute pancreatitis, analysis of literature

Author	Sex	Age	Associate disease	Cause of pancreatitis	Symptoms of PRES	Timing of complete resolution
Hagemann [15] (2004)	F	42	Acute renal failure Acute respiratory failure Chronic alcoholism Hypertension	Alcohol	Disorientation Headache Transient hemiparesis Visual disturbance	15 days
Shen [16] (2008)	M	60	Acute intermittent porphyria	Acute intermittent porphyria	Disorientation Generalized seizures Psychosis	30 days
Yamada [17] (2012)	F	13	Acute renal failure Nephrotic syndrome	Nephrotic syndrome	Headache Generalized seizure Visual disturbance	2 months
Ishikawa [18] (2013)	F	28	Chronic alcoholism	Alcohol	Disorientation Generalized seizures Visual hallucinations	15 days
Nishimoto [19] (2014)	M	18	Acute leukemia Acute renal failure ARDS Sepsis	Steroids	Generalized seizure	20 days
Baek [20] (2014)	M	49	Chronic alcoholism	Alcohol	Disorientation	14 days
Lamour [21] (2014)	F	19	Acute renal failure Antiphospholipid syndrome	Antiphospholipid syndrome	Headache Generalized seizure	Timing not specified
Murphy [22] (2015)	F	40	-	Alcohol	Acute bilateral blindness	2 months
Sigurtà [23] (2016)	M	15	Sepsis	Trauma	Stupor and neck rigidity	10 days
Pinotti (2017)	F	61	-	ERCP	Acute bilateral blindness Visual hallucinations	6 days

## References

1. Hinchey J, Chaves C, Appignani B, Breen J, Pao L, Wang A et al. A reversible posterior leukoencephalopathy syndrome. *N Engl J Med.* 1996; 22;334: 494-500.
2. Granata G, Greco A, Iannella G, Granata M, Manno A, Savastano E et al. Posterior reversible encephalopathy syndrome--Insight into pathogenesis, clinical variants and treatment approaches. *Autoimmun Rev.* 2015; 14: 830-6.
3. Lee VH, Wijdicks EF, Manno EM, Rabinstein AA. Clinical spectrum of reversible posterior leukoencephalopathy syndrome. *Arch Neurol.* 2008; 65: 205-10.
4. S1: Siebert E, Bohner G, Endres M, Liman TG. Clinical and radiological spectrum of posterior reversible encephalopathy syndrome: does age make a difference?--A retrospective comparison between adult and pediatric patients. *PLoS One.* 2014; 16;9: e115073.
5. Legriél S, Pico F, Azoulay E. Understanding posterior reversible encephalopathy syndrome. Volume 1 of the series Annual Update in Intensive Care Unit and Emergency medicine 2011; 631-653
6. Lamy C, Oppenheim C, Méder JF, Mas JL Neuroimaging in posterior reversible encephalopathy syndrome. *J Neuroimaging.* 2004; 14: 89
7. McKinney AM, Jagadeesan BD, Truwit CL. Central-variant posterior reversible encephalopathy syndrome: brainstem or basal ganglia involvement lacking cortical or subcortical cerebral edema. *AJR Am J Roentgenol.* 2013; 201: 631-8
8. Agarwal A, Kapur G, Altinok D. Childhood posterior reversible encephalopathy syndrome: Magnetic resonance imaging findings with emphasis on increased leptomeningeal FLAIR signal. *Neuroradiol J.* 2015; 28: 638-43.
9. Fugate JE, Rabinstein AA. Posterior reversible encephalopathy syndrome: clinical and radiological manifestations, pathophysiology, and outstanding questions. *Lancet Neurol.* 2015; 14: 914-25.
10. Muhammad S, Güresir Á, Greschus S, Scorzin J, Vatter H, Güresir E. Posterior Reversible Encephalopathy Syndrome as an Overlooked Complication of Induced Hypertension for Cerebral Vasospasm: Systematic Review and Illustrative Case. *Stroke.* 2016; 47(2): 519-22.
11. Zhang L, Wang Y, Shi L, Cao J, Li Z, Wáng YX Late postpartum eclampsia complicated with posterior reversible encephalopathy syndrome: a case report and a literature review. *Quant Imaging Med Surg.* 2015; 5: 909-16.
12. Graham BR, Pylypchuk GB Posterior reversible encephalopathy syndrome in an adult patient undergoing peritoneal dialysis: a case report and literature review. *BMC Nephrol.* 2014; 13;15:10
13. Chen S, Hu J, Xu L, Brandon D, Yu J, Zhang J Posterior Reversible Encephalopathy Syndrome After Transplantation: a Review. *Mol Neurobiol.* 2016; 53: 6897-6909.
14. Bartynski WS. Posterior reversible encephalopathy syndrome, part 2: controversies surrounding pathophysiology of vasogenic edema. *AJNR Am J Neuroradiol.* 2008; 29: 1043-9.
15. Hagemann G, Ugur T, Witte OW, Fitzek C. Recurrent posterior reversible encephalopathy syndrome (PRES). *J Hum Hypertens.* 2004; 18: 287-9.
16. Shen FC, Hsieh CH, Huang CR, Lui CC, Tai WC, Chuang YC. Acute intermittent porphyria presenting as acute pancreatitis and posterior reversible encephalopathy syndrome. *Acta Neurol Taiwan.* 2008; 17: 177-83.
17. Yamada A, Atsumi M, Tashiro A, Hiraiwa T, Ueda N Recurrent posterior reversible encephalopathy syndrome in nephrotic syndrome: case report and review of the literature. *Clin Nephrol.* 2012; 78: 406-11.

18. Ishikawa H, Natsume N, Matsui K, Tsuda H. Acute alcohol withdrawal accompanied by posterior reversible encephalopathy syndrome. *Psychiatry Clin Neurosci*. 2013; 67: 189.
19. Nishimoto M, Koh H, Bingo M, Yoshida M, Nanno S, Hayashi Y et al. Posterior reversible encephalopathy syndrome following acute pancreatitis during chemotherapy for acute monocytic leukemia. *Rinsho Ketsueki*. 2014; 55: 552-7.
20. Baek HS, Lee SJ. A case of posterior reversible encephalopathy syndrome associated with acute pancreatitis and chronic alcoholism. *Gen Hosp Psychiatry*. 2015; 37: 192.e3-5.
21. Larmour K, Lewis G, Benson G, Hanko J. The challenges of antiphospholipid syndrome: experience from diagnosis to self-care. *BMJ Case Rep*. 2014; 26;2014.
22. Murphy T, Al-Sharief K, Sethi V, Ranger GS Posterior Reversible Encephalopathy Syndrome (PRES) After Acute Pancreatitis. *West J Emerg Med*. 2015; 16: 1173-4.
23. Sigurtà A, Terzi V, Regna-Gladin C, Fumagalli R. Posterior Reversible Encephalopathy Syndrome Complicating Traumatic Pancreatitis: A Pediatric Case Report. *Medicine (Baltimore)*. 2016; 95: e3758.
24. Kozak OS, Wijdicks EF, Manno EM, Miley JT, Rabinstein AA. Status epilepticus as initial manifestation of posterior reversible encephalopathy syndrome. *Neurology* 2007; 69: 894-7
25. Roth C, Ferbert A. The posterior reversible encephalopathy syndrome: what's certain, what's new? *Pract Neurol* 2011; 11: 136-44
26. Casey SO, Sampaio RC, Michel E, Truwit CL. Posterior reversible encephalopathy syndrome: utility of fluid-attenuated inversion recovery MR imaging in the detection of cortical and subcortical lesions. *AJNR Am J Neuroradiol* 2000; 21: 1199-1206
27. Saez Royula-Gonzalo, F, Gomez-Rodriguez, R., GomezRubio, M., Heredia-Centeno, M., Miranda-Baiocchi, R. and Hernandez-Guio, C., Complications of acute pancreatitis. Frequency, moment of onset and multiplicity, *Rev. Clin. Esp.* 184 280-284.
28. Saija, A., Princi, P., Lanza, M., Scalese, M., Aramnejad, E. and De Sarro, A., Systemic cytokine administration can affect blood-brain barrier permeability in the rat, *Life Sci*. 1995; 56: 775-784.

**Manuscript Information:** Received: March 03, 2018; Accepted: April 26, 2018; Published: April 30, 2018

**Authors Information:** Enrico Pinotti, MD<sup>1\*</sup>; Rubjona Xhani, MD<sup>2</sup>; Simone Famuaro, MD<sup>1</sup>; Marta Sandini, MD<sup>1</sup>; Mauro Montuori, MD<sup>1</sup>; Lorenzo Fumagalli, MD<sup>2</sup>; Carlo Ferrarese, MD<sup>2</sup>; Fabio Uggeri<sup>1</sup>, MD; Luca Nespoli, MD<sup>1</sup>; Luca Gianotti MD, PhD<sup>1</sup>

<sup>1</sup>Department of Surgery, Ospedale San Gerardo, via Pergolesi 33, 20900 Monza (MB), Italy

<sup>2</sup>Department of Neurology, Ospedale San Gerardo, via Pergolesi 33, 20900 Monza (MB), Italy

**Citation:** Pinotti E, Xhani R, Famuaro S, Sandini M, Montuori M, Fumagalli L, et al. Posterior reversible encephalopathy syndrome (PRES) associated with acute pancreatitis. *Open J Clin Med Case Rep*. 2018; 1406.

**Copy right statement:** Content published in the journal follows Creative Commons Attribution License (<http://creativecommons.org/licenses/by/4.0>). © Pinotti E 2018

Journal: *Open Journal of Clinical and Medical Case Reports* is an international, open access, peer reviewed Journal focusing exclusively on case reports covering all areas of clinical & medical sciences.

Visit the journal website at [www.jclinmedcasereports.com](http://www.jclinmedcasereports.com)

For reprints and other information, contact editorial office at [info@jclinmedcasereports.com](mailto:info@jclinmedcasereports.com)