ORIGINAL RESEARCH PAPER

Volume-8 | Issue-8 | August - 2019 | PRINT ISSN No. 2277 - 8179

INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

PANCREATIC ENCEPHALOPATHY - A CASE REPORT

Gastroenterology

Dr Rajat Jhamb Professor, Department of Medicine, Guru Teg Bahadur Hospital, Delhi, India

Dr Anant Parasher* Senior Resident, Department of Medicine, Guru Teg Bahadur Hospital, Delhi, India. *Corresponding Author

Dr Ashish Baweja Post-Graduate Resident, Department of Medicine, Guru Teg Bahadur Hospital, Delhi, India.

Dr Abhishek Vaingankar Post-Graduate Resident, Department of Medicine, Guru Teg Bahadur Hospital, Delhi, India.

ABSTRACT

Pancreatic encephalopathy is a rare complication of acute pancreatitis, which denotes the occurrence of neuropsychiatric abnormalities in the setting of acute pancreatic inflammation, and is frequently under-diagnosed. It presents with neurological symptoms that may persist even after the resolution of all metabolic parameters. Here we present a 28 year old male patient presenting with altered sensorium and focal neurological deficit during the course of acute pancreatitis. The patient was treated conservatively, and later improved with supportive care.

KEYWORDS

Acute Pancreatitis, Encephalopathy, Focal Neurological deficit

BACKGROUND

Pancreatic encephalopathy is a rare complication of acute pancreatitis, which denotes the occurrence of neuropsychiatric abnormalities in the setting of acute pancreatic inflammation, and is frequently under-diagnosed. It presents with neurological symptoms that may persist even after the resolution of all metabolic parameters.[1,2] The syndrome of pancreatic encephalopathy was first described by Rothermich and Von Haam in 1941, and Vogel in 1950 investigated the pathogenesis of this condition. Many cases have been published since then, but the pathophysiology of the disease has not yet been completely elucidated.[3] It is difficult to diagnose an early pancreatic encephalopathy and also to differentiate pancreatic encephalopathy from Wernicke's encephalopathy.[3] Even with the successful treatment of severe acute pancreatitis, 18.2% of patients will still develop encephalopathy, and the mortality rate can reach as high as 67%.[3] The incidence of Pancreatic encephalopathy is much lower in people with simple acute pancreatitis and the diagnosis is non-specific with no explicit standard or reliable diagnostic index for this condition.[2]

CASE REPORT

A 28 year old male presented to the medicine emergency with altered sensorium and a history of vomiting and abdominal pain for six days prior to admission. There was also a history of Ayurvedic drug intake two days prior for the abdominal pain. There was no history of fever, abdominal distension, jaundice, bleeding manifestations, hypertension, diabetes or tuberculosis. The patient's attendants did not give any history of alcohol abuse, smoking or illicit drug use.

On examination, the patient was altered. His blood pressure was 130/80 mm Hg and pulse was 78 beats per minute. There was no pallor, edema, cyanosis, clubbing or lymphadenopathy. Icterus was present in the sclera. CNS examination revealed a Glasgow Coma score of 9/15. Pupils were mid-dilated and sluggishly reacting. Reflexes were normal and the Bilateral Plantar Response was Extensor/ Babinski's Positive. Rigidity was noted in both the upper and lower limbs. Power and Sensory System could not be assessed in view of altered sensorium. Abdominal examination revealed a soft non-tender abdomen and there was no organomegaly. Respiratory and cardiac examinations were unremarkable.

Routine blood counts revealed elevated liver enzymes with SGOT (AST) and SGPT (ALT) being 774 U/L and 3990 U/L respectively, on admission. The total bilirubin was 6.1 mg/dl, with the direct fraction being 1.8mg/dl. Alkaline phosphatase was 185 IU/L. A Provisional diagnosis of Fulminant Hepatitis was made and immediate treatment was started with IV fluids and necessary IV antibiotic cover. Tests for HIV, HBV and HCV done by ELISA were negative and IgM antibodies to HAV and HEV were not found. A urine toxicology screen obtained was negative and an emergency Ultrasound of the whole abdomen did not reveal any significant findings. A Lumbar puncture and CSF analysis was done, which revealed an acellular picture with sugar and protein levels of 85 mg/dl and 34mg/dl respectively. Serum Ceruloplasmin and 24 hour Urinary copper levels were normal. On the third day of admission, the patient's serum amylase and lipase levels were 180 U/L and 300 U/L respectively which increased to 196 U/L and 400 U/L the next day. An emergency CECT Abdomen revealed an Acute Early Interstitial Edematous Pancreatitis, involving the pancreatic tail. NCCT Head was not suggestive of any significant abnormality. Treatment was supplemented with Thiamine 300mg stat as well as on maintenance with dextrose, along with Intravenous fluids. The patient's sensorium improved and the patient gradually became fully conscious and oriented on the fifth day of admission.

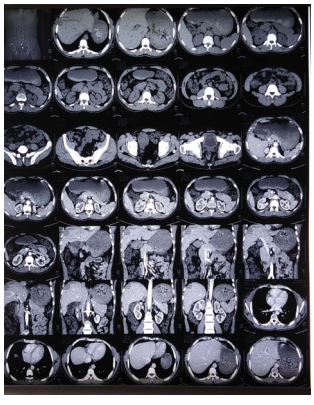

FIG – 1: CECT Abdomen showing Acute Interstitial Edematous Pancreatitis involving the Pancreatic Tail

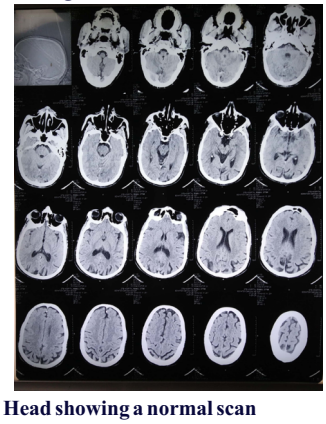

FIG 2: NCCT Head showing a normal scan

DISCUSSION

Pancreatic Encephalopathy is a rare complication of acute pancreatitis. It usually occurs in the early stages of the disease and has a mortality rate of 48.5% to 67%.[4-6] The neurological complications in the restorative or later stages of severe and simple acute pancreatitis are usually secondary to long fasting, multiple vomiting episodes and Total Parenteral Nutrition (TPN) without any thiamine supplementation, collectively termed as Wernicke's encephalopathy.[5,7] Studies have shown that pancreatic encephalopathy is poorly recognized by most physicians[7], and delay in recognition and necessary supportive care can prove to be fatal in these cases.

Pathophysiology

The pathogenesis of pancreatic encephalopathy has been poorly understood. During severe acute pancreatitis, high levels of pancreatic enzymes including trypsin, elastin, lipase and phospholipase A2 (PLA2) enter the circulation and alter the permeability of the Blood Brain Barrier. PLA2, with potent neurotropism, can directly act on the phospholipid layers of brain cells, causing neuronal edema, focal hemorrhagic necrosis, meningeal inflammation with severe demyelinating changes in axons and secondary cell dysmetabolism of neurons, leading to various neuropsychiatric symptoms.[2,8,9] The increased blood-brain barrier permeability can also cause sensitized blood T-cells to enter the brain tissue contributing to demyelination.[8] These patients have also been found to have intracerebral capillary engorgement with cell infiltration and broadened intercellular spaces in the brain parenchyma.[2]

During severe acute pancreatitis, hypoxemia and intravascular fluid depletion can activate the renin angiotensin system, which also plays an important role in the occurrence and progression of pancreatic encephalopathy.[10-12] Additionally, Pro-inflammatory cytokines such as TNF-a, IL-1b, and IL-6 increase the permeability of the blood-brain barrier by damaging it.[13] In addition to these factors, reduced myocardial pumping due to the release of myocardial depressant factor[14,15] and deterioration of circulatory capacity due to vomiting and third spacing of fluid leads to cerebral ischemia and hypoxia.

Histo-pathological findings seen in pancreatic encephalopathy include hemorrhagic foci, demyelination and areas of cerebral malacia.[4] Brain imaging obtained during the acute phase of pancreatic encephalopathy may essentially be normal.[1]

Clinical Features

The early stage of pancreatic encephalopathy is seen within 15 days (usually 2–5 days) after onset of symptoms of acute pancreatitis.[7] The symptoms include confusion, dysarthria, anxiety, muscular aches, short lucid periods and a cyclic progression with remission and relapses. Examination findings include rigidity of all four limbs, hyper-reflexia and asterixis with an occasionally positive Babinski's sign.[2,3] Though the encephalopathy often resolves without leaving any focal deficits, an important long term manifestation of this condition might be cognitive impairment which is demonstrable on the MRI as diffuse cortical and subcortical atrophy.[16]

A report of Boon et al showed the usefulness of MRI in the diagnosis of this disorder. Patchy white matter signal abnormalities, resembling plaques seen in multiple sclerosis, might reflect the lesions that were found in the cerebral white matter of clinically confirmed post-mortem patients.[17]

A close differential diagnosis of Pancreatic encephalopathy is Wernicke's encephalopathy which may occur late in the course of acute pancreatitis as a result of prolonged fasting and thiamine depletion.[16] Encephalopathy complicating pancreatitis may also occur due to hypoxia secondary to pulmonary fat embolism, cerebral fat embolism, or the complicating syndromes of disseminated intravascular coagulation or hyperosmolality.[18]

Treatment

The main therapy for pancreatic encephalopathy is the support and management of severe acute pancreatitis and its complications. In the early stages of disease, it is necessary to control pancreatic secretion and drainage, correct fluid and electrolyte imbalance as well as hypoproteinemia, and provide sufficient caloric support.[2] There is no specific treatment for this condition except supportive care and thiamine.[1] With proper treatment, recovery is uneventful among patients below 40 years of age but may have sequelae such as cerebral infarction in the elderly, especially those above 60 years of age.

Pancreatic encephalopathy carries a high mortality among patients and if recognition and treatment are delayed, there might be potentially serious neurological outcomes.[19] The most common causes of death secondary to encephalopathy include shock, ketoacidosis and multi-organ dysfunction syndrome.

CONCLUSION

Pancreatic encephalopathy is a rarely reported but devastating complication in patients having Acute Pancreatitis. The patients may present with altered behaviour and neurological deficits either early or late in the progression of the disease, and even during resolution. Therefore it becomes necessary to anticipate this phenomenon in all complicated cases of pancreatitis, and provide early and adequate conservative management with fluid support and judicious antibiotic cover, if and as required. The treatment of acute pancreatitis is primarily non-operative except in cases of severe acute biliary pancreatitis with resultant complications such as fluid sequestration and phlegmon formation, in which effective and timely drainage leads to a low mortality rate in the patients who receive surgery. There is also sufficient evidence which suggests that pancreatic encephalopathy can be prevented by administration of Vitamin B1/Thiamine as well as good nutritional replenishment in the early stages of the disease.

***Financial support and sponsorship*:** None

***Conflicts of Interest*:** None

REFERENCES

1. Ruggieri RM, Lupo I, Piccoli F. Pancreatic encephalopathy: a 7-year follow-up case report and review of the literature. Neurol Sci. 2002;23(4):203-5.
2. Pancreatic encephalopathy Joyce A. Akwe, Gloria E Westney, Tatah S Fongeh. Am J Case Rep 2008; 9:399-403.
3. Sharf B, Bental E. Pancreatic encephalopathy. J Neurol Neurosurg Psychiatry. 1971;34(3):357-61.
4. Wu XN. Current concept of pathogenesis of severe acute pancreatitis. World J Gastroenterol. 2000;6(1):32-36.
5. Ding X, Liu CA, Gong JP et al: Pancreatic encephalopathy in 24 patients with severe acute pancreatitis. Hepatobiliary Pancreat Dis Int. 2004; 3(4): 608–11
6. Chen L, Zhang X: Pancreatic encephalopathy and Wernicke encephalopathy.
7. Sun GH, Yang YS, Liu QS, Cheng LF, Huang XS. Pancreatic encephalopathy and Wernicke encephalopathy in association with acute pancreatitis: a clinical study. World J Gastroenterol. 2006;12(26):4224-7.
8. Homma M, Suzuki H, Kusuhara H et al: High affinity efflux transport system for glutathione conjugates on the luminal membrane of a mouse brain capillary endothelial cell line (MBEC4). J Pharmacol Exp Ther,1999; 288: 198–203.
9. Pavelko KD, van Engelen BG, Rodriguez M: Acceleration in the rate of CNS remyelination in lysolecithin-induced demyelination. J Neurosci, 1998; 18: 2498–505
10. Leung PS: Local renin-angiotensin system in the pancreas: the significance of changes by chronic hypoxia and acute pancreatitis. JOP, 2001; 2: 3–8.
11. Leung PS, Carlsson PO: Tissue renin-angiotensin system: its expression, localization, regulation and potential role in the pancreas. J Mol Endocrinol, 2001; 26: 155–64.
12. Leung PS, Chan WP, Nobiling R: Regulated expression of pancreatic renin- angiotensin system in experimental pancreatitis. Mol Cell Endocinol, 2000; 166: 121–28 Case Report 402.
13. Farkas C, Matton J, Nagy Z et al: Experimental acute pancreatitis results in increased blood-brain barrier permeability in the rat: a potential role for tumour necrosis factor and interleukin-6. Neurosci Lett, 1998; 242:147–50.
14. Tenenbein MS, Tenenbein M: Acute pancreatitis due to erythromycin overdose. Pediatr Emerg Care, 2005; 21: 675–76.
15. Ates F, Kosar F, Aksoy Y et al: QT interval analysis in patients with acute biliary pancreatitis. Pancreas, 2005; 31: 238–41.
16. http://www.astrocyte.in/temp/Astrocyte24185-3804612_103406.pdf
17. Boon P, de Reuck J, Achten E, de Bleecker J. Pancreatic encephalopathy. A case report and review of the literature. Clin Neurol Neurosurg. 1991;93(2):137-41.
18. Johnson DA, Tong NT. Pancreatic encephalopathy. South Med J. 1977;70(2):165-7.
19. Ramanathan RS, Ahluwalia T. Acute necrotizing pancreatitis leading to pancreatic encephalopathy in a patient undergoing long-term continuous ambulatory peritoneal dialysis. J Acad Med Sci 2012; 2: 85-7.