Case Report :

Central neurogenic hyperventilation secondary to primary CNS mantle cell lymphoma

Rajesh N. Swarnakar, Deepak I. Muthreja, Akshay D Deotare, Bhavesh J. Vaghani
Department and institution:
Getwell Hospital and Research Institute 20/1, Dr Khare Marg, Dhantoli, Nagpur, Maharashtra 440012

ABSTRACT

In this case report we have described a case of Central neurogenic hyperventilation(CNH) in a 68 year old male patient. The etiology been discovered is Mantle cell lymphoma, a very rare variant of primary CNS lymphoma .As the CNH (central neurogenic hyperventilation) and Mantle cell lymphoma are rare entities, the presence of both of them together in a single patient makes this index case unique for reporting.

Key Words : Central neurogenic hyperventilation (CNH), Mantle cell lymphoma, Primary CNS lymphoma (PCNSL)

INTRODUCTION :

Central neurogenic hyperventilation (CNH) is a rare entity of hyperventilation syndromes. Most of CNH cases are caused by infiltrative tumors in pons and medulla. Here we present a case of CNH caused by Mantle cell lymphoma rare variant of primary CNS lymphoma (PCNSL).

CASE DETAILS :

A 68 year old hypertensive man was admitted in ICU with history of change of voice for 15 days, dry cough for 7 days with breathlessness and drowsiness for 2 days. The patient had no fever, chest pain, or wheeze. He was a non-smoker and a non-alcoholic. There was no significant past medical history. On examination, the patient was found drowsy (but could be easily aroused), but restless with a disproportionately high respiratory rate of 50 per minute while the pulse rate was normal (70 per minute) and blood pressure was 140/90 mmHg. His oxygen saturation was 98% on ambient air. On general examination, there was no other obvious abnormality. The auscultation of chest revealed no abnormality. Although drowsy, there were no other significant neurological findings on examination. A systematic examination of the other systems was normal.

On investigations, the arterial blood gases (ABG) showed a pH of 7.64, PaCO2of 8 mm Hg, PaO2, 85 mm Hg and bicarbonate (HCO3) been as low as 6.8 mEq/L. His hemoglobin level was 13.6mg/dl with hematocrit of 40.5% and the serum sodium, potassium and chloride were 142 mEq/L, 4.2 mEq/L and 106 mEq/L respectively. The initial complete blood count, Liver function test, serum creatinine and thyroid profile were normal. He was HIV seronegative. His serum lactate and salicylate levels were within normal limit. Chest x-ray (PA view) showed patchy opacities which on high resolution computer tomography (HRCT) of chest appeared to be early sub-pleural fibrotic changes corresponding to bilateral lower lobes associated with small areas of patchy consolidations. Computerized tomography (CT) of brain and 2D-Echocardiography did not reveal any abnormalities. Fiber optic bronchoscopy was performed at bedside. It showed left vocal cord palsy and thick purulent secretions within the bronchial segments of both the side. The culture

Corresponding author:
Dr Rajesh Swarnakar
Getwell Hospital and research center
20/1, Dr Khare Marg, Dhantoli, Nagpur,
Maharashtra 440012
Mobile : 09545627978, 09271276116
E.mail: drrajeshswarnakar@gmail.com

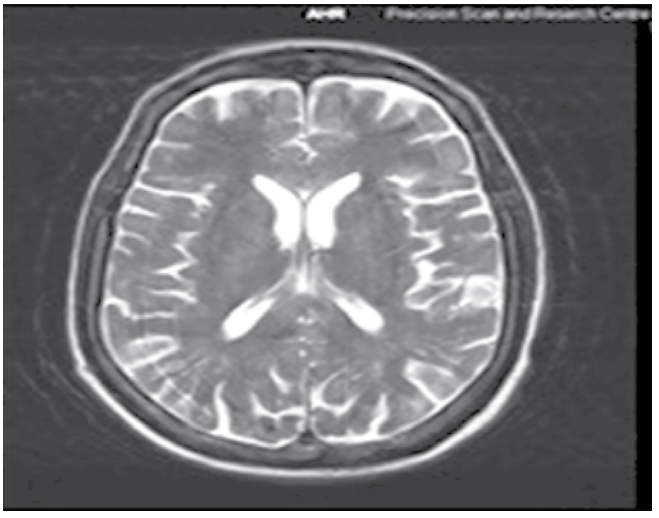
of bronchial wash showed growth of Klebsiella pneumoniae.

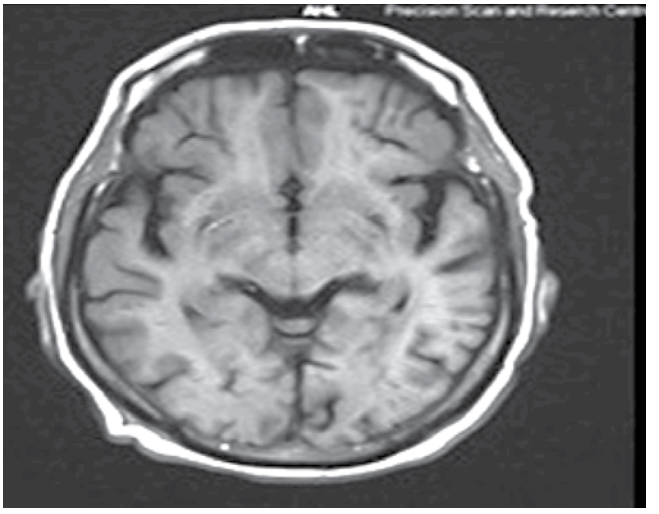
The patient was initially managed with intravenous (IV) antibiotics, which were subsequently changed as per the sensitivity report. He was also treated initially with intravenous sodium bicarbonate and pantoprazole; supportive treatment, whatever required, were continued. Direct laryngoscopy was performed by an otolaryngorhinologist; it showed signs of acute on chronic laryngitis.

The patient remained tachypnoeic even after 7 days of treatment with antibiotics, steroids, and narcotics. His ABG analysis after 7 days showed pH of 7.62, PCO2 as 9 mm Hg, HCO3 as 9.3 mEq/L and PO2 of 110 mm Hg at room air. A nephrologist was consulted in view of metabolic changes on ABG and two settings of hemodialysis were performed following which the ABG showed the signs of correction (pH- 7.58, PCO2- 21 mm Hg, HCO3-19.8 mEq/L,and PO2- 92 mm Hg on room air). A neurologist's opinion was also considered. Magnetic resonance imaging (MRI) brain and cerebrospinal fluid (CSF) study were done. The MRI Brian showed the features representing a metabolic neurodegenerative process/ toxic encephalopathy or encephalitis (Fig 1). CSF contained the following values: glucose level, 31mg/dl, protein level-102 mg/dl with 240 white blood cells of which 50% were lymphocytes and 50% were frank atypical nucleated cells. The CSF was subjected to cytology and it revealed many large atypical cells of lymphoid series; flow cytometry performed revealed their details and pointed to a diagnosis of Mantle cell lymphoma. Hence, a final diagnosis of Central Neurogenic Hyperventilation (CNH) secondary to Primary CNS Mantle Cell Lymphoma was made.

Later patient was intubated and put on mechanical ventilator in view of increase drowsiness and restlessness. Subsequently patient died due to cardio respiratory arrest before we could offer any treatment for mantle cell lymphoma.

Fig 1





DISCUSSION :

Primary central nervous system lymphoma (PCNSL) is a rare variant of extra-nodal non-Hodgkin's lymphoma and accounts 4% of all primary CNS tumors. Primary CNS lymphoma commonly occurs in immune-compromised host. In immunocompetent patients, PCNSL occurs usually during 6th-7th decade of life. Most common sites involved are reported to be the brain, meninges, spinal cord and the eyes; it mostly remains confined to the CNS only. PCNSL localizes to brainstem primarily in 3% cases (1, 2) and about 90% of all PCNSLs are diffuse large B-cell lymphomas while rest 10% are poorly characterized by low grade lymphoma, Burkitt's lymphomas and T-cell lymphoma (3).Our patient had the distinctive feature of central neurogenic hyperventilation associated with Mantle cell

lymphoma; a rare variant of PCNSL. A study by Bateille et al focused on clinical presentations of 248 immunocompetent patients with primary CNS lymphomas and described focal neurological deficits in 70% of patients, neuropsychiatric symptoms in 43%, with 33% having features of raised intracranial pressure, 14% and 4 % showing seizures and ocular symptoms (4).

Plum and Swanson reported the first case of central neurogenic hyperventilation, a rare neurological disorder in 1959. They suggested that "CNH in man results from uninhibited stimulation of both the inspiratory and expiratory centers in the medulla by lateral pontine reticular formation and by laterally located descending neural pathways." (5)There CNH is characterized by hyperventilation that persists during sleep, low arterial PCO2, high arterial PaO2 and high arterial pH in the absence of drugs or metabolic causes.5 Plum reported 18 cases of CNH associated with slowly infiltrating tumors having specified tumor histopathology. There were 9 patients with lymphoma, 6 with slowly growing astrocytoma, with one patient each been contributed by metastatic tumor invading through the skull base, medulloblastoma and aggressive astrocytoma. (6)

CNH is usually seen in comatose patients who have serious brain injuries like head injury, cerebrovascular events, multiple sclerosis and infection. Brain tumor can also produce CNH but it can usually occur in awake patients since the reticular activating system is not affected in the early course of tumor. (7)This condition is extremely uncommon and only 21 cases are reported up till 2005. (8, 9)

REFERENCES :

1. Ferreri AJ, Abrey LE, Blay JY. Summary of statement on primary central nervous system lymphomas from the Eighth International Conference on Malignant Lymphoma.J ClinOncol 2003; 12: 2407-11.
2. Murray K, Kun L, Cox J. Primary malignant lymphoma of the central nervous system: results of treatment of 11 cases and review of the literature. J Neurosurg 1986; 65: 600-7.
3. Batchelor T, Loeffler JS. Primary CNS Lymphoma. J ClinOncol 2006; 24: 1281-8.
4. Bataille B, Delwail V, Menet E. Primary intracerebral malignant lymphoma: A report of 248 cases. J Neurosurg 2000; 92: 261-6.
5. Plum F, Swanson AG. Central neurogenic hyperventilation in man. AMA Arch Neurol Psychiatry1959; 81:535- 549.
6. Plum F. Mechanisms of "central" hyperventilation. Ann Neurol 1982; 11636- 637.
7. Chang CH, Kuo PH, Yang PC. Persistent Severe Hypocapnia and Alkalemia in a 40-Year-Old Woman. CHEST Journal2000; 118 (1): 242–5.
8. Shahar E, Postovsky S, Bennett O. "Central neurogenic hyperventilation in a conscious child associated with glioblastoma multiforme". Pediatric Neurology2004; 30 (4): 287–90.
9. Tarulli AW, Lim C, Bui JD, Saper CB, Alexander MP. "Central Neurogenic Hyperventilation: A Case Report and Discussion of Pathophysiology". Archives of Neurology 2005; 62 (10): 1632–4.