Volume : 3 | Issue : 11 | November 2014 • ISSN No 2277 - 8179

Research Paper

A Rare Case of Primary Angiitis of CNS

Medicine

KEYWORDS :

Dr. Dipen M. Agrawal 2nd Year Resident, Department of medicine, NHLMMC, VS Hospital, Ahmedabad

Dr. Pratik R. Pabani 2nd Year Resident, Department of medicine, NHLMMC, VS Hospital, Ahmedabad

Dr. Mohit P. Kejariwal 2nd Year Resident, Department of medicine, NHLMMC, VS Hospital, Ahmedabad

SUMMARY : A 48 yr old male patient presented with complaints of confusional state, imbalance of walking, memory impairment, tremulousness of limbs, slurring of speech, difficulty in swallowing and slowness of activities since last 2 months ;insidious onset fluctuating course ; preceded by fever for 2 to 3 days, associated with urinary incontinence. On examination patient was conscious disoriented having dysarthric speech with hypertonia ,brisk reflexes, extensor plantar, reduced power in all four limbs .CSF protein was 182.9 mg/dl with normal sugar and cells. MRI Brain shows abnormal asymmetric T2 and FLAIR hyperintensities involving white matter of bilateral frontal lobes ,temporal lobes ,bilateral capsule-ganglionic region ,left mid brain, pons and bilateral cerebellar hemisphere with mild patchy enhancement on post contrast study. CT Brain Angio show mild narrowing of bilateral MCA. Dural & Brain biopsy show small intraparenchymal vessel with transmural chronic inflammation and focal minimal apoptotic debris suggestive of vasculitis. iv predisolone and cyclophosphamide was given and patient is improved with treatment.

BACKGROUND : Primary angiitis of the central nervous system (PACNS) causes focal and diffuse neurologic symptoms due to vasculitis of the intracerebral blood vessels. The broad swathe of symptoms that can mimicPACNS includes angiocentric infections and malignant neoplasms. However, whereas immunosuppressant therapy can be harmful in patients with infections, such cytotoxic based regimens are necessary for treating PACNS, which is otherwise relentlessly progressive and fatal. Therefore, knowledge of the spectrum of demographic, clinical, angiographic, and radiographic features seen in PACNS is necessary for diagnostic accuracy as well as for safely instituting immunosuppressant treatment.

CASE PRESENTATION : 48 yr old male patient presented with complaints of confusional state,imbalance of walking, memory impairment,tremulousness of limbs,slurring of speech, difficulty in swallowing and slowness of activities since last 2 month ,insidious onset fluctuating course, which is preceded by fever for 2 to 3 days,associated with urinary incontinence. On examination patient was conscious disoriented , dysarthric speech with hypertonia ,brisk reflexes, plant extensor, reduced power in all 4 limbs.

INVESTIGATION: HB 11.9, TC 11,500 , PLATELET 3.53 LAKH , S.CREAT 0.94, SGPT 15, NA 134 , K+ 4.3, CSF protein 182.9 mg/dl with normal sugar and cell, CSF negative for VZV,TB PCR, HSV,VDRL,Cryptococcal antigen ,Gram stain ,AFB,India ink, cytology Serum ANA, cANCA, pANCA, Brucella, Ammonia, RPR, HIV, HBsAg, HCV. MRI shows abnormal asymmetric T2 and FLAIR hyperintesity involving white matter of bilateral frontal lobes ,temporal lobes,bilateral capsule-ganglionic region ,left mid brain, pons and bilateral cerebellar hemisphere with mild patchy enhancement on post contrast study. CT Brain Angio show mild narrowing of bilateral MCA. Dural & Brain biopsy show small intraparenchymal vessel with transmural chronic inflammation and focal minimal apoptotic debris suggestive of vasculitis.

TREATMENT AND OUTCOME : iv cyclophosphamide and iv prednisolone was given and patient is improved with treatment.

DISCUSSION : Primary angiitis of the central nervous system (PACNS) is a rare form of vasculitis of unknown cause. The mean age of onset is 50 years, and men are affected twice as often as women. Headache and encephalopathy are the most frequent initial symptoms. Stroke or focal symptoms develop in less than 20% of patients at the onset of disease and are uncommon in the absence of headache or encephalopathy. Symptoms or signs of vasculitis outside of the central nervous system are rare; serologic markers of inflammation are typically normal. Magnetic resonance imaging of the brain is abnormal in more than 90% of patients, but the pattern of abnormal findings is not specific. Cerebrospinal fluid analysis is also usually abnormal because of modest, nonspecific elevations in total protein level or white blood cell count. Angiography has a low sensitivity and low specificity. Most patients suspected of having PACNS have another disease. The diagnosis of PACNS is established by brain biopsy. The differential diagnosis of PACNS is broad and includes reversal cerebral vasoconstriction. In contrast to patients with PACNS, patients with reversal cerebral vasoconstriction are more often young women who experience a thunderclap headache and have a normal cerebrospinal fluid analysis. Patients with biopsy proven PACNS are treated with cyclophosphamide and prednisone.

REFERENCE

1. Salvarani C, Brown RD Jr, Calamia KT, et al. Primary central nervous system vasculitis: | analysis of 101 patients. Ann Neurol. 2007;62(5):442-451. | 2. Calabrese LH, Mallek JA. Primary angiitis of the central nervous system: report | of 8 new cases, review of the literature, and proposal for diagnostic criteria. Medicine | (Baltimore). 1988;67(1):20-39. | 3. Lie JT. Primary (granulomatous) angiitis of the central nervous system: a clinicopathologic | analysis of 15 new cases and a review of the literature. Hum Pathol. | 1992;23(2):164-171. | 4. Moore PM. Diagnosis and management of isolated angiitis of the central nervous | system. Neurology. 1989;39(2, pt 1):167-173. | 5. Vollmer TL, Guarnaccia J, Harrington W, Pacia SV, Petroff OA. Idiopathic granulomatous | angiitis of the central nervous system: diagnostic challenges. Arch Neurol. | 1993;50(9):925-930. | 6. Crane R, Kerr LD, Spiera H. Clinical analysis of isolated angiitis of the central | nervous system: a report of 11 cases. Arch Intern Med. 1991;151(11):2290- | 2294. | 7. Hajj-Ali RA, Furlan A, Abou-Chebel A, Calabrese LH. Benign angiopathy of the | central nervous system: cohort of 16 patients with clinical course and long-term | follow-up. Arthritis Rheum. 2002;47(6):662-669. |