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Takayasu Arteritis and PRES Syndrome - A Rare Case Series

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Abstract
PRES is a very rare manifestation of Takayasu's arteritis. It can present with headache, seizures, altered sensorium with visual disturbances. Two young teenager patients presented in ward with complaints of severe headache, Hypertention , vomiting ,blurring of vision followed by GTCS and altered sensorium for about one month.(1) Their routine investigations like CBC,LFT,RFT appears to be normal with raised ESR.CE-MRI of brain shows focal areas of hyperintensity scattered in white matter signs of PRES.CT angiography of abdominal aorta showed possibility of autoimmune vasculitis with bilateral proximal renal artery stenosis ,critical stenosis on right with small right kidney and decreased enhancement. Patients were put on antihypertensive and antiepileptic medication along with steroids to manage Takayasu's Arteritis with PRES. These patient's respond's symptomatically and were referred to higher centre for recanalisation of renal artery stenosis.

Keywords - *PRES-Posterior reversible encephalopathy syndrome, TA-Takayasu Arteritis, GTCS- Generalised Tonic Clonic seizures, CBC- Complete blood count, LFT- Liver fumction test, RFT-Renal function test.*

Introduction

PRES is a neuroradiological condition associated with headache, seizure, altered sensorium, visual disterbences and characteristic lesion on neuroimaging. It was first described by Hinchey et al in 1996.Pres as the presenting manifestation of Takayasu arteritis is quite rare.(2)

Case Series

Case 1: A 15 yrs old female presented with complaints of severe headache associated with projectile vomiting, blurring of vision and altered sensorium since around one month.

Patient was admitted and management carried out. Routines investigations along with CECT Abdomen, CE-MRI scan brain, fundus examination was done.

Patient's hemogram was normal, Hb 13.0 g/dl, liver profile, renal profile and Lipid profile was normal. Urine routine and microscopy was normal.ESR 60 mmFHR.

HIV and HbsAg negative.

Echocardiography shows LVH.

Doppler shows dampand wave and severely reduced velocity of origin of right renal artery and distal part of the renal artery not visualised with small atrophic right kidney s/o Takayasu arteritis.[3]

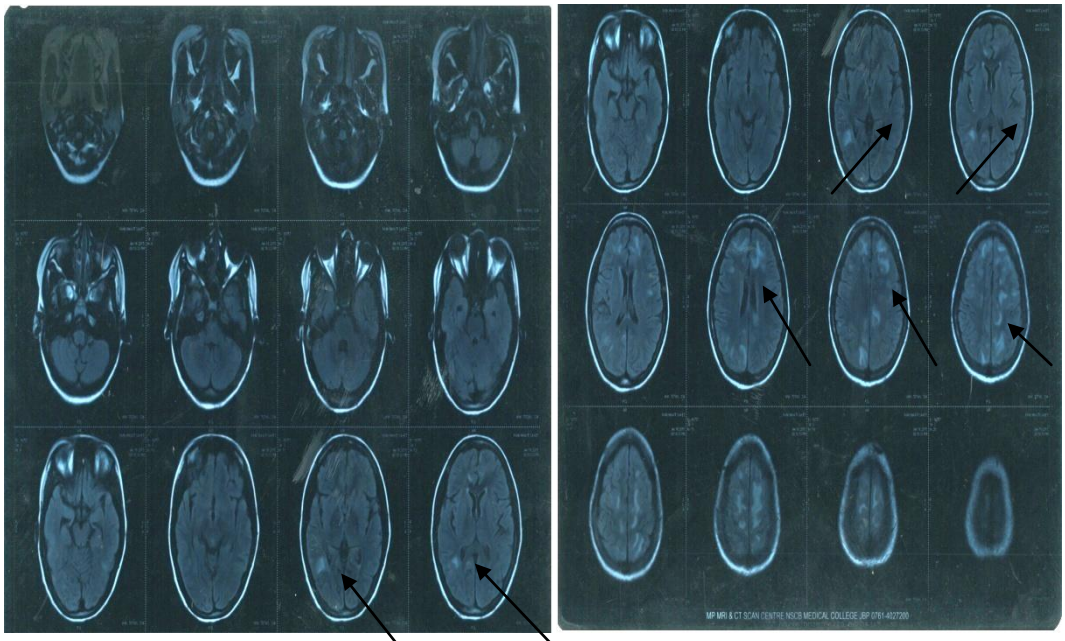
Fundus examination signs of hypertensive retinopathy present.

CE-MRI Brain(16 Jan): Signs of flair signals in white matter of both high parietal ,left parietal ,b/l frontal and left occipital region with no restriction of diffusion s/o PRES.(2) CECT Angiography of abdominal aorta shows possibility of takayasu arteritis.[4]

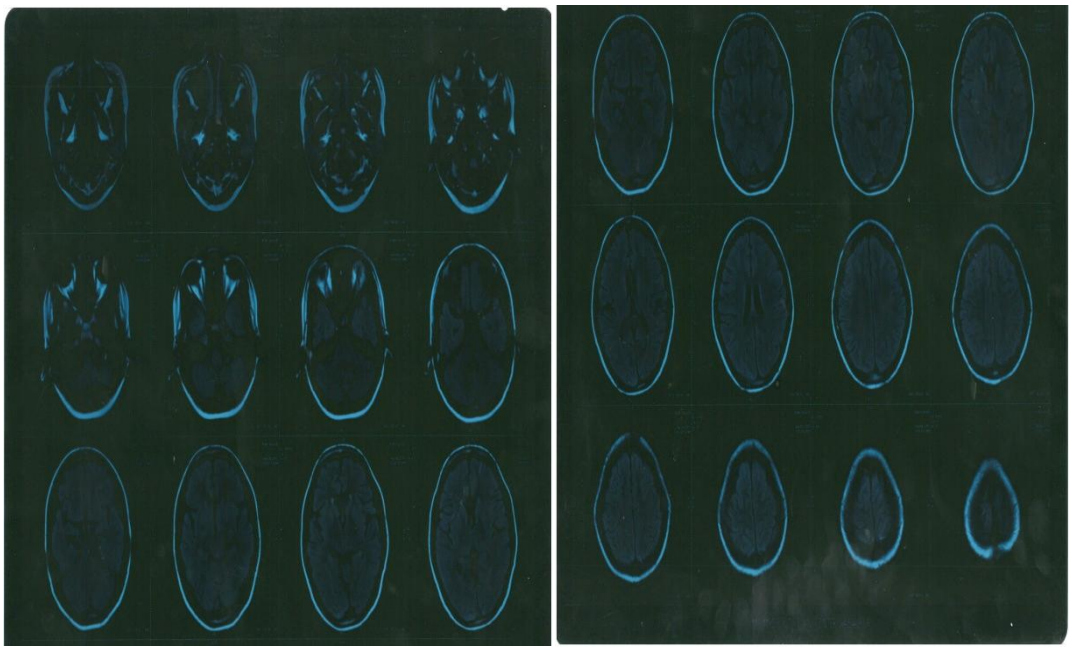
Repeat CEMRI brain (4 feb): No abnormal parenchymal enhancement or focal lesion.

CE-MRI BRAIN IMAGES

On admission (16 Jan 2015) [2] : Changes of PRES seen.



On discharge(04 Feb 2015): Complete resolution.



Case 2: A 16 yrs old male patient comes with complaints of severe headache, vomiting ,abnormal body movement and altered sensorium, blurring of vision and abdominal pain since one month.

Patient was admitted and management carried out. Routines investigations along with CE-CT abdomen, CE-MRI scan brain, fundus examination done.

Patient's hemogram was normal Hb-14.3 g/dl, liver profile, renal profile and Lipid profile was normal. Urine routine and microscopy was normal. ESR 50 mm FHR.

HIV and HbsAg Negative

Echocardiography no abnormality seen

Doppler (aorta + renal artery) shows circumferential wall thickening with luminal narrowing seen in abdominal aorta at region just distal to origin of renal arteries and also showing severe stenosis in the proximal part of right renal artery with small right kidney.This favours the diagnosis of PRES with takayasu arteritis type IV(3). CECT Angiography of abdominal aorta shows possibility of takayasu arteritis.[4]

Fundus examination was normal.

CE-MRI Brain signs of PRES.

Past history –Not significant

Family history- Not significant

Outcome

Patient was put on oral Nifedipine, Prazosin, Phenytoin, and Steroids. Patient improved his headache, vomiting, and no seizure episodes after that. Then patient was discharged on request and advised to go higher centre for revascularisation surgery for stenosis of renal artery.

Discussion

History : 1908 in Japan (7) by Hinchey et al.

TA, a granulomatous panarteritis, affects the aorta and its major branches, typically before the age of 40 years. The disease predominates in women, with a female-to-male ratio of up to 10:1. Because the diagnosis is often delayed, substantial arterial injury accures [1] .TA may be accompanied by symptoms of Raynaud phenomenon or upper extremity claudication, and carotidynia occurs in up to 25% of patients. The aorta may be involved throughout its length, and even though any branches can be diseased, the most commonly affected are the subclavian and common carotid arteries. Stenotic / occlusive arterial lesions are found in more than 90% of patients, whereas aneurysms are reported in approximately 25%. The pulmonary arteries are involved in up to 50% of patients, and aortic valve regurgitation and coronary arteritis may occur. Renal artery stenosis occurs in 28-75% of the cases.

PRES is a clinicoradiological syndrome. The syndrome is not always reversible and can cause permanent damage if not treated immediately. It is not always confined to posterior regions of brain and white matter. It can involve anterior regions of brain and also can involve the gray matter. [8]

The clinical manifestations are headache, altered consciousness, visual disturbances and seizures. The neurologic syndrome is usually preceded by a hypertensive crises.[9]

The pathophysiology is not clear but two contradictory hypotheses have been proposed.The first hypothesis is due to impaired autoregulation with cerebral hyperperfusion and the second is due to endothelial dysfunction with cerebral hypoperfusion. Both mechanisms result in blood brain barrier dysfunction with cerebral vasogenic oedema.[10]

Diagnosis

PRES as the presenting symptom of TA is quite rare. On imaging (CT/MRI) the brain shows regions of symmetric hemisphere oedema. The parietal and occipital lobes commonly affected followed by frontal lobe, inferior temporal, occipital junction and the cerebellum. The posterior part of the brain is more affected due to relative paucity of sympathetic innervations. The lesions of PRES are best visualized with imaging with Flair sequence.

Treatment

Treatment of PRES is solely directed towards the underlying cause. These patients require

antihypertensives, antiepileptics along with steroids [5] to manage Takayasu arteritis with PRES. To control the cerebral edema anti cerebral edema measures also required.[6]

Important Guidelines

As the PRES as a presenting manifestation of Takayasu arteritis is quite rare and the syndrome is not always reversible. So when any suspicion of PRES occurs in any patient clinician must be very watchful for early diagnosis (by relevant investigations) and treatment; as the condition is reversible when diagnosed early and treated properly.

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