ISSN: 2581 – 3633
PubMed - National Library of Medicine - ID: 101745081

International Journal of Medical Science and Advanced Clinical Research (IJMACR)

Available Online at: www.ijmacr.com

Volume – 6, Issue – 1, Janaury - 2023, Page No. : 94 - 100

Takayasu Arteritis Presenting As Pres: Posterior Reversible Enchephalopathy Syndrome

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How to citation this article: Dr Kousar Begum, Dr Bindu C B, Dr Muthuraj N, Dr. Ashwath K S, "Takayasu Arteritis Presenting As Pres: Posterior Reversible Enchephalopathy Syndrome", IJMACR- January - 2023, Volume – 6, Issue - 1, P. No. 94 – 100.

Type of Publication: Case Report

Conflicts of Interest: Nil

Abstract

Takayasu arteritis (TA) is a rare, systemic, chronic inflammatory condition causing granulomatous vasculitis of medium-sized and large arteries of unknown etiology. Posterior Reversible Encephalopathy Syndrome (PRES) and seizure are rare complications. We report a case of Takayasu arteritis with known seizure disorder presented with status epilepticus and Posterior reversible encephalopathy syndrome and segmental pulmonary embolism.

Keywords: Takayasu Arteritis, Posterior Reversible Encephalopathy Syndrome, Status Epilepticus.

Introduction

Takayasu arteritis (TA) is a primary vasculitis of unknown etiology that affects medium- sized and large arteries, primarily the aorta, its main branches, and the pulmonary arteries.[1] The other commonly affected arteries are coronary, renal and internal carotid. Histopathologically, affected vessels initially show mononuclear cell infiltrations in the adventitia and granulomas with Langerhans cells in the media, followed by disruption of the elastin layer and subsequent medial and intimal fibrosis leading to stenosis, occlusion, dilatation, and aneurysm formation in the affected vessels.[2] The etiopathogenesis of this disease suggest an autoimmune basis along with genetic and environmental factors also play an important role.[3] Posterior reversible encephalopathy syndrome (PRES) is a rare complication of Takayasu Arteritis. It is a neuroradiological condition associated with headache, seizure, altered sensorium, visual disturbance and typical lesion on MRI brain.[4]

Case Report

A 35 year old lady who is a known seizure disorder not on regular medications presented with fever for 1 day andheadache since 3 days withmutiple episodes of generalised tonic clonic seizures without gain of consciousness between the episodes. On arrival patient was in post-ictal state, with no pulse in the upper limbs and BP not recordable and lower limb pulses being very feeble,however BP was recordable in right popliteal artery and with left carotid was just palpable.Physical examination revealed no focal neurodeficits, with normal cardiorespiratory examination. Initially patient was treated with IV antiepileptics, and IV fluids . Metabolic parameters were within normal limits. LP-CSF done i/v/o suspected meningoencephalitis were normal. In view of persistent low GCS, MRI brain to rule out structural lesions was done and revealed white matter changes in the subcortical matter in posterior parietal lobe region suggestive of PRES - ?vasculitis. .Patient was subjected to CT aortogram revealed dilated ascending aorta multiple foci of moderate luminal narrowing in right subclavian artery and proximal left common carotid, vertebral, subclavian artery with collateral formation with normal abdominal , renal vasculature. Luminal filling defect was noted in the segmental pulmonary artery of left upper lobe and right lower lobe. In view of above, diagnosis of large vessel vasculitis- Takayasu arteritis was made. EEG done in view of seizure revealed generalised spike-wave epileptiform discharges. Fundoscopy was normal and 2DECHO done was normal. Patient recovered in 72 hours with antioedema measures.MRI Brain angiography was done to look for intracerebral arterial stenosis and was found to be normal. Evaluation for pulmonary embolism was done with ANA screening and was 1+ and with Cytoplasmic-antineutrophil cytoplasmic antibodies(c-ANCA) and perinuclear - antineutrophil cytoplasmic antibodies (p-ANCA) was negative and was thought to be a part of vasculitis. Serum vitamin B12, homocysteine levels were normal. Rheumatologist opinion was taken and started on Methotrexate, folic acid, oral prednisolone and oral anticoagulation for pulmonary embolism.

Lab and Imaging Reports

Table 1

Parameters	Values	Units	Normal Range
Hb	10.8	mg/dl	12-14
TC	5080	/cu mm	4000-11000
DC-Neutrophils	80	%	60-75
Platelets	3.65	Lakh/cu mm	1.5-4
LFT-TB/DB AST/ALT	0.6/0.2 44/48	mg/dl IU/dl	0-1.2/0-0.2 <40
RFT	38/0.9	mg/dl	15-40/0.5-1.10
S.ELECTROLTYES	135/4.5/110	mg/dl	135-145/3.5-5/95-110
ABG-ph/pco2/po2/hco3	7.35/42/90/20	-/mmhg/mmhg/-	7.35-7.45/35-45/60-100/22-24
RBS	85	mg/dl	80-140

Urine routine	Normal		
ESR	36	Mm/ 1 hour	0-20
S. Calcium	8.2	mg/dl	8.5-10
TSH	4.5	IU/ml	0.5-4.5
HIV	negative	-	Negative
HbsAG	Negative	-	Negative
Anti-HCV	negative	-	Negative
ANA screening	1+	-	Negative
c-ANCA	negative	-	Negative
p-ANCA	negative	-	Negative
Peripheral smear	Normocytic normochromic anemia	-	-
HsCRP	14.6	mg/dl	0-6
Procalcitonin	**0.5**	ng/ml	0-0.08

Figure 1: Dilated ascending aorta multiple foci of moderate luminal narrowing in right Subclavian artery and proximal left common carotid, vertebral, Subclavian artery with collateral formation with normal abdominal, renal vasculature

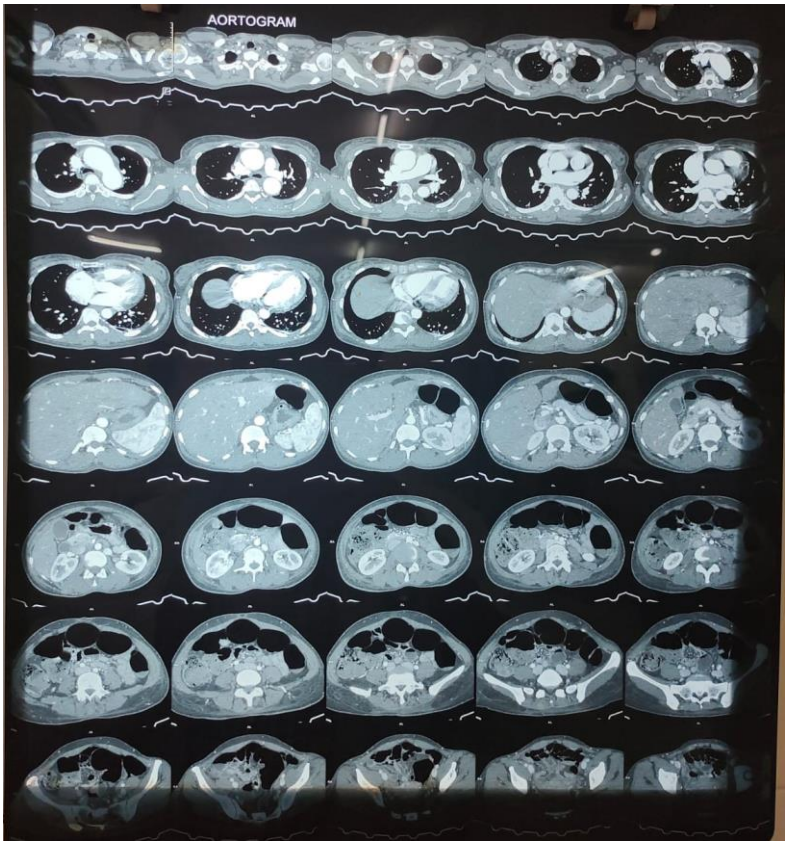


Figure 2: Luminal filling defect was noted in the segmental pulmonary artery of left upper lobe and right lower lobe.

MRI Brain Images

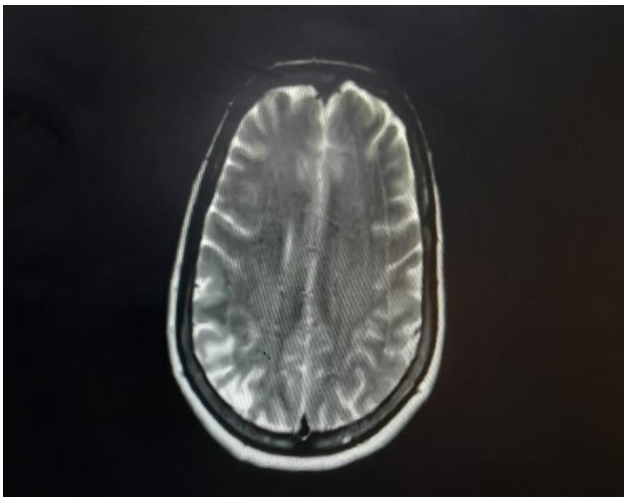


Figure 3a: MRI Brain - T2 Weighted Image

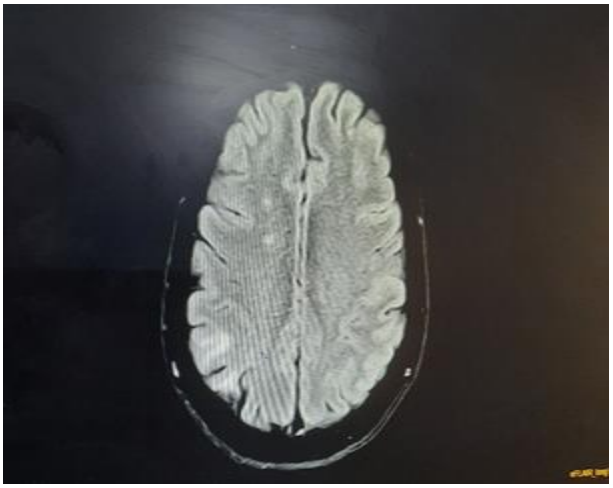


Figure 3b: MRI brain T2 FLAIR IMAGE showing white matter changes in the subcortical white matter in posterior parietal region with no restriction on ADC suggestive of PRES

Discussion

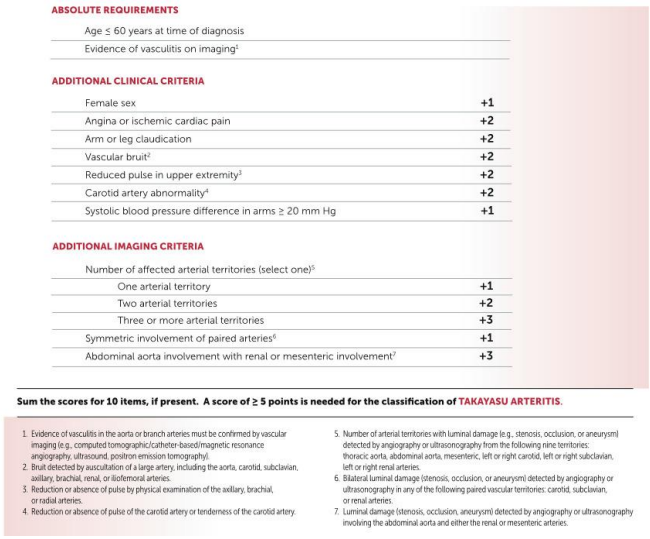
In 1908, Dr. Mikito Takayasu, professor of ophthalmology at Kanazawa University Japan, presented

the case of a 21 years old woman with characteristic fundal arteriovenous anastamoses.[5] In the same year, Onishi and Kagoshima each described similar cases associated with absent radial pulses.[6]

In 1920, the first postmortem case of a 25 years old woman demonstrated panarteritis and suggested that retinal ischaemia caused the fundal changes. In 1951, Shimizu and Sano summarised the clinical features of this "pulseless disease".[7] Takayasu arteritis, a granulomatous panarteritis, typically affects before the age of 40 years, predominates in women with a female-to-male ratio of up to 10:1.[8] Histologically, In the acute phase, T cells, B cells, granulocytes, dendritic cells, and macrophages infiltrate the medial layer of vessel wall that produce inflammatory cytokines and mediators that leads to inflammatory response and tissue injury.[9]Some class II antigen-presenting HLA molecules (particularly HLAB52 and HLA-DR4) have been implicated.[10]

We report a case of 35 years with known case of seizure disorder with constitutional symptoms, headache, fever and one episode of seizure lasting for 1 minute. Patient fulfills the diagnostic criteria of Takaysu arteritis on the basis of clinico-radiological findings.[10] Approximately 10% of patients are asymptomatic and others mostly presented with headache (50%-70%), malaise (35%-65%), arthralgia (28%-75%), fever (9%-35%) and weight loss (10%- 18%).[11] In our case MRI brain showed typical findings of Posterior reversible encephalopathy syndrome. PRES is a rare complication associated with Takaysu arteritis, that typically presents with headaches, seizures, painless visual loss, confusion, altered sensorium, and typical findings on neuroimaging.[12,13] MRI brain show vasogenic edema most frequently in the occipital lobes, followed by the parietal, frontal and temporal lobes.[14] In PRES, seizures are the most common clinical presentation (66– 87 %), followed by headache (28–53 %), visual abnormalities (20–42 %), and nausea or vomiting (42 %).[15] Our patient presented with all of these symptoms . Available clinical data shows that all patients with Takaysu arteritis and PRES presented with seizures but an uncommon manifestation of isolated Takaysu arteritis. Vasculitis-induced loss of cerebral autoregulation, accumulation of toxic metabolites, changes in blood flow, acidosis, hypoxemia, and blood–brain barrier dysfunction are the proposed mechanisms in the pathogenesis of PRES as well as seizure.[16,17] Our case is unique to be reported as it presented with rare manifestation of Takayasu arteritis in the form of PRES and had a significant clinical improvement with steroid, anticoagulation, antiepileptic and methotrexate.

Criteria for diagnosis



ABSOLUTE REQUIREMENTS	
Age ≤ 60 years at time of diagnosis	
Evidence of vasculitis on imaging[1]	
ADDITIONAL CLINICAL CRITERIA	
Female sex	+1
Angina or ischemic cardiac pain	+2
Arm or leg claudication	+2
Vascular bruit[2]	+2
Reduced pulse in upper extremity[3]	+2
Carotid artery abnormality[4]	+2
Systolic blood pressure difference in arms ≥ 20 mm Hg	+1
ADDITIONAL IMAGING CRITERIA	
Number of affected arterial territories (select one)[5]	
One arterial territory	+1
Two arterial territories	+2
Three or more arterial territories	+3
Symmetric involvement of paired arteries[6]	+1
Abdominal aorta involvement with renal or mesenteric involvement[7]	+3

Sum the scores for 10 items, if present. A score of ≥ 5 points is needed for the classification of TAKAYASU ARTERITIS.

1. Evidence of vasculitis in the aorta or branch arteries must be confirmed by vascular imaging (e.g., computed tomographic/catheter-based/magnetic resonance angiography, ultrasound, positron emission tomography).
2. Bruit detected by auscultation of a large artery, including the aorta, carotid, subclavian, axillary, brachial, renal, or iliofemoral arteries.
3. Reduction or absence of pulse by physical examination of the axillary, brachial, or radial arteries.
4. Reduction or absence of pulse of the carotid artery or tenderness of the carotid artery.
5. Number of arterial territories with luminal damage (e.g., stenosis, occlusion, or aneurysm) detected by angiography or ultrasonography from the following nine territories: thoracic aorta, abdominal aorta, mesenteric, left or right carotid, left or right subclavian, left or right renal arteries.
6. Bilateral luminal damage (stenosis, occlusion, or aneurysm) detected by angiography or ultrasonography in any of the following paired vascular territories: carotid, subclavian, or renal arteries.
7. Luminal damage (stenosis, occlusion, aneurysm) detected by angiography or ultrasonography involving the abdominal aorta and either the renal or mesenteric arteries.

American College of Rheumatology 2022 CRITERIA

Here in this case the score being 7, with absolute requirements and additional criteria (female sex, reduced pulse in upper extremity, carotid artery abnormality, vascular bruit)

Treatment

1. T.LEVETIRACETAM 500mg 1-0-2 x daily
2. T.WARFARIN 2MG 0-0-1 x Daily @ 6pm
3. T.METHOTREXATE 7.5MG once weekly
4. T.FOLIC ACID 5MG 1-0-0 x Daily
5. Not to swim/drive Watch for bleeding manifestations
6. Regular INR monitoring monthly

Conclusion

We report the case with a rare case of Takayasu arteritis and which presented with even rare manifestation of the disease in the form of posterior reversible encephalopathy syndrome . The diagnosis was made on the basis of clinic-radiological and cytological findings. It's a potentially treatable condition, so physicians should be aware of its varied clinical presentation.

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