International Journal of Research in Medical Sciences
Kumar M et al. Int J Res Med Sci. 2021 Jul;9(7):2121-2124
www.msjonline.org
pISSN 2320-6071 | eISSN 2320-6012

DOI: https://dx.doi.org/10.18203/2320-6012.ijrms20212537

Case Report

Posterior reversible encephalopathy and status epilepticus in Takayasu arteritis

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Received: 11 May 2021
Accepted: 07 June 2021

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ABSTRACT

Takayasu arteritis (TA) is a rare, systemic, chronic inflammatory condition causing granulomatous vasculitis of medium-sized and large arteries of unknown etiology. A possible relationship between Takayasu arteritis and tuberculosis has been suggested but not proven until now. Posterior Reversible Encephalopathy Syndrome (PRES) and seizure are rare complications. We report a case of Takayasu arteritis with lymph node tuberculosis presented with status epilepticus and Posterior reversible encephalopathy syndrome.

Keywords: Takayasu arteritis, Status epilepticus, Posterior reversible encephalopathy syndrome, Tuberculosis

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] Among the environmental factors, Mycobacterium tuberculosis (MT) has been suggested to be associated with it, but In spite of the clinical relationship between both, no evident link has been proved till now.[4] Takayasu arteritis are known to have augmented immune response to Mycobacterium tuberculosis antigens, specifically to its 65 kDa Heat shock protein.[5] 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.[6]

CASE REPORT

A 16 years old girl presented to our emergency on 27th March 2021, with worsening headache, recurrent vomiting, bilateral painless visual loss and status epilepticus. She had four months history of moderate to severe holocranial headache associated with nausea and/ or vomiting, fever, weight loss and decreased appetite. Seen by local physician and found to have hypertension and lymph node tuberculosis, started with antihypertensive and Anti-Tubercular drug. On examination she was drowsy, pulse right radial 100/minute, Left Radial and femoral feeble, bilateral carotids normal. Blood pressure in right arm 240/150 mmHg and in left arm 150/130 mmHg. Cardiac examination showed normal heart sounds. No bruit was heard. Respiratory examination normal. Other system examinations were within normal limits. Fundus revealed arteriovenous crossing changes.

Laboratory findings summarized in table 1. Erythrocyte sedimentation rate (ESR) was at 38 mm/h, C-reactive protein (CRP) was at 41.7 mg/L. Serum ferritin 10.17 ng/ml.

Table 1: Lab parameters.

	27.03.2021	01.03.2021	04.03.2021	07.03.2021
TLC (/cmm)	31000	7200	6100	4900
Hb (g/dl)	9.7	8.1	8.7	8.5
Platlets (lacs/ml)	6.24	3.35	2.87	2.88
Blood Urea (mg/dl)	70	54	30	25
S. Creatinine (mg/dl)	1.7	1.2	1.0	0.9
Na^+ (meq/L) / K^+(meq/L)	138/5.1	138/4.4	137/4.9	138/3.9
ALT/AST (IU/L)	25/34	21/43	72/80	60/53
Total bilirubin (mg/dl)	0.9	0.6	0.6	0.6
Random blood sugar (mg/dl)	85	112	109	95

Viral markers were negative for hepatitis B and C, human immunodeficiency virus, cytomegalovirus and Epstein-Barr virus. The serological study for anti-nuclear antibody (ANA), cytoplasmic anti-neutrophil cytoplasmic antibodies (c-ANCA), and perinuclear anti-neutrophil cytoplasmic antibodies (p-ANCA), antiphospholipid and anticardiolipin antibodies were negative. Cerebrospinal fluid examination was normal. USG Kidney- normal size and shape with normal echogenicity. Doppler showed reduced flow and narrowing in left renal artery and left subclavian. On MRI brain, hyperintensity noted predominantly in sub cortical white matter involving bilateral frontal, temporal and occipital lobe. Also involving deep white matter bilaterally without any restriction diffusion or blooming. No contrast enhancement seen.

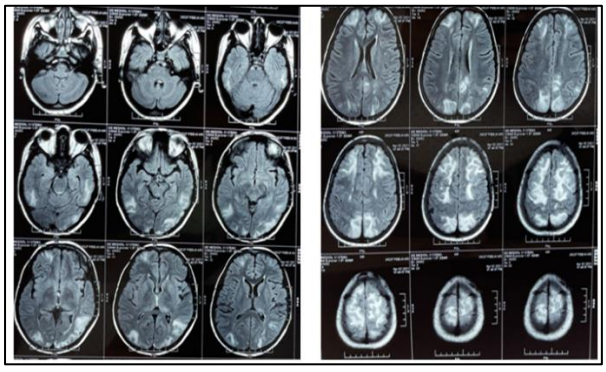

Figure 1: FLAIR image showing hyperintense lesion in occipital, frontal and temporal lobes.

Bilateral gangliocapsular region, thalamus, brainstem, ventricles and cerebellum appear normal in signal intensity. MR angiogram brain normal. Findings suggestive of Posterior reversible encephalopathy syndrome (PRES). [Figure 1 and 2] CT angiography showed marked attenuation with diffuse thickening of wall of left subclavian and non-visualization of left renal artery with progressive attenuation of abdominal aorta, suggestive of large vessel vasculitis. (Figure 3)

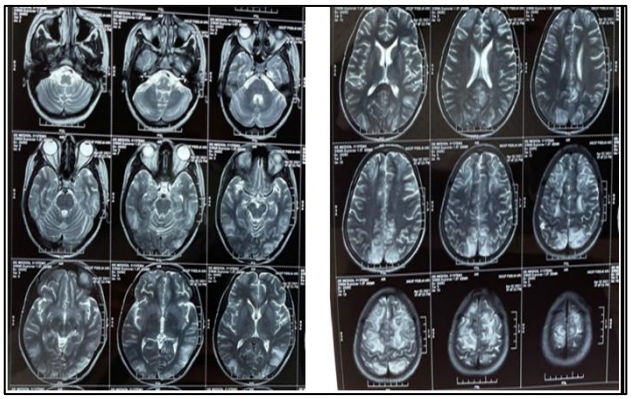

Figure 2: T2W image showing hyperintense lesion in occipital, frontal and temporal lobes.

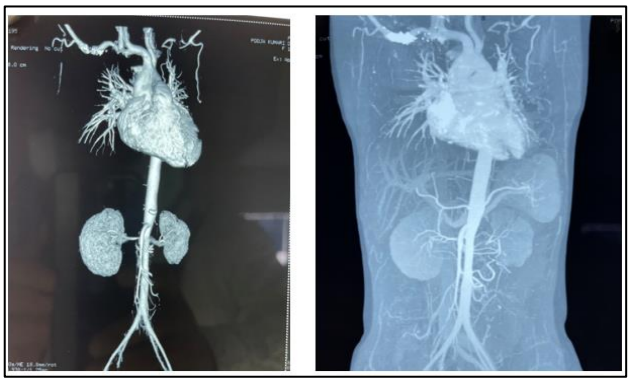

Figure 3: CT angiogram showing marked attenuation with diffuse thickening of wall of left subclavian and non-visualization of left renal artery.

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.[7] In the same year, Onishi and Kagoshima each described similar cases associated with absent radial

pulses.[8] 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".[9] 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.[10] 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.[11] Some class II antigen-presenting HLA molecules (particularly HLA-B52 and HLA-DR4) have been implicated.[12] In our case Takayasu arteritis was associated with cytologically proven cervical lymph node tuberculosis. In a study by Lim et al found much higher frequencies of tuberculosis associated with Takayasu arteritis compared to the general population.[13] Lupi-Herrera et al. in a retrospective study showed that 21% of Takayasu arteritis patients had tubercular lymphadenitis whereas only 1% had pulmonary tuberculosis.[14] Another study of 267 Korean patients with Takayasu arteritis, 47 had Tuberculosis, with a high prevalence of tubercular lymphadenitis (12/47, 25.5%).[13] Tubercular lymphadenitis seems to be more common in young female patients and most common involvement of cervical lymph node.[15]

We report a case of 16 years hypertensive girl with constitutional symptoms, headache, vomiting, bilateral painless visual loss with status epilepticus. 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%).[16] In our case MRI brain showed typical findings of Posterior reversible encephalopathy syndrome. CT angiography showed marked attenuation with diffuse thickening of wall of left subclavian and non visualization of left renal artery. 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.[17,18] MRI brain show vasogenic edema most frequently in the occipital lobes, followed by the parietal, frontal and temporal lobes.[19] 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 %).[20] Our patient presented with all of these symptoms over a period of few months. Available clinical data shows that all patients with Takaysu arteritis and PRES presented with seizures but an uncommon manifestation of isolated Takaysu arteritis. Hypertension-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.[21,22] Our case is unique to be reported as it presented with rare manifestation of Takayasu arteritis in the form of PRES and status epilepticus. Had a significant clinical improvement with steroid, antiplatelet, antiepileptic and anti-tubercular drugs.

CONCLUSION

We report the case with a rare co-occurrence of Takayasu arteritis and lymph node tuberculosis which presented with even rare manifestation of the disease in the form of posterior reversible encephalopathy syndrome and status epilepticus. 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.

ACKNOWLEDGEMENTS

We are grateful to the Department of Radiology, Patna Medical College, Patna for their contribution.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Johnston SL, Lock RJ, Gompels MM. Takayasu arteritis: a review. Journal of Clinical Pathology. 2002;55(7):481-6.
2. Vaideeswar P, Deshpande JR. Pathology of Takayasu arteritis: A brief review. Annals of Pediatric Cardiology. 2013;6(1):52-8.
3. Brunner J, Feldman BM, Tyrrell PN, Kuemmerle-Deschner JB, Zimmerhackl LB, Gassner I, et al. Takayasu arteritis in children and adolescents. Rheumatology (Oxford) 2010;49:1806-14.
4. Duzova A, Türkmen O, Cinar A, Cekirge S, Saatci U, Ozen S. Takayasu's arteritis and tuberculosis: A case report. Clin Rheumatol 2000;19:486-9.
5. Aggarwal A, Chag M, Sinha N, Naik S. Takayasu arteritis: role of Mycobacterium tuberculosis and its 65 kDa heat shock protein. Int J Cardiol. 1996;55(1):49-55.
6. Zaki SA, Chavan V, Shanbag P. Unusual presentation of Takayasu's Arteritis as posterior reversible encephalopathy syndrome. Ann Indian Acad Neurol. 2011;14(3):214-6.
7. Takayasu M. A case with peculiar changes of the retinal central vessels [in Japanese]. Acta of the Opthalmic Society of Japan. 1908;12:554-5.
8. Numano F. Introductory remarks for this special issue of Takayasu arteritis. Heart Vessels. 1992;(Suppl. 7):3-5.
9. Shimizu K, Sano K. Pulseless disease. Journal of Neuropathology and Clinical Neurology. 1951;1.37-1.47.
10. Arend WP, Michel BA, Bloch DA. The American College Of Rheumatology 1990 criteria for the classification of Takayasu arteritis. Arthritis Rheum. 1990;33:1129.

11. Arnaud L, Haroche J, Mathian A, Gorochov G, Amoura Z. Pathogenesis of Takayasu's arteritis: a 2011 update. Autoimmun Rev. 2011;11:61-7.
12. Johnston SL, Lock RJ, Gompels MM. Takayasu arteritis: a review. J Clin Pathol. 2002;55:481-6.
13. Lim AY, Lee GY, Jang SY, Gwag HB, Choi SH, Jeon E-S et.al. Comparison of clinical characteristics in patients with Takayasu arteritis with and without concomitant tuberculosis. Heart Vessels. 2016;31:1277-84.
14. Lupi-Herrera E, Sánchez-Torres G, Marcushamer J, Mispireta J, Horwitz S, Vela JE. Takayasu's arteritis. Clinical study of 107 cases. Am Heart J. 1977;93:94-103.
15. Handa U, Mundi I, Mohan S. Nodal tuberculosis revisited: a review. J Infect Dev Ctries. 2012;6:6-12.
16. Park MC, Lee SW, Park YB. Clinical characteristics and outcomes of Takayasu's arteritis: analysis of 108 patients using standardized criteria for diagnosis, activity assessment, and angiographic classification. Scand. J. Rheumatol. 2005;34(4):284-92.
17. Wakida K, Morita H, Sakurai T, NishidaH. A case of posterior reversible encephalopathy syndrome associated with Takayasu's arteritis. Rinsho Shinkeigaku. 2016;56(3):174-9.
18. Camara-Lemarroy CR, Lara-Campos JG, Perez-Contreras E, Rodríguez-Gutiérrez R, Galarza-Delgado DA. Takayasu's arteritis and posterior reversible encephalopathy syndrome: a case-based review. Clin Rheumatol. 2013;32(3):409-15.
19. Li K, YangY, GuoD, SunD, LiC. Clinical and MRI features of posterior reversible encephalopathy syndrome with atypical regions: a descriptive study with a large sample size. Front Neurol. 2020;24(11):194.
20. Lee VH, Wijdicks EF, Manno EM, Rabinstein AA. Clinical spectrum of reversible posterior leukoencephalopathy syndrome. Arch Neurol. 2008;65:205-10.
21. Yaffe K, Ferriero D, Barkovich AJ, Rowley H. Reversible MRI abnormalities following seizures. Neurology. 1995;45:104-8.
22. Ong B, Bergin P, Heffernan T, Stuckey S. Transient seizure- related MRI abnormalities. J Neuroimaging 2009;19:301-10.

Cite this article as: Kumar M, Anand V, Nadeem R. Posterior reversible encephalopathy and status epilepticus in Takayasu arteritis. Int J Res Med Sci 2021;9:2121-4.