Case Report

Indian Journal of Emergency Medicine
Volume 4 Number 2, April - June 2018
DOI: http://dx.doi.org/10.21088/ijem.2395.311X.4218.19

A Case of Rheumatoid Cerebral Vasculitis Presenting to the ED

Mayank Kumar[1], Pankaj Jhaldiyal[2], Indranil Das[3]

Author's Affiliation:
[1]PGY2 [2]HOD & Senior Consultant [3]Attending Consultant, Department of Emergency Medicine, Max Hospital, Dehradun, Uttarakhand 248001, India.

Corresponding Author:
Mayank Kumar,
PGY2, Department of Emergency Medicine, Max Hospital, Dehradun, Uttarakhand 248001, India.
E-mail: drmkmayank@gmail.com

Received on 23.03.2018,
Accepted on 23.04.2018

Abstract

Central nervous system involvement in rheumatoid arthritis is infrequent. The most frequent neurological manifestations of rheumatoid arthritis are peripheral neuropathy and cervical spinal cord compression due to subluxation of the cervical vertebrae. Cerebral rheumatoid vasculitis is an uncommon and serious complication which can be life-threatening Neurological involvement in RA is rare, present in only 1% of patients. Disorders of the central nervous system (CNS) include cervical myelopathy, vasculitis, RNs located within the CNS, or meningitis. Stroke also occurs with increased frequency. CNS vasculitis is extremely rare. The diagnosis is supported by magnetic resonance imaging (MRI), alone or with magnetic resonance angiography (MRA), showing the segmental vascular stenosis characteristic of vasculitis. Peripheral neuropathy is usually manifested as sensorimotor neuropathy or mononeuritis multiplex. The underlying mechanism is small vessel vasculitis of the vasa vasorum of the nerves with ischaemic neuropathy and demyelinisation as part of the rheumatoid vasculitis (RV) syndrome. Rheumatoid vasculitis typically affects small and medium-size blood vessels. It is associated with high rates of premature mortality with up to 40% of patients dying by 5 years as well as significant morbidity due to both organ damage from vasculitis and consequences of the treatment. High levels of circulating immune complexes have been observed in patients with rheumatoid vasculitis, and in particular high serum levels of rheumatoid factor are often detected at the time of onset of vasculitis. Deposition of immune complexes most likely contributes to small vessel inflammation and organ damage. Anti CCP levels also tend to be higher in patients with RA who have systemic vasculitis than in those who do not. Histologically, rheumatoid vasculitis involves blood vessels of the small arteries, and all layers of the vessel wall are infiltrated by neutrophils, lymphocytes, and plasma cells.

Keywords: Cerebral Vasculitis; Rheumatoid Arthritis; Cerebral Magnetic Resonance Imaging; Central Nervous System.

Case Report

A 71 years old female was admitted with complaints of involuntary limb movements with aphasia and disorientation and seizure like activity since one day. Pt had history of recent onset seizure disorder since past 3 months and a recent episode of tia. Patient is a case of rheumatoid arthritis since 30-40 yrs (on treatment from tab. folitrax 7.5 mg weekly once and tab.hcq 200 mg bd daily).

Initial vitals

Breathing Assessment

Respiration(RR/min): 30/min

Laboured: YES

SpO2: 90% on Room Air

SpO2: 100% on O2

Pulse: 100/min

BP: 140/70

Temperature: 101 F

Cardiac Monitor: sinus rhythm

GRBS: 140mg/dl

Her neurological examination: plantar reflexes were b/l extensor.

Recent lab reports : hb:10 tlc:18.7 plat:306 p-anca and ana profile negative csf viral panel reports were negative. Csf culture reports were sterile. Esr:75 anti-ccp:1620 (18/10/2017) 1390(09/11/2017) gene xpert for tb negative csf: tlc:80, neutrophil:75, lymphocyte:25,glucose:51, protein:107 meningoencephalitis panel:negative autoimmune panel: negative.

CT Head / Brain–Ischemic changes in bilateral periventricular and centrum semiovale with moderate ventriculomegaly. Suspected loss of gray white matter differentiation in the right high frontal region.

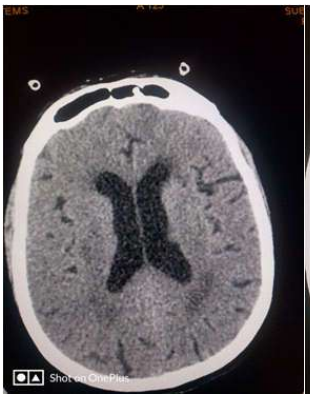

Fig. 1:

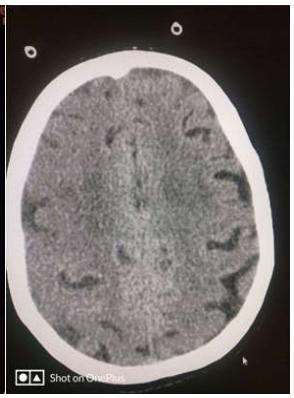

Fig. 2:

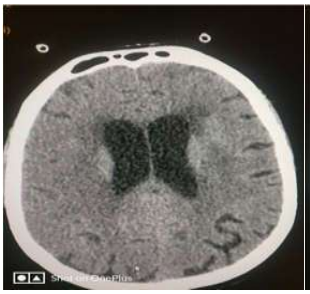

Fig. 3:

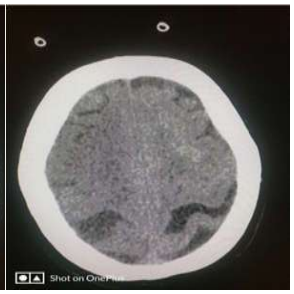

Fig. 4:

Eeg: suppression of background activity with burst focal spike and wave discharges over the right hemisphere suggestive of focal epilepsy.

Mri Brain with Contrast: Diffuse pachymeningeal enhancement along bilateral high frontoparietal convexity with suggestion of CSF loculations and adhesions in aqueductal region with subacute infarct in right thalamus as described.

Patient was put on 4 oral antiepileptics (phenytoin, clobazam, leviteracetam and lamotrigine)and inj methylprednisolone. Antibiotics inj ceftriaxone and antiviral inj acyclovir-course completed. Pt had intermittent focal seizure controlled with medical management. Pt was stable at the time of discharge.

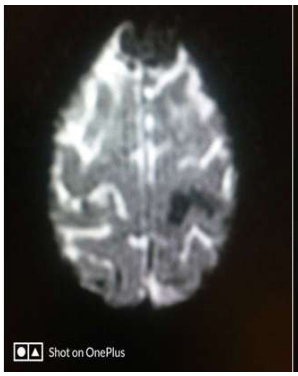

Fig. 5:

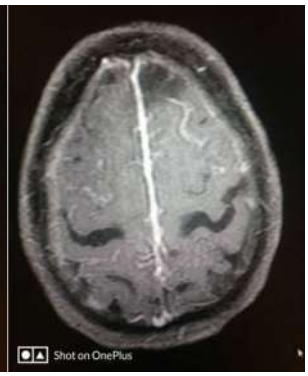

Fig. 6:

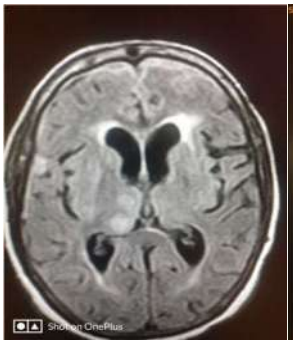

Fig. 7:

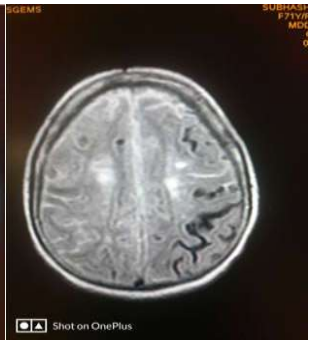

Fig. 8:

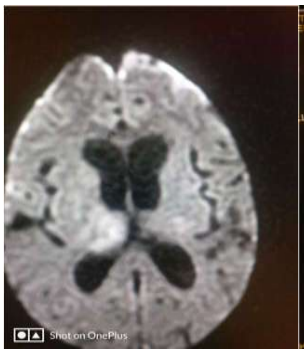

Fig. 9:

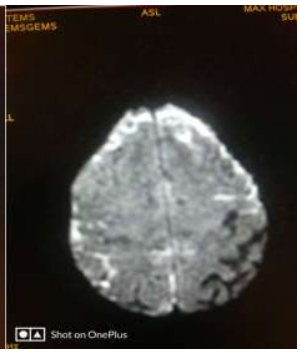

Fig. 10:

Discussion

Rate of occurrence of cerebral vasculitis in patients with RA is 1% to 8%. Neurological manifestations in patients with RA can be due to inflammatory central nervous system lesions. Cerebral vasculitis is usually associated with severe general signs as well as prominent extra-articular manifestations with minimal joint manifestations. Neurological involvement in RA includes atlantoaxial subluxation, polymyositis, mononeuritis multiplex, peripheral neuropathy, rheumatoid nodules in the central or peripheral nervous system, and rheumatoid vasculitis causing stroke and/or neuropathy The neurological manifestations of rheumatoid cerebral vasculitis include focal signs such as hemiplegia, partial epilepsy, cranial nerve involvement, or visual field loss, altered consciousness, confusion, and cognitive impairment or dementia. Neurological involvement in rheumatic disease is associated with high morbidity and, in some cases, can be life-threatening. Early assessment and a high index of suspicion for recognized complications are essential in managing such patients. Although serious neurological complications in rheumatic disease appear to be rare, few studies have been conducted on their prevalence. Studies of larger cohorts of patients in multi-center settings are required to assess the management of such patients more fully.

References

1. Jose m gobernado, carlos leiva, jorge rabano, jose c alvarez-cermeno, a fernandez-molina Recovery from rheumatoid cerebral vasculitis, Journal of Neurology, Neurosurgery, and Psychiatry 1984;47:410-413.
2. Adam Joseph Brown*, Susan M Staugaitis, Alex Wu, Leonard H. Calabrese; Central Nervous System Vasculitis Complicating Rheumatoid Arthritis in a Patient on a TNF Inhibitor: a Causal Association? Case Report and Systematic Review; J Vasc 2016;1:2.
3. Paloma Vela, extra-articular manifestations of rheumatoid arthritis,:EMJ Rheumatol. 2014;1:103-112.
4. Vasculitis and Vasculopathy in Rheumatic Diseases Mislav Radicì and Josipa Radic DOI: 10.5772/55056.
5. Ramos-Casals M, Brito-Zerón P, Muñoz S, Soria N, Galiana D, et al. Autoimmune Diseases Induced by TNF-Targeted Therapies Analysis of 233 Cases. Medicine (Baltimore) 2007;86:242-51.
6. Calabrese LH, Molloy E, Berger J. Sorting out the risks in progressive multifocal leukoence-phalopathy. Nat Rev Rheumatol 2015;11:119-23.
7. Jarrett SJ, Cunnane G, Conaghan PG, Bingham SJ, Buch MH, et al. Anti-tumor necrosis factor-alpha therapy-induced vasculitis: case series. J Rheumatol. 2003 Oct;30(10):2287-91.
8. Saint Marcoux B, De Bandt M; CRI (Club Rhumatismes et Inflammation) Vasculitides induced by TNFalpha antagonists: a study in 39 patients in France. Joint Bone Spine 2006;73:710-13.
9. Baek W, Lee SG, Kim YS, Kim JH, Jun JB, et al. Fatal varicellazoster virus vasculopathy associated with adalimumab therapy. Arch Neurol 2012;69:1193-1196.
10. Vannucchi V, Grazzini M, Pieralli F, Giannotta M, Biagioni C, et al. Adalimumab-induced lupus erythematosus with central nervous system involvement in a patient with Crohn's disease. J Gastrointestin Liver Dis 2011;20:201-03.
11. Pons C, Montala N, Valverde J, Brell M, Ferrer I, et al. Isolated cerebral vasculitis associated with rheumatoid arthritis. Joint Bone Spine 2010;77:361-63.
12. Martínez-Taboada VM, Val-Bernal JF, Pesquera LC, Fernández-Llanio NE, Esteban JM et al. Demyelinating disease and cutaneous lymphocitic vasculitis after etanercept therapy in a patient with rheumatoid arthritis. Scand J Rheumatol 20006;35: 322-23.
13. Spath NB, Amft N, Farquhar D. Cerebral vasculitis in rheumatoid arthritis. QJM 2014;107:1027-29.
14. Hajj-Ali RA, Calabrese LH. Diagnosis and classification of central nervous system vasculitis. J Autoimmun J Autoimmun. 2014 Feb-Mar;48-49: 149-52.
16. Hajj-Ali RA, Singhal AB, Benseler S, Molloy E, Calabrese LH. Primary angiitis of the CNS. Lancet Neurol 2011;10:561-72.
17. Pirani CL, Bennett GA. Rheumatoid arthritis, a report of three cases progressing from childhood and emphasizing certain systemic manifestations. Bull Hosp Joint Dis. 1951;12:335–367. [PubMed].
18. Sokoloff L, Bunim JJ. Vascular lesions in rheumatoid arthritis. J Chronic Dis. 1957;5:668–687. doi: 10.1016/0021-9681(57)90075-9. [PubMed] [Cross Ref].
19. Johnson RL, Smyth CJ, Holt GW, Lubchenco A, Valentine E. Steroid therapy and vascular lesions in rheumatoid arthritis. Arthritis Rheum. 1959;2:224–229. doi: 10.1002/1529-0131(195906)2:3<224::AID-ART1780020305>3.0.CO;2-I. [PubMed] [Cross Ref].
20. Steiner JW, Gelbloom AJ. Intracranial manifestations in two cases of systemic rheumatoid disease. Arthritis Rheum. 1959;2:537–545. doi: 10.1002/1529-0131(195912)2:6<537::AID-ART1780020608>3.0.CO;2-Q. [PubMed] [Cross Ref].
21. Ouyang R, Mitchell DM, Rozdilsky B. Central nervous system involvement in rheumatoid disease. Report of a case. Neurology. 1967;17:1099–1105. doi: 10.1212/WNL.17.11.1099. [PubMed][Cross Ref].

22. Ramos M, Mandybur TI. Cerebral vasculitis in rheumatoid arthritis. Arch Neurol. 1975;32:271–75. doi: 10.1001/archneur.1975.00490460087014. [PubMed] [Cross Ref].

23. Watson P, Fekete J, Deck J. Central nervous system vasculitis in rheumatoid arthritis. Can J Neurol Sci. 1977;4:269–72. [PubMed].

24. Kiss G, Kelemen J, Bely M, Vertes P. Clinically diagnosed fatal cerebral vasculitis in long-standing juvenile rheumatoid arthritis. Virchows Arch. 2006; 448:381–83. doi: 10.1007/s00428-005-0106-0.[PubMed] [Cross Ref].

25. Rodriguez Uranga JJ, Chinchon Espino D, Serrano Pozo A, Garcia Hernandez F. Pseudotumoral central nervous system vasculitis in rheumatoid arthritis. Med Clin (Barc) 2006;127:438–39. doi: 10.1157/13092774. [PubMed] [Cross Ref].

26. Mrabet D, Meddeb N, Ajlani H, Sahli H, Sellami S. Cerebral vasculitis in a patient with rheumatoid arthritis. Joint Bone Spine. 2007;74:201–204. doi: 10.1016/j.jbspin.2006.08.004. [PubMed][Cross Ref].

27. Caballol Pons N, Montalà N, Valverde J, Brell M, Ferrer I, Martínez-Yélamos S. Isolated cerebral vasculitis associated with rheumatoid arthritis. Joint Bone Spine. 2010;77:361–63. doi: 10.1016/j.jbspin.2010.02. 030. [PubMed] [Cross Ref].

28. Ohno T, Matsuda I, Furukawa H, Kano T. Recovery from rheumatoid cerebral vasculitis by low-dose methotrexate. Intern Med. 1994;33:615–20. doi: 10.2169/internalmedicine.33.615. [PubMed][Cross Ref].

29. Sofat N, Malik O, Higgens CS. Neurological involvement in patients with rheumatic disease. QJM. 2006;99:69–79. [PubMed].

30. Yukio A, Sunako K, Eiichiro U, Kazuhiro I, Yoichiro H, Makoto U, Masayuki A. Involvement of the central nervous system in rheumatoid arthritis: its clinical manifestations and analysis by magnetic resonance imaging. Intern Med. 1995;34:188–191. doi: 10.2169/internalmedicine.34.188. [PubMed][Cross Ref].

31. Watts RA, Mooney J, Lane SE, Scott DG. Rheumatoid vasculitis: becoming extinct? Rheumatology. 2004;43:920–23. doi: 10.1093/rheumatology/keh210. [PubMed] [Cross Ref].

32. Pease CT, Haugeberg G, Morgan A, Montague B, Hensor EM, Bhakta BB. Diagnosing late onset rheumatoid arthritis, polymyalgia rheumatica, and temporal arteritis in patients presenting with polymyalgic symptoms. A prospective long-term evaluation. J Rheumatol. 2005;32:1043–46. Erratum in: J Rheumatol 2005;32:1852. [PubMed].

33. Singleton JD, West SG, Reddy VV, Rak KM. Cerebral vasculitis complicating rheumatoid arthritis. South Med J. 1995;88:470–74. doi: 10.1097/00007611-199504000-00019. [PubMed] [Cross Ref].

34. Cupps TR, Moore PM, Fauci AS. Isolated angiitis of the central nervous system. Prospective diagnostic and therapeutic experience. Am J Med. 1983;74:97–105. [PubMed].