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A CASE REPORT OF RECURRENT SPINAL LESION WITH RETINAL VASCULITIS - UNCOMMON PRESENTATION OF COMMON DISEASE

Dr. Naga KarthikVanukuri[1], Dr. Maheedhar Reddy Nardla[2], Dr. Philo Hazeena[3], Dr. Sundar Shanmugam[4], Dr. Shankar Venkatasubramanian[5],

[1]Senior Resident, Department of neurology, Sri Ramachandra Institute of Higher Education and Research (SRIHER), Porur, Chennai.

[2]Senior Resident, Department of Neurology, Sri Ramachandra Institute of Higher Education and Research (SRIHER), Porur, Chennai.

[3]Assistant Professor, Department of neurology, Sri Ramachandra Institute of Higher Education and Research (SRIHER), Porur, Chennai.

[4]Associate professor, Department of neurology, Sri Ramachandra Institute of Higher Education and Research (SRIHER), Porur, Chennai.

[5]Professor and Head of Department, Department of neurology, Sri Ramachandra Institute of Higher Education and Research (SRIHER), Porur, Chennai.

Conflicts of Interest: Nil

Corresponding author: Dr. Maheedhar Reddy Nardla

Abstract:

Intramedullary neurosarcoidosis /tuberculosis may be the first and only manifestation of the disease and clinical course may mimic an idiopathic inflammatory demyelinating syndrome both clinically and on neuroimaging results. We report a case of 26 yrs. old male with recurrent episodes of vision loss due to retinal vasculitis and urinary incontinence with paraparesis due to spinal intramedullary granulomatous lesion probably neurosarcoidosis or tuberculosis. A high index of suspicion and a search for granulomatous lesion at extraneural sites are required for an early diagnosis. Even in the absence of systemic symptoms, targeting sites other than neural tissue for potential biopsy can result in the identification of the disease process with substantially less morbidity than sampling neural tissue. A high index of suspicion for the diagnosis is required because early intervention is associated with a favourable outcome.

Keywords: Eales disease, Retinal vasculitis, spinal intramedullary lesion

Introduction

This case of recurrent intramedullary lesion with retinal vasculitis is presented for the rarity and posing a diagnostic challenge to the clinician. The differential diagnosis varied from granulomatous lesion to the connective tissue disorder and demyelinating lesions [(1)]. In this case the diagnosis is delineated by biopsy of extraneural tissue suggesting granulomatous disease Eales disease (tubercular -protein hypersensitivity) [(2).] We report this case not only because of rarity of presentation but also as it requires the high index of suspicion and also early diagnosis have the favourable outcome and it has many misleading masquerades which may delay the diagnosis leading to increased morbidity.

Case report*:*

A 26yrs old male presented with a history of urinary incontinence and weakness of all four limbs (LL's>UL's) which was sudden in onset and progressive, over the period of 30 days. He was diagnosed as transverse myelitis and treated with steroids after which the symptoms had improved gradually with persisting mild symptoms. In 2014 October the patient observed floaters and curtain falling in front of left eye with vision loss for which an ophthalmologist opinion was taken and was diagnosed as retinal vasculitis on FFA and laser photocoagulation was done. Vision recovery was full. In 2012 patient had developed sudden onset of weakness of both lower limbs with urinary incontinence and decreased sensation from mid thorax level, similar episode again in 2016 both of which improved with steroids. In 2017 patient again had left eye vision loss with floaters for which patient had undergone laser photocoagulation again and vision recovered. The patient was diagnosed as demyelinating disorder at outside hospital and was being treated with 1gm of methyl prednisolone every month since May 2017 (ANA, Anti-NMO done negative). In 2018 patient presented to our neurology department with weakness of both lower limbs with tripping of toes and fall with sensory loss below D9 level (Left> Right). No history of penile or oral ulcers, rash, arthralgias. No history of hearing loss. No s/o other cranial nerve symptoms. No history fever or weight loss. On examination patient vitals were normal. Cardiovascular, respiratory and abdominal examination was normal. The patient's higher mental functions were normal.Optic nerve-Bilateral visual acuity - normal. Colour vision normal.Fundoscopy was showing left eye-post laser status with tractional bands Right eye-Normal.

Motor system examination was showing features of increased tone in both lower limbs (LT>RT), Pyramidal pattern of weakness is present (LT>RT) DTR-exaggerated from D9 Level, Plantar bilateral extensor with absent superficial reflexes. Sensory system-All modality of sensation was decreased from D9 level. No signs of cerebellum or meningeal involvement. Blood investigation-CBC- normal.LFT, RFT–normal. Serum B12 levels-normal. Serum ACE levels,Serum Ca^{2+} level-normal.CSF analysis showing mild elevation in proteins of 51mg/dl and lymphocytosis with10 cells and normal ADA levels. HIV-negative. Line Immunoassay-negative. C-ANCA, P-ANCA-Negative. Montauxtest-Positive, TB PCR-negative, Gamma interferon-negative. CSFOCB and S.OCB-negative. Anti NMO antibodies-Negative, S.MOG-Negative. Fundus photography done showing post laser status with traction bands (figure1). MRI done-2018 showing intramedullary lesion for D9-D11 level (figure2). Old MRI done in 2016 having T1-T5-intramedullarylesion and in 2012 having C7-C6-intramedullary lesionT2 hyperintensity with cord expansion (figure3). CT thorax and neck done has Enlarged peri-hilar with pre tracheal and carinal lymph node. Cervical lymph node biopsy showing histopathological features of granulomatous lesion with langerhans cells and large caseating necrotic tissue s/o tuberculousetiology (figure4). The patient was started on Anti tubercular therapy and steroids along with physiotherapy. Patient has been followed up regularly for one year and occasionally thereafter with repeat imaging showed no new spinal lesions and decrease in extent of lesion with last visit on March 2020 patient having sustained clinical improvement with medical management and rehabilitation measures like physiotherapy and bladder exercises.

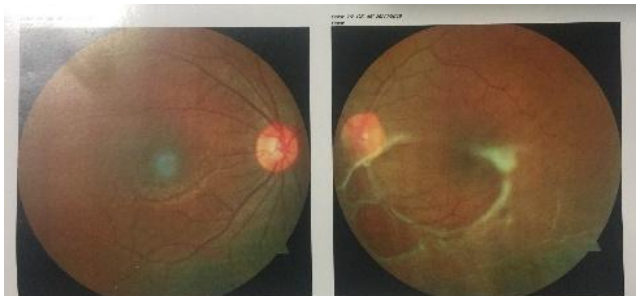

Figure 1: Fundus photo post laser with traction bands

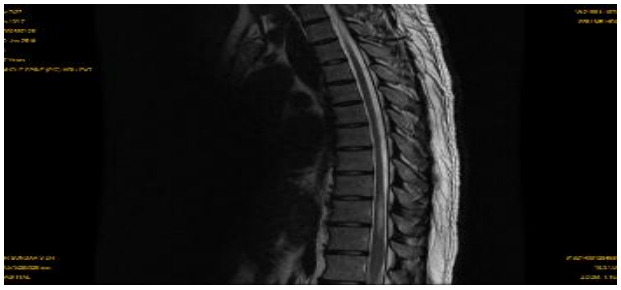

Figure 2: MRI spine D7-D11 intramedullary lesion (2018)

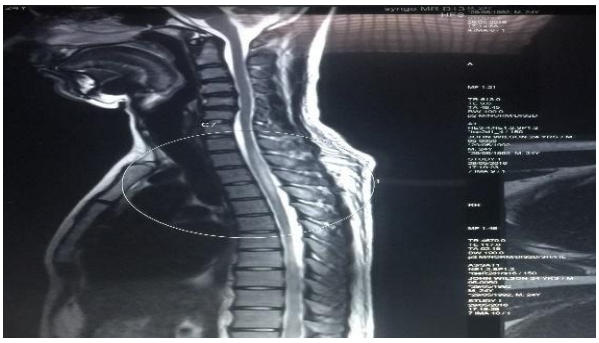

Figure 3: MRI spinal intramedullary C7-D6 lesion (2012)

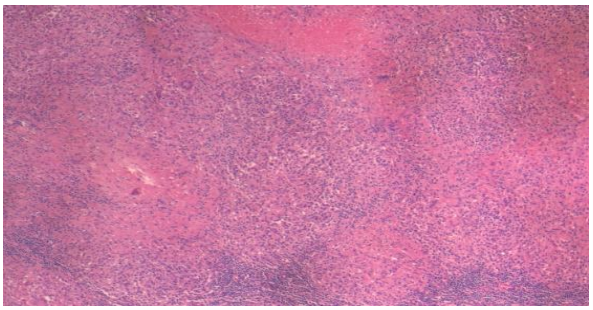

Figure 4: Cervical lymph node biopsy caseous necrosis

Discussion

The spinal Intramedullary lesion along with retinal vasculitis which was responding to steroids therapy and recurring on withdrawal of steroids was a very uncommon presentation posing a diagnostic challenge to treating clinician. The differential diagnosis for this type of presentations are systemic vasculitis seen as wegeners granulomatous, PAN, Churg-strauss and cryoglobulinemia, Behçet disease, SLE which were ruled out by considering clinical presentation and negative immunological profile. The Fundus Fluorescein angiography showing leakage of dye due to inner blood retinal barrier breakdown and staining of blood vessels walls by Fluorescein can be seen in sarcoidosis, multiple sclerosis and Eales disease.[(1)]Susacs syndrome can also cause this type of presentation by microangiopathy of arterioles of brain, retina, cochlea but in this case classical triad was not present hence not considered. CMV retinitis /toxoplasmosis with immunocompromised status can also present with retinal vasculitis with spinal lesions but was unlikely in our patient and CSF was not suggestive. Demyelinating lesion was not considered as diagnostic criteria for MS/NMO/NMO Spectrum disorders was not satisfied and OCB and NMO antibodies werenegative.[(2)]In this case extraneural tissue (cervical node) biopsy was showing caseating necrosis with langerhans cells suggesting tubercular pathogenesis of Eales disease (tuberculo-protein hypersensitivity) which requires to be treated with ATT and steroids. Mishra et al, [(3)] Singhalet al,[(4)] Das et al,[(5)] and sidharthSankar et al,[(6)] described similar cases in their experiences of Eales disease with neurological complications.

Conclusion

The spinal intramedullary lesion along with retinal vasculitis is a rare presentation and has an extensive differential diagnosis. The treating clinician needs to have a high index of suspicion and has to perform exhaustive workup to delineate the cause early. The features of FFA (Leakage of dye) along with other imaging studies and CSF analysis can provide a greater clue in diagnosis of the disease. Targeting the extraneural sites for potential biopsy can substantially reduce the morbidity for the patients. The early diagnosis and intervention with ATT and steroids has favourable outcome in Eales disease.

References

1. Ahmed M. Abu EL-Asrar, Carl Pet al. DD Of Retinal Vasculitis ,Middle East Afr J Opthalmology 2009 16(4):202-218.
2. El-AsrarAM ,AL Kharashi SA. Br J Opthalmology 2002;86:1248-51.
3. Mishra UK, Jha S, Kalitha j Stroke : A rare presentation of eales disease. A case report.Angiology 1996; 47:73-6.
4. Singhal BS, Dastur DK. Eales disease with neurological involvement J Neurol Science 1976; 27:313-21.
5. Das T, Bisswas J, Kumar A , Namperumalsamy P, et al. Eales disease. Indian J Opthalmology 1994;42:3-18.
6. SidharthSankar et al Neurology India vol 61, no4, july-august 2013.