Resistant Neuro-sarcoidosis: An Uncommon form of Sarcoidosis

Category: Medicine & Medical Specialties; Poster Presentation
Disclosure: The authors did not report any financial relationships or conflicts of interest
Supplemental Video

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Introduction: Neuro-sarcoidosis (NS) is a rare manifestation of sarcoidosis with only 5-10 % prevalence among the patients. Both central and peripheral nervous systems can be affected including involvement of meninges, parenchyma, spinal cord etc. NS is diagnosed by clinical manifestations along with neurodiagnostic testing which include leptomeningeal enhancement in contrast enhanced MRI. Lumbar puncture shows pleocytosis, elevated protein and low glucose levels. Glucocorticoids are the first line therapy. Immunomodulators like mycophenolate and infliximab can be used if patient worsens despite aggressive steroid therapy.

Case Presentation: We are presenting the rare case of resistant NS in a 68year old Caucasian lady involving cervical spine and cerebellum. The patient came with complaints of increasing generalized weakness and not being able to bear weight on her legs for 2 weeks. Physical examination showed weakness in both lower extremities, right (3/5) being weaker than the left (4/5). MRI scan of the brain and spine showed persistent leptomeningeal enhancement similar to older MRI with nodular enhancement in the posterior aspect of the cerebellum.

Diagnosis: The symptoms first started over a year ago. MRI brain done that time showed leptomeningeal enhancement in brainstem and enhancing nodules at C1 cord. CT chest showed enlarged mediastinal lymph nodes which were biopsied and they showed granulomatous inflammation with necrosis. Lumbar puncture was notable for lymphocytic pleocytosis (28), low glucose (39) and high protein (108). The patient was diagnosed with NS and started on high dose methylprednisolone followed by tapering dose of prednisone and methotrexate 4 times a week. Despite these, the patient presented with worsening features of NS and hence was diagnosed as a resistant case.

Management: Infliximab was added to the above medications. She receives Infliximab infusions 6-8 weekly and has received 4 infusions till now. She is clinically asymptomatic now and performs Activities of Daily Living independently.

Learning Objectives

1. To discuss about Neuro-sarcoidosis which is a relatively uncommon manifestation of sarcoidosis. It is diagnosed by combination of clinical features along with neurodiagnostic testing. It is typically associated with leptomeningeal enhancement with an abnormal CSF study showing lymphocytic pleocytosis, increased protein and decreased glucose levels. CSF opening pressure is also raised in some patients.
2. This case shows that NS can be found in Caucasian population and some rare cases may be resistant to steroids in which case immunomodulators may need to be initiated.