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Sudden-Onset Bilateral Mydriasis in a Young Girl

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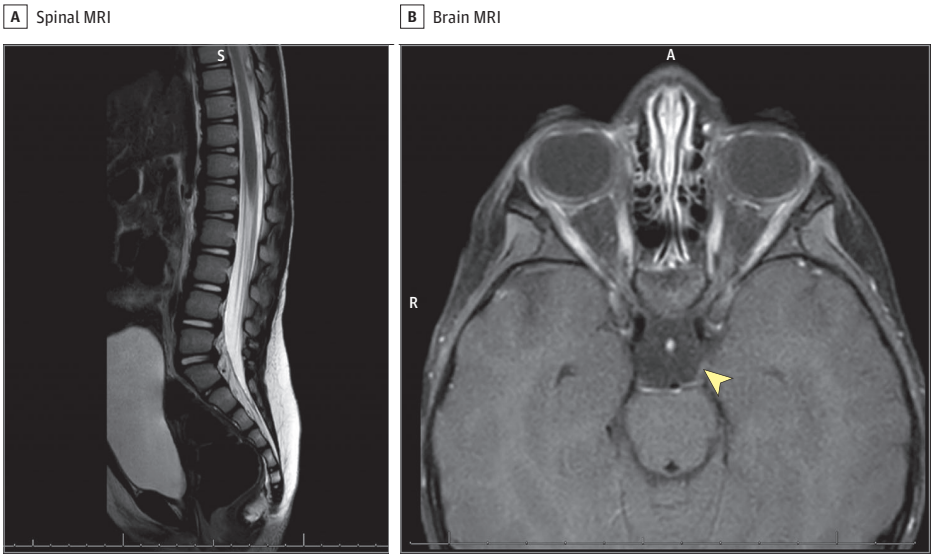

Figure. Magnetic resonance imaging of the spine (A) showing diffuse enhancement of the lower thoracic and equina nerve roots and of the brain (B) showing asymmetric enhancement of the left oculomotor nerve (arrowhead).

A 3-year-old girl with no medical history presented to the emergency department with 1 day of abnormal gait and bilateral mydriasis. Two weeks prior, she had upper respiratory symptoms and bacterial conjunctivitis treated with topical ofloxacin. On examination, pupils were fixed and dilated to 8 mm, extraocular movements were intact, and fundus examination was normal. She had brisk reflexes, poor coordination, and a wide-based unsteady gait. Bloodwork revealed leukocytosis and an elevated erythrocyte sedimentation rate; cultures remained without growth. Lumbar puncture with cerebrospinal fluid studies, computed tomography scan, and magnetic resonance imaging of brain and orbits were normal. The patient was discharged 3 days later following improvement in lethargy and gait with a diagnosis of acute cerebellar ataxia.

When she presented to ophthalmology for 1-week follow-up, her visual acuity was central, steady, and maintained in both eyes. She demonstrated sluggish but reactive pupils and new-onset ophthalmoplegia. She was unable to move either eye in any direction, including with doll's head maneuver. She was readmitted to the hospital and demonstrated diminished reflexes bilaterally and unsteady gait. Repeat magnetic resonance imaging demonstrated diffuse enhancement of the lower thoracic and cauda equina nerve roots and enhancement of the left oculomotor nerve (**Figure**).

WHAT WOULD YOU DO NEXT?

- **A.** Repeat blood cultures and start antibiotics
- **B.** Repeat lumbar puncture with cerebrospinal fluid studies
- **C.** Order anti-GQ1b antibody and start intravenous immune globulin
- **D.** Start intravenous corticosteroids

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Diagnosis

Miller Fisher syndrome

What to Do Next

C. Order anti-GQ1b antibody and start intravenous immune globulin

Discussion

Miller Fisher syndrome is a rare, acute, and self-limited disorder that is considered a variant of Guillain-Barré syndrome. While the exact pathophysiology is unknown, it is thought to be an inappropriate autoimmune response to a preceding infection caused by molecular mimicry of viral components to peripheral nerve antigens. It presents with a clinical triad of ophthalmoplegia, ataxia, and areflexia.[1] Bilateral mydriasis is frequently present in typical Miller Fisher syndrome cases, but an initial presentation with bilateral mydriasis as the only symptom has rarely been reported.[2] Other atypical symptoms could include headache, facial palsy, taste impairment, tachycardia, and hypertension.[3,4]

Miller Fisher syndrome is typically associated with infectious etiologies. The most common bacterial pathogen is *Campylobacter jejuni*, followed by *Haemophilus influenzae*.[5] Viral cases have been reported with Epstein-Barr virus, influenza virus, HIV, and varicella-zoster virus.[5] Autoimmune and neoplastic etiologies have rarely been reported,[1,6] as well as cases following vaccinations.[7]

Imaging is important to rule out other possible diagnoses, such as brainstem abnormalities or compressive lesions.[2] Magnetic resonance imaging results of the brain are usually normal, but there can be enhancement of the cranial or spinal nerve roots.[8] Workup should include lumbar puncture with cerebrospinal fluid analysis to assure the absence of an infectious etiology for symptoms. Cerebrospinal fluid analysis can be normal or can indicate high protein with low white blood cell counts (albuminocytologic dissociation).[4] In this case, the patient had previously had an extensive workup, including blood cultures (choice A) and lumbar puncture with cerebrospinal fluid studies (choice B), which were unrevealing. Given there were no additional symptoms suggesting an infectious etiology, these tests were of low utility.

The most likely diagnosis at this time was Miller Fisher syndrome, the most appropriate next step would be to order the associated antibody and start intravenous immune globulin (choice C). The anti-GQ1b antibody is detected in the serum and has an 80% to 90% sensitivity for this condition.[4,8] In practice, it may take several days to weeks following serum collection for the laboratory to process, so if the patient is clinically presenting with this syndrome, then intravenous immune globulin should be started prior to the official laboratory result. This laboratory test is not necessary for diagnosing the disease but can be helpful in narrowing a diagnosis. Generally, this condition is self-limited with a good prognosis, and recurrence is rare. Patients are typically treated with intravenous immune globulin or, less preferably, plasmapheresis. There is no proven improvement in overall prognosis, but it does shorten the time to recovery compared to supportive care alone.[9] Starting steroids (choice D) is not an appropriate option as steroids are no longer recommended, given they do not hasten recovery or prove effective for this condition.[10]

Patient Outcome

The patient was treated with 2 doses of intravenous immune globulin prior to discharge. Anti-GQ1b antibodies were positive with a 1:400 titer. She presented to the ophthalmology clinic 3 months later. At that time, extraocular movements were full without strabismus. Pupils remained dilated at 6.5 mm in the dark but were mildly reactive to 5.5 mm in the light. Visual acuity was 20/25 and 20/20 in the right and left eyes, respectively. Neurologically, the patient was nearly at her baseline with normal reflexes and gait.

ARTICLE INFORMATION

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