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UNILATERAL DIAPHRAGMATIC PALSY MIMICKING A PULMONARY SPACE OCCUPYING LESION IN A CASE OF NEUROMYELITIS OPTICA

Neurology

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ABSTRACT

The article presents a 15year old young female known case of Neuromyelitis Optica Spectrum Disorders who was admitted with complaints of quadriparesis and respiratory failure. Although she improved with steroids but her dyspnea persisted. And on evaluation she was found to have unilateral diaphragmatic paralysis which was confirmed on fluoroscopy.

KEYWORDS

Neuromyelitis Optica spectrum disorders, respiratory failure, diaphragmatic palsy

Neuromyelitis Optica spectrum disorders (NMOSD) previously known as Devic's disease or Neuromyelitis optica (NMO) is an aggressive demyelinating autoimmune disease. It is distinct from Multiple Sclerosis. It is characterised by presence of autoantibodies to aquaporin-4 and targets optic nerves and spinal cord. Respiratory failure is a feared complication of NMOSD due to neurogenic respiratory failure following involvement of medulla or due to pneumonia, atelectasis and pulmonary thromboembolism. Here we a report a case of NMOSD with respiratory failure due to diaphragmatic palsy following cervical myelitis.

15 year old female was admitted with complaints of sudden onset progressive quadriparesis with bladder and bowel involvement from 3 days. She also had associated respiratory distress progressing into respiratory failure. She was intubated and put on mechanical ventilator. She had bilateral optic neuritis with longitudinally extensive transverse myelitis a year back that improved with IV steroids, after which patient was lost to follow up. She presented a month back with paraplegia. This time she was evaluated further and found to have antibodies against aquaporin-4 in serum. She was diagnosed to have NMOSD and put on tapering dosages of steroid with azathioprine to prevent relapse.

A month later she presented with quadriparesis, bowel and bladder incontinence with respiratory failure. Her chest radiograph (figure 1) revealed homogenous opacity at right mid and lower zone. Routine blood investigations revealed leucocytosis. MRI spine revealed hyperintense intramedullary signals extending from cervico-medullary junction to dorsal cord upto D10 level. Cervical cord was swollen. In contrast to previous scans there was significant increase in cord signals with increase in cord edema (figure 2).

She was given intravenous methyl prednisolone one gram for five days followed by oral taper. Her azathioprine dose was increased. Plasma exchange was planned but couldn't be performed due to monetary issues but with intravenous steroids and conservative management her quadriparesis improved. She was successfully extubated but her respiratory distress persisted. A repeat X ray revealed homogenous opacity in right side involving mid and lower zone. Possibilities of pleural effusion or any space occupying lesion were kept. A CT thorax with contrast was done which didn't reveal any effusion or space occupying lesion.

Fluoroscopy was done, which revealed no movement of right diaphragm with normal movement on left side. This was correlated with MRI Spine, which on assessment revealed asymmetric involvement of cervico-dorsal cord right > left (figure 2) which explains unilateral (right) diaphragmatic palsy.

Although the neurological symptoms had resolved and patient was out of respiratory failure, the diaphragmatic palsy on the right side had failed to resolve in this case.

NMOSD is a distinct clinical entity from MS because of its unique immunologic features. The discovery of a disease-specific serum NMO-immunoglobulin G (IgG) antibody that binds aquaporin-4 (AQP4) has expanded the understanding of clinicians.

Revised consensus criteria were published in 2015. The diagnosis of NMOSD is now based on the presence of core clinical characteristics, AQP4 antibody status, and magnetic resonance imaging (MRI) features [1]. Mortality rates are high in NMOSD. This is most frequently secondary to neurogenic respiratory failure, that occurs due to extension of cervical lesions into the brainstem[2].

Unilateral diaphragmatic paralysis is more common than bilateral involvement. It doesn't lead to much symptoms but is often discovered incidentally on a chest radiograph [3]. Causes of unilateral diaphragmatic paralysis in adults include phrenic nerve injury, viral infections (Herpes zoster, poliomyelitis), cervical spondylosis, cervical compressive tumours, blunt neck trauma or surgery, and brachial plexopathies [3]. Clinician should be aware of following differentials: subpulmonic pleural effusion, diaphragmatic eventration, diaphragmatic hernia, and unilateral subdiaphragmatic processes like abscess, splenomegaly.

In one series, respiratory dysfunction was reported in 22% patients after onset of NMO [4]. In 16% of these patients, respiratory failure was related to a relapse whereas 7% required invasive mechanical ventilation. In another series from literature[5], respiratory failure that caused by acute cervical myelitis occurred 19 times (33%) in 16 relapsing patients and was responsible for death in 15 (93%) of these patients. Only two patients with monophasic NMO (2%) had this complication, with both patients recovering [6]. Bennji et al has reported one such case with left hemiparesis and unilateral diaphragmatic palsy but that was not associated with respiratory failure [7].

This patient presented with multiple relapses and in the present admission she developed respiratory failure. Even after intubation her dyspnoea persisted. Evaluation of which lead us to diagnosis of unilateral diaphragmatic paralysis. It is important to be aware of such a rare presentation as ignorance of which can lead to unnecessary investigations. The diaphragmatic paralysis can be attributed to high asymmetrical cervical cord involvement leading to phrenic nerve damage.

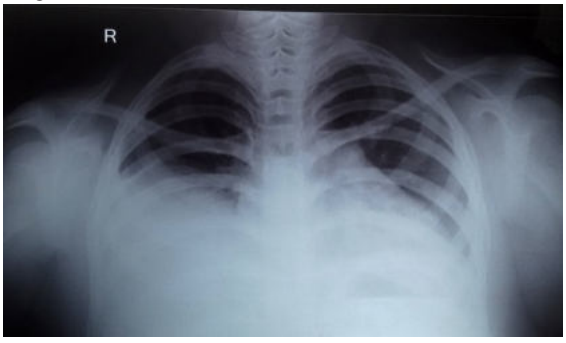

Figure 1 : Antero posterior radiograph showing homogenous opacity in right mid and lower zone

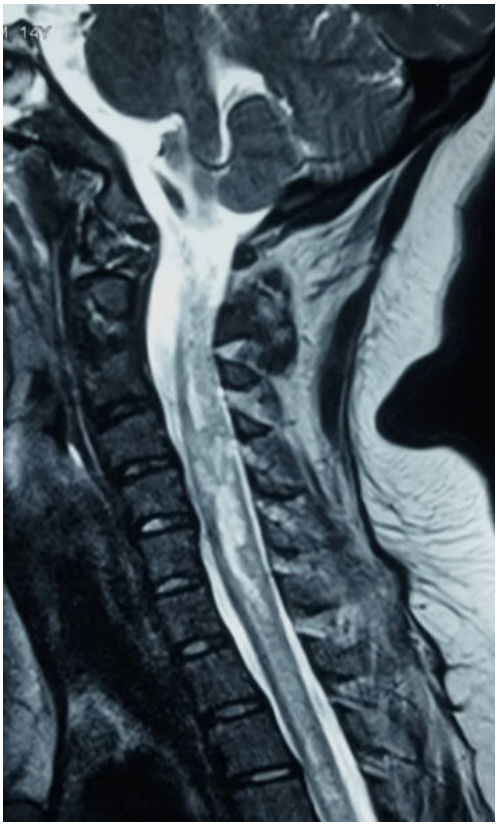

Figure 2 : Sagittal MRI view of the patient showing longitudinally extensive transverse myelitis in cervical cord with extension upto the cervico-medullary junction.

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