Rare Cause of Type 2 Respiratory Failure: Arnold Chiari Malformation

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Sir,

Chronic obstructive airway disease is the foremost cause of type 2 respiratory failure, but there are other causes of this including chest wall disease, impaired neuromuscular transmission and diminished drive. Syringomyelia i.e. Impaired neuromuscular transmission is a rare cause (0.01%) of type 2 respiratory failure. Here i am discussing a case who presented with hypercapnic respiratory failure and on evolution found to be afflicted with extensive syrinx involving cervical spinal cord. A prediabetic 45 yrs old lady presented in our casualty with complaints of cough with expectoration, generalized body weakness, unable to walk since 10 days prior to admission, and swelling over feet since 2 days. On investigations she was found to have type 2 respiratory failure with cor pulmonale. There was no evidence of pulmonary embolism. CBC, liver and kidney function tests were normal. RA factor, and ANA were negative. She was managed with nebulization, BIPAP, antibiotics, iv steroids and other supportive treatment. She responded well and discharged on BIPAP 20/10 and other supportive management. On follow up visit she was doing well with respect to her respiratory effort, but still she was not able to stand and walk on her own and complained of increase weakness in both upper arms. MRI brain and spine was ordered for her neurological weakness. This revealed a syringomyelia with myelomalacia at cervical cord. Syringomyelia and other neuromuscular disorders leads to increased load versus strength in respiration. This type of hypoventilation causes symptoms like excessive daytime somnolence, poor quality sleep, and orthopnea.

In retrospect we should give a thought of rare disorders if diagnosis of COAD was not confirmed as a cause of type 2 respiratory failure. It is a surgically correctable cause.

Neuromuscular disorders usually present as sensory motor weakness. Their presentation as respiratory failure is very rare. There are only few case reports of syringomyelia presenting as respiratory failure. The clinical course of chronic hypoventilation follow a typical sequence: an asymptomatic stage + Pao2 and Pco2 normal--- >nocturnal hypoventilation -->Finally if vital capacity drops further, daytime hypercapnia develops. This increase in Pco2 leads to compensatory increase in plasma bicarbonate and obligatory decrease in Pao2, resulting in hypoxemia. This can further leads to secondary erythropoiesis. The combination of chronic hypoxemia and hypercapnia induces pulmonary vasoconstriction, leading to pulmonary hypertension, right ventricular hypertrophy , and right heart failure.

Take home massage is to keep our vision wide until you get a suitable diagnosis

References