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SUCCESSFUL TREATMENT OF ANTY-SINTHETASE SYNDROME PULMONARY MANIFESTATIONS WITH RITUXIMAB

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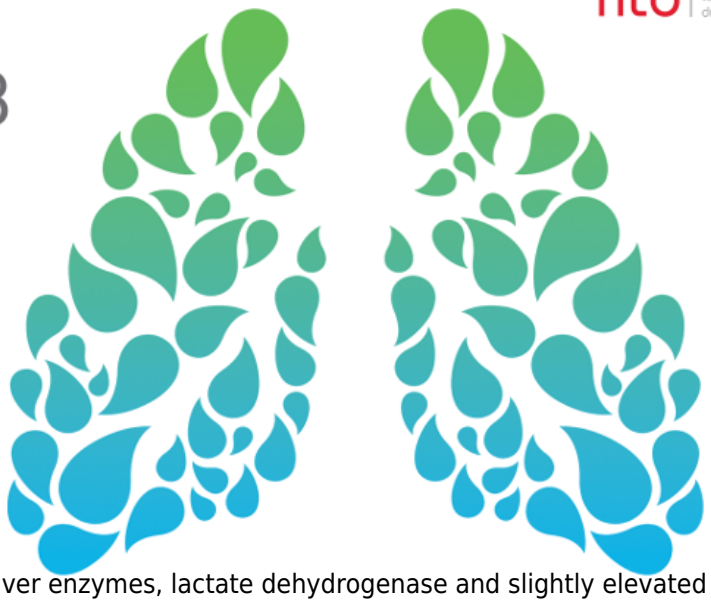
Objective: Introduction: Anti-synthetase syndrome is a rare autoimmune disease characterised by polymyositis/dermatomyositis associated with antisynthetase antibodies, arthritis and interstitial lung disease (ILD). Anti–Jo-1 antibody being the most common. Corticosteroids are first line treatment but usually additional immunosuppresive drug is used.

Case history: 40-yers old, previously healthy female patient with symptoms of joint swallowing, malaise and occasionally dry cough which started 6 months before. She was initially admitted in hospital due to progressive dyspnea and fever but in the next few days her condition deteriorated and she was transferred to ICU were she was intubated and mechanically ventilated. Chest X ray showed extensive bilateral infiltrates. In lab findings there



were elevated C reactive protein, procalcitonin, liver enzymes, lactate dehydrogenase and slightly elevated creatine kinase. On CT pulmonary pulmonary angiography there was no signs of pulmonary embolism but extensive consolidates were described and finding was consistent with adult respiratory distress syndrome. Bronchoscopy was performed and pathology finding was cryptogenic organizing pneumonia. Immunology findings were positive for JO-1, anti Ro52, AGLM and AMHM-M2 antibodies. Pathology finding of muscular biopsy shoved inflammatory myopathy.
she was treated with broad spectrum antibiotics. Initial laboratory findings and her history were suggestive for autoimmune disease and treatment with methylprednisolone 1 g for 3 days and intravenously immunoglobulins was started. There was no significant improvement in her condition so we decided to treat her with rituximab 1000 mg. She received two doses of rituximab 14 day apart. After first dose her condition improved with partial regression of bilateral infiltration. During the stay in ICU she had several complications: superimposed infections and bilateral pneumothorax, pneumomediastinum and bilateral subcutaneous emphysema after percutaneous tracheotomy so she needed chest tub insertion for drainage. She successfully recovered with resolutions of bilateral pulmonary infiltrates and after 26 days she was weaned from mechanical ventilation. Treatment with methotrexate and methylprednisolone was continued. Two months after she was discharged there was no signs of interstitial opacities on her chest X ray, her spirometry finding was normal and diffusion capacity for carbon monoxide was reduced.
Conclusion: Patients with anti-synthetase syndrome have a higher incidence of pulmonary involvement. Clinical suspicion is crucial for early diagnosis and treatment especially when we have patients with acute respiratory distress syndrome. Rituximab is one of the treatment options and in our case it was effective and safe treatment in patient with severe respiratory insufficiency and superimposed infections.