An unusual case presentation of graft versus host myositis secondary to bone marrow transplant in a toddler

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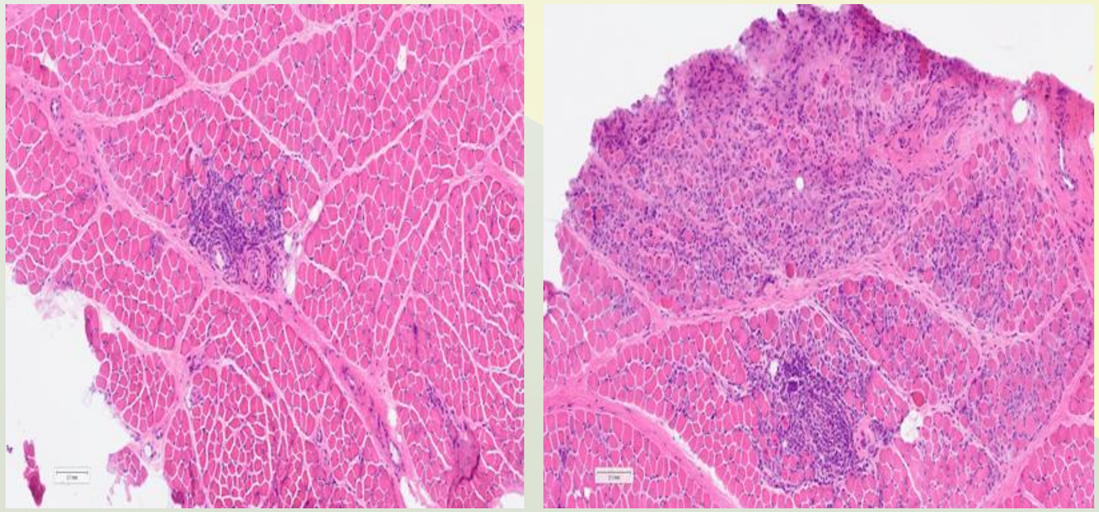
Introduction- An unusual case presentation of graft versus host myositis secondary to bone marrow transplant in a toddler

Material- A 21-month-old toddler presented to the PICU following a second episode of drowsiness and respiratory depression requiring intubation for hypoxia and hypercapnia. Examination revealed hypotonia in all four limbs with pan muscle power of 2/5 and diminished knee and ankle reflexes. After extubation, she was established on overnight NIV due to hypoxia and significant hypercapnia. At 7 months of age, she received a bone marrow transplant for subacute combined immunodeficiency. Three months before this presentation, bulbar weakness was observed with a weak cough, nasal voice, dysphagia, weight loss, and drowsiness. Differentials like Demyelination, Congenital myasthenia syndrome, Central hypoventilation syndrome and congenital myopathy were considered.

Methods- Extensive investigations performed including MRI Brain and spine, R14 genetic testing, Phox gene, infection screen and myasthenia screening were unremarkable. Muscle biopsy showed findings consistent with the diagnosis of graft versus host disease.



Results - She received IVIG, high-dose pulsed methylprednisolone with a taper to maintain oral prednisolone, methotrexate, and fortnightly subcutaneous intravenous immunoglobulin. With the normalisation of Creatinine Kinase from 2402 to 276, she demonstrated significant biochemical and clinical improvement becoming symptom-free.

Discussion- This acute unusual presentation required multiple immunosuppression over several months The differentiation among aforementioned differentials are clinically difficult; the temporal sequence of clinical events and response to immunosuppression can be extremely helpful.

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Disclosures: None