Acute Bilateral Hand Weakness and Swelling in Myelodysplastic Syndrome

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Patient/ Setting

A 78-year old male with myelodysplastic syndrome (MDS) was referred to an outpatient electromyography clinic for further evaluation with complaint of acutely diminished grip strength, hand swelling and diffuse joint pain.

Assessment

The patient was referred to EMG clinic for evaluation of bilateral median and ulnar neuropathies. He had relevant history of MDS with progressive symptoms over the previous year requiring multiple transfusions. On presentation to clinic, he reported impaired mobility and activities of daily living. He complained of acute weakness of bilateral grip strength and associated swelling of hands and wrists as well as diffuse joint pain that began one week before starting medical management with Vidaza for MDS. A detailed history of progression of symptoms was obtained. Manual motor testing demonstrated 4/5 strength in all muscle groups with intact sensation to light touch throughout all limbs equally. He had symmetric swelling and warmth of bilateral wrists, hands and fingers with fullness appreciated in the interphalangeal and metacarpophalangeal joints. Additionally he had diffuse swelling in bilateral ankles and feet. After a thorough history and physical examination were performed, the decision was made not to perform EMG and Rheumatology referral was made for further evaluation of possible inflammatory arthritis. He was subsequently started on steroids with improvement in symptoms.

Conclusion

EMG is frequently described as an extension of the physical exam. A detailed and thorough history and physical examination can assist practicing physiatrists appropriately utilize EMG, especially in patients with uncommon clinical presentations of rare disorders.

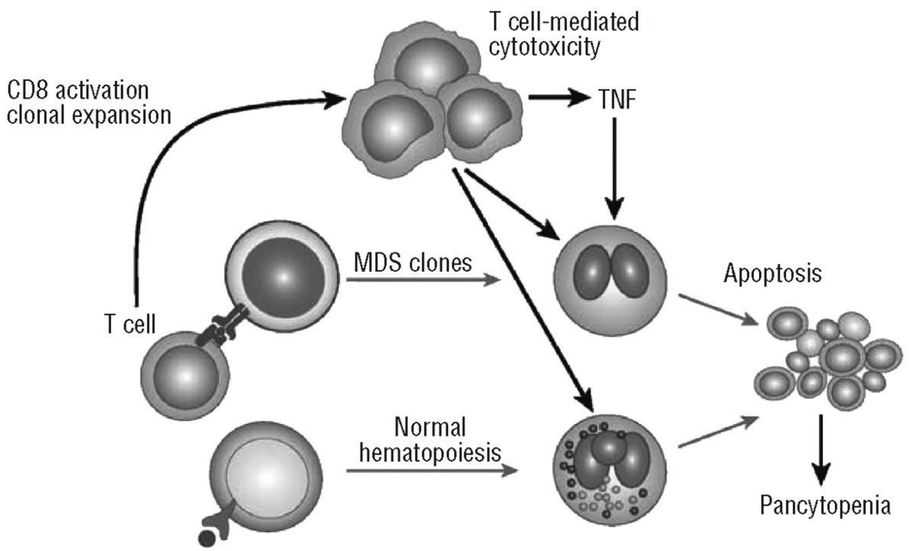

Figure 1: Marrow failure in some MDS is associated with autoimmunity, T-cell mediated myelosuppression and cytokine-induced cytopenias. Myelodysplastic syndromes clones express a neoantigen or overexpress an antigen. This triggers expansion of T cell clones cytotoxic for the myelodysplastic syndromes cell. Activated T cells secrete cytokines, TNFα and IFNγ, which promote apoptosis of normal progenitor cells suppressing hematopoiesis..

Discussion

Autoimmune conditions are uncommon complications of MDS and may affect up to 25% of patients with MDS. The most common autoimmune conditions in patients with MDS are chronic rheumatic heart disease, rheumatoid arthritis, pernicious anemia, psoriasis and polymyalgia rheumatic. In general, autoimmune conditions in MDS do not necessarily lead to increased mortality; however, they may be associated with adverse outcomes and have a significant impact on quality of life. Inflammatory arthritis in MDS is rare, has variable presentation, and may be seronegative without joint destruction. Electrodiagnostic studies are not commonly utilized in patients with MDS, though EMG may be useful in diagnosing concomitant disease states. In the setting of MDS, steroids are often the treatment of choice for autoimmune or inflammatory manifestations as disease-modifying agents may be associated with cytopenias.

References

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