INTRODUCTION: THYMOMAS AND MCD

- Thymomas are uncommon mediastinal tumors arising from the thymus, representing only 0.2-1.5% of all malignancies
- Roughly 40% of thymomas are associated with paraneoplastic syndromes, 5.7% of which are secondary to nephritic syndrome
- Of these cases, the most common renal pathology is Minimal Change Disease (MCD) at 47%
- Approximately 49 cases of MCD associated with thymomas are cited in the literature.
- Due to their rarity, determination of a standard treatment protocols is still under investigation

First-Line Therapy: Corticosteroids
Second-Line Therapy: Limited to Case Reports

MINIMAL CHANGE DISEASE: WORKUP AND MANAGEMENT

- Suspect MCD in any adult patient presenting with acute-onset signs and symptoms of nephrotic syndrome
- Nephrotic Syndrome: Edema, Substantial Proteinuria >3.5g/24hr, Hypoalbuninemia <30g/L
- Microscopic hematuria is common
- Recall: Hypercoagulability, Hyperlipidemia, Increased risk of Infection
- Biopsy: diffuse podocyte effacement
- Glucocorticoid Therapy leads to complete remission in 80-95% of adults with MCD
- Glucocorticoid-sparing regimens are available

CASE DESCRIPTION

- 37-year-old female with PMHx significant for metastatic thymoma (Stage IIB, 2012), status-post resection, with subsequent recurrence (2016), and paraneoplastic myasthenia gravis presented with periorbital, abdominal and lower extremity swelling
- Workup revealed nephrotic-range proteinuria (25g in 24hrs) and urine protein/creatinine >9.4
- Renal biopsy showed MCD
- Treatment with steroids was initiated, but her hospital course became complicated with concerns for myasthenic crisis and shock
- She underwent two rounds of IVIG in conjunction with corticosteroids, but her renal function continued to deteriorate. Pro/Cre Ratio worsened to >12
- Other complications: transudative pleural effusions, blood loss anemia secondary to hematologic, abdominal cellulitis, neutropenia
- She was offered various chemotherapeutic interventions, most of which she declined.
- She was ultimately started on Rituximab with resulting stabilization of her renal disease, and Lanreotide for treatment of her recurrent thymoma

DISCUSSION

- MCD associated with thymoma is rare, with speculation limited to case reports
- The cause of MCD in patients with thymic disease is unclear, but evidence suggests dysregulation of the immune system may play a role.
- Steroid treatment is generally first-line, but other interventions such as chemotherapy and immunomodulation may be utilized in steroid-resistant and recurrent disease
- Responses to such immunosuppressive agents such as Rituximab, in combination with thymoma-specific management appears favorable
- Treatment requires individualized planning, discussions with patients regarding the benefits and risks of these medications, and considerations for a patient’s other comorbidities.

REFERENCES