Thymoma and autoimmunity: a case of multiple sequelae

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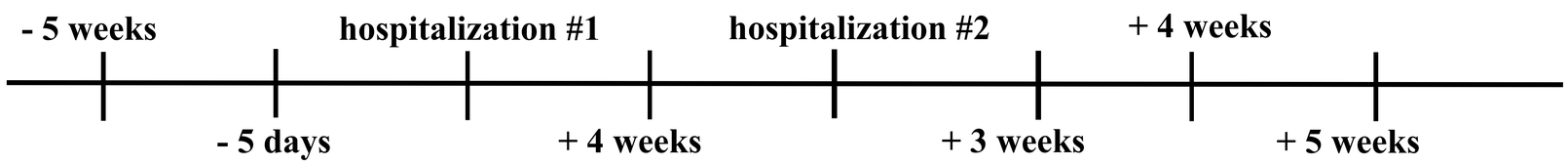
INTRODUCTION

- Thymoma, although rare, is the most common mediastinal mass in adults.
- Most thymoma present with autoimmune diseases (AD).
- Myasthenia gravis is classically associated with thymoma, but many other AD associations have been described.

HISTORY

69-year-old male with history of atrial fibrillation and hypertension who was found to have a mediastinal incidentaloma and one month later presented with petechial rash and epistaxis.

CLINICAL TIMELINE



- 5 weeks from hospitalization #1: mediastinal incidentaloma discovered on CT chest (Figure 1)
- 5 days from hospitalization #1: onset of petechial rash and epistaxis
hospitalization #1: pancytopenia, bone marrow biopsy diagnosis of aplasia (Figure 2), thymoma diagnosed, transfusions
+ 4 weeks from hospitalization #1: onset of proximal muscle weakness, fasciculations, myokymia; admitted 2 days later
hospitalization #2: diagnosis of neuromyotonia, thymectomy, persistent transfusion-dependence
+ 3 weeks from hospitalization #2: start of cyclosporine and equine antithymocyte globulin
+ 4 weeks from hospitalization #2: cyclosporine discontinued due to adverse reaction, tacrolimus started
+5 weeks from hospitalization #2: resolution of neuromyotonia, persistent transfusion-dependence

DISCUSSION

Pathophysiology

- Auto-reactive T cells originating in the thymus are thought to be the cause of thymoma-associated autoimmune disease (AD), although the precise mechanism is unknown.
- Known AD associations include myasthenia gravis, systemic lupus erythematosus, neuromyotonia (Isaac's syndrome), autoimmune cytopenias, limbic encephalitis, autoimmune hepatitis, autoimmune thyroid disease, and cutaneous autoimmune disorders including pemphigus and lichen planus.

Management

- Thymectomy may or may not resolve thymoma associated AD, and some cases of AD may present *after* thymectomy.
- When surgical management of AD fails, immunosuppression should be pursued.

REFERENCES

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VISUAL DATA

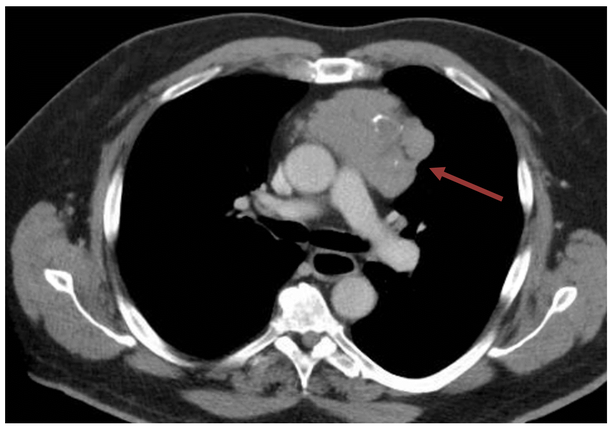

Figure 1: Thymoma (red arrow)

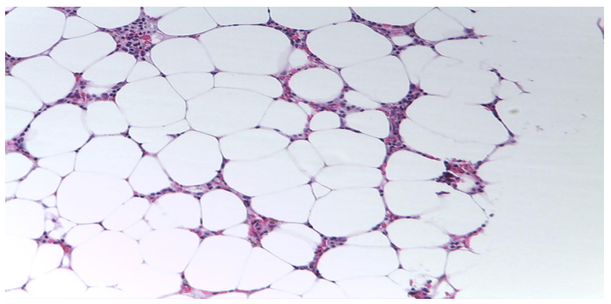

Figure 2: Severe bone marrow aplasia