How Rare Can It Get? Unveiling the Dual Diagnosis of Sarcoidosis and IgG4-RD

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INTRODUCTION:

Sarcoid and IgG4-RD are rare conditions. They are characterized by heterogeneous organ involvement but differ significantly in their pathophysiology. This case illustrates the uniqueness of the immune system as two vastly different conditions may have some similarities in presentation.

CASE PRESENTATION:

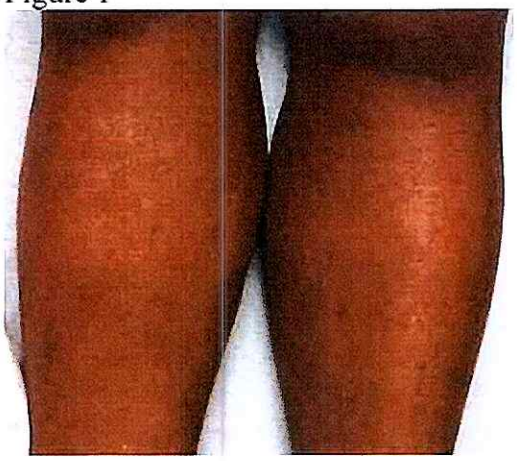
62-year-old female with a history of sarcoid diagnosed in 1998 after presenting with erythematous rash and shortness of breath. Mediastinal lymph node biopsy showed non caseating granulomas. She was treated with high dose prednisone which improved her clinical picture.

She presented recently with facial swelling, lower extremity pruritic rash (figure 1), 40 pound weight loss, decreased appetite, nausea and dry eyes.

Table 1

Laboratory test	Before Treatment	After Treatment
Serum IGG4	1249 mg/dL	76 mg/dL
Creatinine	3.2 mg/dL	0.9 mg/dL
Urine Protein	50 mg/dL	Negative
Urine Protein/Creatinine	3.12 mg/mg	0.06 mg/mg
AST	180 IU/L	22 IU/L
ALT	83 IU/L	8 IU/L
Alkaline Phosphatase	763 IU/L	56 IU/L
Sedimentation Rate	130 mm/Hr	17 mm/Hr
C-Reactive Protein	5.8 mg/dL	0.1 mg/dL

Figure 1



Chest x-ray unremarkable. CT neck/soft tissue showed bilateral lacrimal gland enlargement. CT abdomen and pelvis showed bilateral kidney enlargement with diffuse infiltrate/nodular hypodense pattern, along with enlarged abdominal lymph nodes. Dermatology was concerned about vasculitic process given her rash. Kidney function continued to deteriorate and she was placed on pulse steroids followed by steroid taper. Kidney biopsy showed chronic sclerosing tubulointerstitial fibrosis, storiform fibrosis, at least 50% of IgG-positive plasma cells to be IgG4 positive with more than 12 IgG4 positive cells/hpf. She was diagnosed with IgG4-RD and started on Rituximab. Constitutional symptoms, renal function, liver tests, serum IgG4 and rash improved greatly.

DISCUSSION:

The challenge in distinguishing between Sarcoid and IgG4-RD is well-documented. A notable study in 2011 evaluated 44 patients with suspected sarcoidosis and elevated serum IgG4, finding that two were ultimately diagnosed with IgG4-RD. No studies have demonstrated occurrence of both conditions simultaneously. This case highlights the diagnostic pitfalls that can be involved in distinguishing the two systemic multiorgan conditions and the importance of a comprehensive evaluation and close inspection of biopsy specimen to obtain the correct diagnosis.

References:

1. Tsushima K, Yokoyama T, Kawa S, Hamano H, Tanabe T, Koizumi T, Honda T, Kawakami S, Kubo K. Elevated IgG4 levels in patients demonstrating sarcoidosis-like radiologic findings. Medicine (Baltimore). 2011 May;90(3):194-200. doi: 10.1097/MD.0b013e31821ce0c8. PMID: 21512409.