

Case 9

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Clinical Abstract:

At age 41 this previously healthy heterosexual man was found to have enlarged cervical lymph nodes on routine physical examination. Biopsy showed noncaseating granulomas. He received no specific treatment.

A year later he developed night sweats, fatigue, malaise, generalized lymphadenopathy, joint pains and right ankle edema. CXR showed a diffuse interstitial lung process. Biopsy of an inguinal node revealed non-caseating granulomas. Prednisone and Motrin were begun. There was improvement in symptoms and he returned to his job as a sales manager. Over the next 4 years on Motrin, he did well, although the edema gradually increased to involve hands, feet, arms, legs, lower abdomen and genitalia.

4 years later he complained of inability to maintain an erection and loss of libido. He had lost 25 lbs. In November night sweats, chills and joint pains returned with dyspnea on exertion. Laboratory investigations included Hct 34.9, WBC 9.5 (84P/9B/6L), ESR 63, platelets, PT, PTT, Liver function tests WNL; Angiotensin-converting enzyme 82 (nl-44-125); Rheumatoid factor, ANA, PPD negative. Echocardiogram: mitral regurgitation, aortic insufficiency. Right ventricle biopsy: fulminant myocarditis with diffuse lymphocytic infiltrate. Open lung biopsy: non-caseating granulomas.

In December, episodic heart block with syncopal episodes resulted in pacemaker placement. He returned home on a tapering dose of steroids. One month later, protein-losing enteropathy, thought due to intestinal lymphangiectasia, was diagnosed. Small bowel biopsy was negative.

The following June worsened joint pain, difficulty sleeping, increased dyspnea, and general malaise developed, accompanied by severe proximal muscle weakness and mild peripheral neuropathy. Climbing stairs and getting out of a chair became difficult. Weakness was attributed to steroid myopathy but tapering was not followed by improvement. Dyspnea worsened. Pneumonia required intubation and mechanical ventilation. Lung biopsy was consistent with adult respiratory distress syndrome. Cardiac catheterization suggested constrictive pericarditis. He subsequently developed Herpes zoster in the left groin, and pancreatitis. Muscle weakness was so severe that he became nonambulatory. On admission to a rehabilitation facility there was diffuse muscle atrophy. He expired 3 months later.

Autopsy Findings: Pleural and pericardial thickening, prominent and fibrotic pulmonary interstitium; most organs grossly normal; skeletal muscle pale.

Material Submitted: One H&E- and one unstained section of deltoid, psoas or intercostal muscle

Points for Discussion: Diagnosis. ? Similar cases.