

## ANALYZING A CASE WITH AN ARCANE DIAGNOSIS: A Review of Wegener's Granulomatosis

by Nursine S. Jackson, RN, MSN, CS

Certainly, any endeavor into a medical negligence case is an arduous task, but what should an LNC do when she is asked to review a case involving a disease process with which she (and virtually the entire medical community) have little familiarity? This is the sort of challenge that inspires creativity in developing patterns for case analysis that could serve in all future casework. For me, research in a "failure to diagnose Wegener's Granulomatosis" case served as such an impetus.

My ultimate goal was to determine if the case had merit. My immediate goal was to comprehend the disease, its symptoms, diagnostics, and "standards of care", so that I could perform the initial client interview, then analyze the medical records, and speak knowledgably with experts. Ultimately, I would have to distill all of the information and my analysis into a primer for the attorney's understanding.

To start, I performed a quick review of all of the gold standard treatises. First, I utilized the internet, focusing on a search through MDConsult [URL: <http://www.mdconsult.com>], which is a prescription online medical research resource containing 35 full medical texts, MEDLINE and other search engine capabilities. It also provides immediate access to many full text articles from 48 scientific journals and Clinics. After this initial review, I visited Falk Library, the medical library at the University of Pittsburgh, to perform a more thorough review of the scientific literature by perusing the wall of non-circulating medical treatises. Additionally, I called several rheumatologists with expertise in the field, whom I found in my literature review, and briefly discussed the issues of the case—free of charge.

Following this research process, I compiled a concise summary of the disease process with authoritative citations. Into this review of the literature, I inserted explanations of many medical terms in brackets to make easy reading for the attorney. See Figure 1.

Next I developed a checklist of the common symptoms associated with Wegener's Disease, from which I could conduct my initial interview with the representative family members, and then perform a thorough search of the medical records for evidence of the disease. See Figure 2. Incorporating this checklist of symptoms into my initial interview template, I was prepared to confront the task of performing a case review.

Finally, after reviewing all of the records reflecting the baseline status, plus all of the records reflecting treatment for the problems associated with Wegener's Disease, I was able to assist the attorney in determining whether this case had enough merit to justify formally hiring an expert rheumatologist to perform a review. Most significant to my work as an LNC, I had developed a method to analyze all future cases and present them to the attorney in a useful package.

### **Figure 1.**

#### A Brief Review of the Literature

- Wegener's disease is the result of an abnormal immune response, manifested by inflammation and necrosis of blood vessels (called necrotizing vasculitis). This

process characteristically involves the upper and lower respiratory tracts and the kidneys, with additional frequent involvement of the skin, eyes, ears, and the heart.

- It is a rare disease with an unknown etiology, though an infectious origin is suspected because its process can be altered with antibiotic therapy.
- It is more common in males and may occur at any age.
- It often presents with symptoms referable to the upper respiratory tract, (sinusitis, nasal obstruction) or, less commonly, the lower respiratory tract.
- Abnormalities in routine laboratory tests are nonspecific. Diagnosis is made by demonstrating necrotizing granulomatous lesions in the sinuses and upper airways, fibrinoid appearance of lesions in the lungs, and renal problems from glomerulonephritis.

(See: Horwitz, O. *Diseases of the Blood Vessels*. Phila: Lea & Febiger, 1985, p. 260.)

#### Prognosis:

- Without treatment-one year survival is 15%.
- With steroids and immunosuppressive agents-remission is induced in 90%.

#### Treatment:

- Prednisone and cyclophosphamide [Cytosan, an antineoplastic agent] are recommended as initial treatment until symptoms resolve.
- Continuing treatment (usually steroids) for one year after diagnosis; can yield a continuing sustained remission. Relapses (25-30%) can occur and are treated with corticosteroids. Surgical resection of solitary lung nodules is an option.

(See: Stern, J. H. *Internal medicine*. 4<sup>th</sup> ed. Philadelphia: Mosby, 1994, pp. 1707-1709.)

#### **Figure 2.**

##### Common Symptoms Associated with Wegener's Granulomatosis:

1. Upper respiratory symptoms present in 20-40%: chronic ulcerative lesions, which are refractory to treatment, epistaxis [nosebleed], "saddle" nose from destruction of cartilage, and chronic sinusitis.
2. Eye symptoms occur in 40% of patients: Proptosis [abnormal protrusion of the eyeball(s)], visual loss from corneal damage or uveitis.
3. Otologic [ear] symptoms are frequent (20 – 40%) and often first to appear: otitis media, mastoiditis (often with destructive bone lesions and associated cranial nerve abnormalities), external otitis with earlobe perforation, ear pain, and sensorineural deafness.
4. Lung involvement (These symptoms are often absent): cough, dyspnea, chest pain or hemoptysis [coughing up blood from the lungs]-occasionally massive, abnormal chest x-ray showing Wegner's like nodules.
5. Renal [kidney] diseases: may present as a fulminant [sudden and severe] course of renal failure within days of onset of symptoms and is the leading cause of death. Proteinuria may persist for long periods in patients who are otherwise in remission.
6. Skin lesions are observed in approximately 40% of patients: subcutaneous nodules, purpura (palpable), and chronic ulcerations.
7. Nervous system involvement occurs in less than ¼ of patients: both peripheral nerve and cranial nerves are equally affected.
8. Cardiac involvement in less than 10%: acute pericarditis, coronary arteritis, congestive heart failure (rare).

9. Non-deforming arthritis and arthralgia, usually of large joints, occur in 2/3 of patients.

(See: Stern, J. H. *Internal medicine*. 4<sup>th</sup> ed. Philadelphia: Mosby. 1994, pp. 1707-1708.)