Relapsing Polychondritis: More Than a Swollen Ear

1,2,3 Marcela A. Ferrada, MD, 4Ninet Sinaii, PhD, MPH, 2Thomas Christie, CEO, 3Shubhasree Dutta Choudhury, MD, and 3James Katz, MD

1National Institutes of Health Clinical Center. 2Relapsing Polychondritis Awareness and Support Foundation. 3National Institute of Arthritis and Musculoskeletal and Skin Diseases.

Background

- Relapsing polychondritis (RP) is a rare connective tissue disorder characterized by recurrent episodes of chondritis.
- This disease is unpredictable and clinical manifestations can be variable resulting in a delay in diagnosis.
- Patients can have significant involvement of organs other than the nasal bridge and ear, including larynx, tracheobronchial tree, multiple joints, vasculature, heart valves, and kidneys.
- We aimed to evaluate different patterns of the disease by performing an international survey of patients with RP.

Methods

- Data were acquired using a questionnaire based on known clinical symptoms and other possible clinical presentations of relapsing polychondritis.
- We obtained an exemption from OHSRP, since the survey was anonymous and met criteria for exclusion from full IRB review per CFR 46 and NIH policy.
- The survey was administered by the Relapsing Polychondritis Awareness and Support Foundation via emails to patients who had previously agreed to participate in surveys, and posting the link on their website and multiple support groups sites.
- The survey was open to the public on 2/23/2016 and closed on 8/8/2016.

Results

Figure 1. Survey of Patients with Relapsing Polychondritis

- Total number of survey respondents: N=304
- 16 patients excluded as follows:
  - 9 age younger than 18
  - 7 no DOB reported
- 87% female
- 29% non-US country of origin
- 7 no DOB reported

Figure 2. Time of Onset of Symptoms to Diagnosis

- Figure 2. The majority of the patients had symptoms for up to 5 years or more before RP was diagnosed.

Figure 3. Who Made the Diagnosis

- Figure 3. Half of the patients were diagnosed by a non-rheumatologist physicians.

Figure 4. Initial and Overall Symptoms

- Figure 4. Initial symptoms of RP can include voice changes, shortness of breath and costochondritis. It appears that the symptoms can progress.

Figure 5. ER visits due to RP Symptoms Prior to Diagnosis

- Complications of Relapsing Polychondritis:
  - 21% of patients were on disability due to complications of RP
  - 16% of patients had tracheomalacia
  - 13% reported being intubated due to RP

Therapy Prescribed after RP Diagnosis

- The most common medications included prednisone (n=180), methotrexate (n=94), TNF-blockers (n=18), azathoprine (n=42).
- Other medications included hydroxychloroquine, mycophenolate mofetil, dapsone, and NSAIDs.

Conclusions

- In our population of patients who self-identified as having RP, the majority are female and the time to diagnosis is commonly greater than five years.
- Non-rheumatologist physicians commonly encounter patients with RP.
- Voice changes and shortness of breath are common presenting symptoms of RP.
- The limitations of this study include inability to validate self-reported claims of a diagnosis of RP, and selection and recall biases.
- The strength of our study includes the anonymous web-based strategy employed, which enabled us to capture a large and geographically diverse population.