Clinical Presentations of Relapsing Polychondritis: More Than a Swollen Ear

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Background/Purpose: Relapsing polychondritis (RP) is a rare and in some cases fatal autoimmune disease that can affect multiple organs including cartilaginous structures. The disease is unpredictable and clinical manifestations may be variable resulting in a delay to diagnosis. Patients may have involvement of organs other than the nasal bridge and ear, including larynx, tracheobronchial tree, joints, vasculature, heart valves and kidneys. In this report we present the results of the largest survey to date evaluating disease patterns in an international cohort.

Methods: Data was acquired using an internet-based questionnaire aimed at cataloguing a variety of possible clinical presentations of RP. We obtained an Office of Human Subjects Research Protections ‘Exclusion from IRB Review’ certificate. The data was anonymous therefore met criteria for exclusion per the requirements of 45 CFR 46 and NIH policy. The Relapsing Polychondritis Awareness and Support Foundation administered the survey by email solicitation to patients that previously agreed to be contacted and by posting the link online. The survey was open to the public on 2/23/2016 and the data for this analysis was extracted on 4/11/2016.

Results: 193 surveys total were captured and 13 were excluded either for age less than 18 or no age reported. 180 surveys were included in this analysis. The mean age was 49.5 (SD 11.9). 86.5% were female. 91% identified as “white” and 31.5% reported country of origin other than the USA. The approximate mean age at diagnosis was 43.4 years (SD 12.7). A non-rheumatologist physician made the diagnosed in 47.5%. 54% of the patients saw more than 3 physicians prior to establishing a diagnosis and only 15% underwent cartilage biopsy to support the diagnosis. 49% of the patients had symptoms for more than 3 year before diagnosis. 55% of patients went to the emergency room prior to diagnosis because of RP symptoms. Common initial symptoms included dizziness, eye inflammation, constochondritis, and shortness of breath, nose pain, and voice changes. Some patients also reported fatigue, flu-like symptoms, fever and
Difficulty swallowing as initial symptoms. Complications of RP included disability (25%), tracheomalacia (16%) and intubation related to RP (12%).

**Conclusion:** This is the largest RP study to collate patient and disease characteristics in an international self-reported cohort. We found that the majority was female and there was a high incidence of initial symptoms other than ear and nose inflammation. The time to diagnosis was greater than 5 years. We learned that non-rheumatologist physicians commonly encounter patients with RP and sometimes these encounters occur in the emergency room. We also found that RP could have devastating complications including tracheomalacia and disability. Physicians must be alert to underappreciated presenting symptoms such as voice changes and shortness of breath. The limitations of this study include both the inability to validate self-reported claims of a diagnosis of RP and recall bias. The strength of our study includes the anonymous web-based strategy, which enabled us to capture a larger, and less geographically constrained, population.

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