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Sjogren's Syndrome in Pediatrics: Underdiagnosed and Occasionally Life-Threatening

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Sjogren's syndrome is an autoimmune disease that targets the exocrine glands, most commonly involving the lacrimal and salivary glands.

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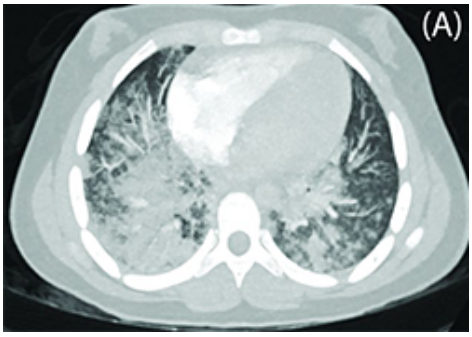

Sjogren's syndrome is an autoimmune disease that targets the exocrine glands, most commonly involving the lacrimal and salivary glands. The classic symptoms are xerophthalmia (dry eyes) and xerostomia (dry mouth), and these symptoms are a key part of the diagnostic criteria; however, affected patients can exhibit a wide array of symptoms.[1] Other common symptoms include chronic fatigue, chronic joint pain without inflammation, and "brain fog", which encompasses poor concentration and memory impairments. On a diagnostic basis, patients with Sjogren's syndrome commonly have positive anti-nuclear antibody along with either anti-Ro antibodies, anti-La antibodies, or both anti-Ro and anti-La. Sjogren's is common in the general population, with an incidence of 0.5-1%, but the median age of presentation is around 45 years and it is not commonly diagnosed in childhood. Given that Sjogren's typically has an insidious onset, dryness may be hard to appreciate or relay in pediatric and adolescent patients, and further that sicca symptoms may not be the initial features of the disease in pediatric patients, such that childhood and adolescent onset Sjogren's syndrome remain underdiagnosed.

In this month's *Pediatrics*, Wang et al (10.1542/peds.2020-042127) report an extreme presentation of childhood Sjogren's in an individual with pulmonary hemorrhage and lymphoid interstitial pneumonia as the presenting features of Sjogren's syndrome in an 11-year-old girl who was admitted to the intensive care unit. In the absence of any signs of infection, the clinical team astutely screened for autoimmune disease and her ANA, anti-Ro, and anti-La antibody tests were positive. A salivary ultrasound showed classic remodeling despite the patient not complaining of dryness, solidifying the diagnosis. Thankfully the individual recovered from the hemorrhage and did well on an immunosuppressive regimen. The case highlights the need for a low index of suspicion in order to make an accurate and timely diagnosis in children with Sjogren's. One of the most specific childhood symptoms of Sjogren's is recurrent parotitis, and this finding or other classic systemic features like lymphadenopathy, recurrent fevers, or diffuse joint pain should trigger autoimmune

testing or referral to a rheumatologist for evaluation. Pediatricians and subspecialists need to have an awareness of the disease in order to arrive at a timely diagnosis and treatment plan for affected children.

Reference:

1. Patel and Shahane, “The epidemiology of Sjogren’s syndrome,” *Clinical Epidemiology*, 2014. PMID: 25114590.

- https://pediatrics.aappublications.org/content/147/5/e2020042507
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