

This podcast can be accessed at www.pedscases.com, Apple Podcasts, Spotify, or your favourite podcasting app.

Post Streptococcal Reactive Arthritis (PSRA)

Developed by Shinia Van and Dr. Lillian Lim for PedsCases.com.
July 7th, 2023

Introduction:

Hi, my name is Shinia Van, and I am a third-year medical student at the University of Alberta. Under the guidance of Dr. Lillian Lim, an Assistant Professor in Pediatric Rheumatology at the Stollery Children's Hospital, this podcast will cover an approach to post-streptococcal reactive arthritis, or PSRA.

The podcast will review the following learning objectives:

1. Present the clinical presentation of PSRA including key questions to ask on history and pertinent physical exam findings.
2. Recognize the differences between PSRA and acute rheumatic fever (ARF).
3. Discuss an approach to investigations and management for PSRA and lastly;
4. Identify possible complications of PSRA.

Clinical Presentation

Let's consider a clinical case: Your preceptor in the pediatric emergency department asks you to see Jane, a 10-year-old female with a 4-day history of right shoulder pain and left ankle pain. Jane describes her right shoulder pain to be a 3 out of 10 severity but has progressively gotten worse to a sharp and persistent 8 out of 10, with swelling that makes it difficult for her to sleep. There has been no obvious trauma or injuries on history. Over the last few days, she noticed a similar pain progression with her left ankle which is making it more difficult for her to walk, with stiffness in the morning. The pain does not affect any other joints. She has tried Tylenol and it has not provided her with any improvement in her symptoms. Jane has been unable to go to school since the pain first started and stays at home with her mother.

Her mother mentions that about one week ago, she had a fever of 38.5C for two days, a sore throat, swollen lymph nodes and mild abdominal pain, but denies having a cough or rhinorrhea. Jane went to a walk-in clinic but cannot recall a throat swab being done. According to her mother, these symptoms all resolved in a few days without any treatment.

Developed by Shinia Van and Dr. Lillian Lim for PedsCases.com.
July 7th, 2023

Jane has not had any unexpected weight loss, night sweats, chills, pain waking her up at night, regression of developmental milestones, or new onset neurological symptoms such as paresthesias. Her review of systems was unremarkable, including no bowel or bladder dysfunction, difficulty breathing, or gastrointestinal symptoms such as diarrhea, or bloody stools. She denied any recent travel, sick contacts, or animal exposures.

On review of her past medical history, Jane is healthy and has no previous surgeries, chronic medical conditions, allergies, or regular medications. Her family history was unremarkable for any autoimmune conditions or recurrent infections.

Physical Examination

Starting first with vital signs, Jane has a blood pressure of 108/70 and a HR of 100 beats/minute. Her respiratory rate is 20 breaths/minute, and her temperature today is 37C. Jane appears well, with no signs of acute distress and is responsive to the questions we have asked her.

Moving on to the specific MSK exam, we assess the joints above and below as well as the unaffected contralateral side. Upon inspection, the right shoulder appears swollen around the acromioclavicular joint, with painful and limited passive range of motion. Jane's left ankle is also noticeably swollen and painful on palpation. She can weight bear but has an antalgic gait. Both joints are neurovascularly intact.

Her remaining physical exam was unremarkable, including no other abnormal joints. She does not have any rashes. Of note, her chest exam is unremarkable with normal S1/S2 sounds, no murmurs or additional sounds radiating towards the axilla or carotid.

Differential Diagnoses:

Our differential diagnosis can include acute and chronic conditions. Autoimmune conditions like juvenile idiopathic arthritis (JIA) are important to consider, though arthritis caused by JIA is usually chronic and lasts at least six weeks. In Jane's case, she does have features of synovial inflammation with joint pain, swelling, and stiffness at two joints, though for only about a week. Given her lack of constitutional or systemic symptoms, chronic conditions like lupus and malignancies are much less likely.

Her joint symptoms started about one week after onset of her infectious symptoms, suggesting that her arthritis is likely post-infectious. Important differential diagnoses to consider are septic arthritis, which needs to be identified urgently, though Jane is well-appearing, afebrile, and able to weight bear.

Other causes include reactive and post-inflammatory processes such as transient synovitis, reactive arthritis, and post-streptococcal reactive arthritis. Jane does not have risk factors to suggest other infection-related etiologies such as tuberculosis or Lyme disease, nor any features to suggest a traumatic injury.

Given the recent streptococcus infection, it is important to distinguish between post-streptococcal reactive arthritis (PSRA) and acute rheumatic fever (ARF). We will discuss some of the key similarities and differences of PSRA and ARF, but please refer to the Acute Rheumatic Fever podcast for more details. Both conditions affect kids of similar age (roughly between ages 5 until late adolescence), though PSRA may also affect young adults in their 20s-30s. Both diseases may show mild to moderate elevation in inflammatory markers.

However, there are several key differences. The timing from onset of infection to joint symptoms is different. PSRA typically happens in 7 to 10 days after the streptococcus infection, whereas ARF oligoarthritis usually presents much later after 10 to 28 days. The arthritis in PSRA also tends to be more persistent and additive by incrementally affecting more joints and is less responsive to initial anti-inflammatory treatment and often requires longer-term therapy, sometimes with additional treatments. In contrast, the arthritis in acute rheumatic fever is migratory, transient, and responds quickly and effectively with initial ASA or NSAID therapy.

The presence or absence of physical exam findings are also important. In Jane's case, she does not have an erythema marginatum rash, or symptoms suggestive of carditis, subcutaneous nodules, or chorea, all of which would be more in keeping with ARF. It is important to keep the Jones criteria in mind to help diagnose or rule out ARF.

Investigations

The investigations that we want to order would focus on bloodwork including a complete blood count and differential, CRP to measure inflammation, a throat swab if not yet done, and anti-streptolysin O test (ASOT). Her white blood cell count was normal, and her CRP was mildly elevated at 20. The throat swab for Group A streptococcus was negative, indicating no current active infection, but the ASOT was moderately elevated at 450, which demonstrates a recent streptococcus infection. Given the short duration of symptoms, the diagnosis of JIA is much less likely, and an ANA was not ordered.

Diagnostic imaging in the context of Jane's presentation does not seem necessary at this point, especially given the low suspicion for an injury such as a fracture. Imaging tests such as x-rays would not provide any significance or changes to our management plan and are often more helpful to rule out other diagnoses.

Management

Given all this information, our preferred diagnosis is PSRA. To manage her condition, the American Heart Association recommends first-line treatment with oral penicillin V for 10 days. For Jane's joint pain due to her arthritis, NSAIDs such as naproxen are helpful, though in some refractory cases, intra-articular or low-dose oral systemic corticosteroids may be considered. The role for disease-modifying anti-rheumatic drugs

(DMARDs) have not been well evaluated but are used sometimes in severe refractory cases.

It is important to discuss and provide education to Jane and her mother about secondary prevention. There is debate about whether prophylactic treatment is effective in preventing carditis, which is typically seen in ARF. A small proportion of patients with PSRA have been reported to develop valvular heart disease. The evidence for the use of secondary prophylaxis is class IIb, level C – meaning that some experts recommend PSRA patients receive secondary prophylaxis for up to one year, after which time it can be discontinued if there is no clinical evidence for carditis. However, the effectiveness of secondary prophylaxis in PSRA is not well established.

In Jane's case, we recommend follow-up after initial treatment before deciding whether prophylaxis is necessary, given the fact that carditis is less common in cases of PSRA and she has normal physical exam findings. Jane and her mother agree with the treatment plan including penicillin antibiotics for streptococcal eradication and NSAIDs for the affected joints. While most cases resolve within a few weeks, a scheduled follow up is important to assess for treatment response, which can take up to two months. Most patients with PSRA only have the one episode and will see resolution in the weeks to come.

This concludes our visit with Jane. Here are some of the key points to take away from this podcast:

1. A thorough history to understand classic presentations of oligoarthritis in a pediatric population are important. Remember to consider post-infectious causes, and that the duration of timing of symptoms can help us differentiate between various etiologies.
2. Recognize some of the key similarities and differences between PSRA and ARF on our differential. You may also refer to the podcast on ARF for more information.
3. Laboratory investigations including a CBCd, CRP, streptococcal throat swabs, and ASOT are important to characterize the infection. Imaging studies, like x rays or ultrasound, are not indicated to diagnose PSRA. If there is concern of chest pain or carditis, an ECG would be helpful for assessing for conditions like acute rheumatic fever.
4. Identify that PSRA can be self-limiting and will usually resolve in a few months. Antibiotic treatment for streptococcus infection is important, and NSAIDs and sometimes intraarticular steroid injections can be used for the arthritis. There is a possibility of recurrence in PSRA, so be mindful when considering if prophylactic therapy is necessary. PSRA has been reported in some cases to lead to carditis, so proper guidance and treatment for streptococcal sore throat is highly encouraged.

Thank you for listening to our PedsCases podcast on PSRA. We hope you took away a thing or two from the podcast, and that you can check out other topics of your interest with our other great selection of PedsCases.

References

Ahmed, S., Padhan, P., Misra, R., & Danda, D. (2021). Update on Post-Streptococcal Reactive Arthritis: Narrative Review of a Forgotten Disease. *Current rheumatology reports*, 23(3), 19. <https://doi.org/10.1007/s11926-021-00982-3>

van der Helm-van Mil A. H. (2010). Acute rheumatic fever and poststreptococcal reactive arthritis reconsidered. *Current opinion in rheumatology*, 22(4), 437–442. <https://doi.org/10.1097/BOR.0b013e328337ba26>

Wilson, W., Taubert, K. A., Gewitz, M., Lockhart, P. B., Baddour, L. M., Levison, M., Bolger, A., Cabell, C. H., Takahashi, M., Baltimore, R. S., Newburger, J. W., Strom, B. L., Tani, L. Y., Gerber, M., Bonow, R. O., Pallasch, T., Shulman, S. T., Rowley, A. H., Burns, J. C., Ferrieri, P., ... Quality of Care and Outcomes Research Interdisciplinary Working Group (2007). Prevention of infective endocarditis: guidelines from the American Heart Association: a guideline from the American Heart Association Rheumatic Fever, Endocarditis, and Kawasaki Disease Committee, Council on Cardiovascular Disease in the Young, and the Council on Clinical Cardiology, Council on Cardiovascular Surgery and Anesthesia, and the Quality of Care and Outcomes Research Interdisciplinary Working Group. *Circulation*, 116(15), 1736–1754. <https://doi.org/10.1161/CIRCULATIONAHA.106.18309>