CASE REPORT

UNUSUAL PRESENTATION OF ACUTE CORONARY SYNDROME IN A 22-YEAR-OLD MALE WITH JUVENILE IDIOPATHIC ARTHRITIS: A CASE REPORT

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This case report documents the clinical journey of a 22-year-old male who was initially diagnosed with Rheumatoid Arthritis (RA) and later, at the age of 11, with Juvenile Idiopathic Arthritis (JIA). The patient manifested atypical symptoms, primarily marked by sudden and intense chest pain accompanied by sweating, leading to a collapse during his emergency room visit. Medical evaluation revealed an extensive myocardial infarction affecting the anterior wall. The patient underwent a successful percutaneous coronary intervention targeting the left anterior descending artery. This case underscores the rare incidence of acute coronary syndrome in young patients with a history of JIA.

Keywords: Acute Coronary Syndrome; Juvenile Idiopathic Arthritis (JIA); Myocardial Infarction; Rheumatoid Arthritis

Citation: Sallar T, Ammar A, Hussain F, Awan R, Muntaha ST, Ahmed M, Sial JA. Unusual Presentation of Acute Coronary Syndrome in a 22-Year-Old Male with Juvenile Idiopathic Arthritis: A Case Report. Pak Heart J. 2024;57(01):67-70. DOI: https://doi.org/10.47144/phj.v57i1.2513

INTRODUCTION

Juvenile Idiopathic Arthritis (JIA) is a chronic inflammatory arthritis predominantly affecting children and adolescents. It represents the most prevalent inflammatory arthritic condition within this demographic. Notably, a significant proportion of individuals, exceeding one-third, continue to experience active symptoms into adulthood, underscoring the long-term implications of JIA.[1]

Rheumatoid Arthritis (RA), on the other hand, is a chronic systemic inflammatory disorder known for its capacity to affect multiple organs, including the heart's pericardium, myocardium, and endocardium. Patients with RA are at a doubled risk of myocardial infarction and stroke, likely due to the interplay between systemic inflammation and traditional cardiovascular disease (CVD) risk factors.[2]

The inflammatory process, characterized by the production of pro-inflammatory cytokines such as Interleukin-1 (IL-1), Tumor Necrosis Factor-alpha (TNF-alpha), and Interleukin-6 (IL-6), is a common pathological mechanism shared by the formation of unstable atherosclerotic plaques and rheumatic synovitis.[3] In this report, we present a rare case of myocardial infarction (MI) in a 22-year-old male patient with a confirmed diagnosis of Juvenile Idiopathic Arthritis.

CASE REPORT

The patient, a 22-year-old male, was diagnosed with Juvenile Idiopathic Arthritis (JIA) at the age of 11. His musculoskeletal examination revealed significant bone deformities in both hands, characterized by ulnar deviation and joint swelling (Figure 1). He had been prescribed Prednisolone 5mg daily and was previously on Methotrexate and Sulfasalazine, which were discontinued three years prior under medical advice.

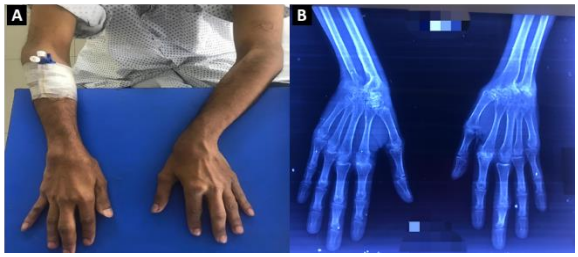

Figure 1: Swelling is evident in multiple proximal interphalangeal joints (PIPs) bilaterally, displaying ulnar deviation and subluxation, with a more pronounced effect on the right (A) and X-ray of the hands reveals periarticular osteopenia and diminished joint space in proximal interphalangeal joints (PIPs), metacarpophalangeal joints (MCPs), as well as radiocarpal joints (B)

He presented to the emergency room (ER) with a 10-hour history of burning chest pain and associated sweating. The pain was gradual in onset but progressively worsened. While in the ER, he

developed ventricular fibrillation, necessitating DC cardioversion. Subsequently, he went into asystole and underwent cardiopulmonary resuscitation (CPR). After approximately six minutes, return of spontaneous circulation (ROSC) was achieved, and he was successfully intubated.

Post-intubation electrocardiogram (ECG) revealed ST elevation in leads I, AVL, and V1-V6 (Figure 2a). Following a thorough evaluation, he was diagnosed with an extensive anterior wall myocardial infarction (MI). He received dual antiplatelet therapy (aspirin and clopidogrel) and heparin in the emergency department and was then transferred to the cardiac catheterization lab. There, primary percutaneous coronary intervention (PCI) of the proximal left anterior descending artery (LAD) was performed using a drug-eluting stent (3.5 x 38 mm). The post-PCI ECG showed resolution of ST elevations (Figure 2b).

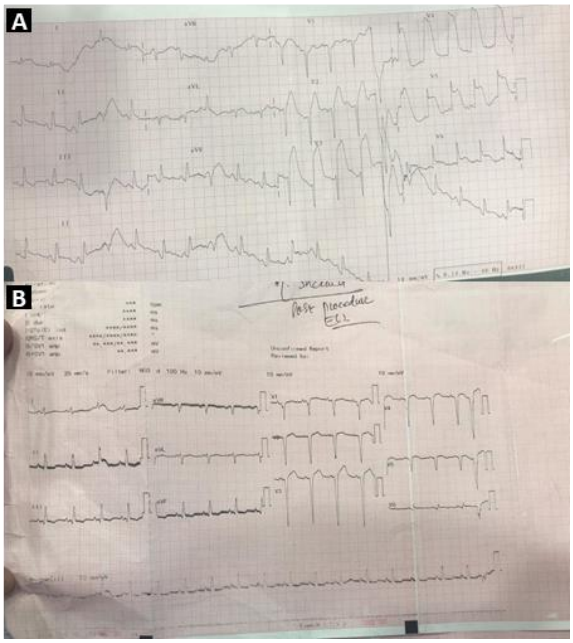

Figure 2: ECG recorded upon arrival in the Emergency Room, illustrating elevation from lead V1 to V6 (A) and post-PCI ECG exhibiting the resolution of ST changes (B)

In the cardiac catheterization lab, coronary angiography was performed. Figure 3A (left) shows the coronary angiography with Right Anterior Oblique (RAO) view, revealing a total occlusion of the LAD with a grade 5 thrombus. Subsequently, primary PCI was performed on the proximal LAD using a drug-eluting stent (3.5 x 38 mm). The post-PCI angiography, as seen in Figure 3B (right), demonstrates the LAD with patent flow following stent placement.

A post-procedure echocardiogram indicated an ejection fraction of 35%, with akinesis of the left ventricular apex, anterior wall, and anterior interventricular septum. The ventricles were of normal size and function. The patient was subsequently transferred to the Cardiac Critical Care Unit, where he developed bradycardia. Telemetry indicated a complete heart block, leading to the planning of a Temporary Pacemaker (TPM) insertion. During this procedure, he experienced ventricular tachycardia, requiring DC cardioversion. A relook angiography confirmed a patent LAD stent, and he was returned to the Cardiac Critical Care Unit.

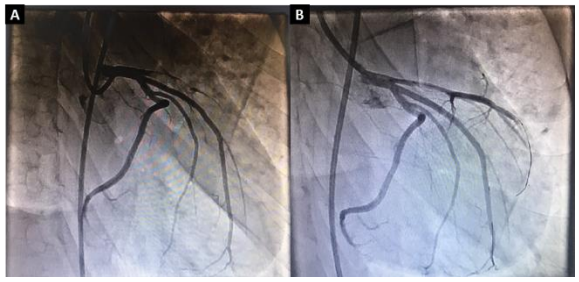

Figure 3: Coronary angiography with Right Anterior Oblique (RAO) view, revealing a total occlusion of the Left Anterior Descending artery (LAD) with a grade 5 thrombus (A) and post-PCI angiography, demonstrating the LAD with patent flow following stent placement (B)

By the next day, he reverted to normal sinus rhythm, and the TPM was removed following an electrophysiology review. His hospital stay was uneventful thereafter, and he was discharged with the following medications: dual antiplatelets, a beta-blocker, an angiotensin-converting enzyme inhibitor, a potassium-sparing diuretic, and a high-intensity statin.

At his one-month follow-up, the patient reported doing well. He was able to exercise without experiencing angina and remained compliant with his medications. His rheumatoid status was stable, evidenced by a negative anti-cyclic citrullinated peptide (anti-CCP) and a positive anti-neutrophil antibodies (ANA) profile. Consent for this case report was obtained from the patient.

DISCUSSION

The established correlation between RA and CVD underscores the significant cardiovascular risks in individuals with chronic inflammatory conditions. However, the long-term cardiovascular risk in patients with JIA is not as clearly defined. It is well known that sustained systemic inflammation, a hallmark of both RA and JIA, can accelerate atherosclerosis, thus elevating the risk of cardiovascular diseases in these populations.

The European League Against Rheumatism (EULAR)

recommends annual cardiovascular risk evaluations for adults with RA. However, similar guidelines for assessing and managing cardiovascular risk in JIA patients are not established. Given the increasing evidence of heightened cardiovascular risk in adult inflammatory arthritis, more research is needed to explore the potential link between JIA and increased cardiovascular risk, especially as the prolonged duration of JIA may place these patients at a higher risk for cardiovascular diseases.

Several factors contribute to the accelerated progression of cardiovascular disease in RA patients, including a higher incidence of conventional cardiovascular risk factors such as diabetes mellitus, hypertension, and smoking. Studies have shown that patients with RA have higher body fat percentages compared to healthy individuals with the same Body Mass Index (BMI), although it's unclear if obesity is more prevalent in adults with inflammatory arthritis. Furthermore, reduced physical activity levels in these patients, alongside the use of corticosteroids and NSAIDs, may exacerbate cardiovascular disease progression. Chronic inflammation in RA patients has been shown to have an additive effect on established risk factors, leading to stiffer artery walls in patients with chronic inflammation and hypertension.

Interestingly, the risk of cardiovascular disease remains elevated in RA patients even after correcting traditional contributory factors. Cardiovascular morbidity and mortality in RA are significantly associated with increased inflammatory markers and disease activity. Elevated C-reactive protein (CRP) levels, for instance, have been linked to higher cardiovascular-related mortality and poorer outcomes in patients, even those without RA, emphasizing the direct role of inflammation in the etiology of coronary artery disease.

The risk of cardiovascular disease in adult arthritis raises the question of a similar increased risk in JIA. Although the pathogenesis of JIA is not completely understood, there is considerable overlap in the pathophysiology between adult forms of arthritis and JIA. Common predisposing factors, such as genetic and environmental influences, are observed in various adult rheumatic diseases and JIA. For instance, Rheumatic Factor (RF) positive JIA shares similarities with adult RA, while systemic-onset JIA parallels adult Still's disease.

Patients with JIA might be at an elevated risk for cardiovascular disease since inflammation is integral to the pathophysiology of both conditions. Elevated CRP levels in young and otherwise healthy individuals have been associated with increased surrogate markers of cardiovascular disease, like carotid intima-media thickness (cIMT) and brachial artery flow-mediated dilatation (FMD).[4] Inflammation may exacerbate atherosclerosis through the combined effect of chronic inflammatory states and traditional risk factors.[5]

RA-associated inflammation might accelerate atherosclerosis through endothelial dysfunction. The production of reactive oxygen species triggered by pro-inflammatory cytokines such as Interleukin-1 (IL-1), Interleukin-6 (IL-6), and Tumor Necrosis Factor (TNF) plays a significant role in this process.[6]

Inflammation is also central to the pathogenesis of JIA, with systemic and polyarticular JIA subtypes showing more pronounced systemic inflammation. The synovium of individuals with JIA contains various inflammatory cells, including T cells, B cells, macrophages, dendritic cells, and plasma cells.[7] CD4+ T helper cells may facilitate these inflammatory reactions. Additionally, neovascularization in synovial tissue, primarily driven by Vascular Endothelial Growth Factor (VEGF) and osteopontin, is noteworthy. Importantly, patients with ongoing JIA have shown elevated levels of pro-inflammatory adhesion molecules, which are associated with endothelial activation, cardiovascular risk, and vascular dysfunction.[8]

The suppression of TNF in clinical treatments aligns with the role of pro-inflammatory cytokines in JIA's progression. Moreover, the inhibition of IL-6 and IL-1 has been effective in alleviating symptoms in the systemic subtype of JIA. The epidemiology of JIA-associated cardiovascular risk necessitates further exploration. Current data suggests that the prevalence of cardiovascular complications in JIA patients may be underrecognized. Longitudinal studies are required to better understand the incidence and progression of cardiovascular diseases in this population, informing future guidelines for cardiovascular risk assessment and management in JIA patients.

CONCLUSION

In conclusion, it is imperative to maintain a vigilant approach in monitoring patients with juvenile arthritis for subtle signs of cardiovascular risk, employing a comprehensive assessment that includes inflammatory markers. Given the current gaps in knowledge, there is a critical need for extensive and detailed data collection, coupled with thorough studies focusing on cardiovascular outcomes in individuals affected by juvenile arthritis. This proactive approach will not only enhance our understanding of the intricate relationship between juvenile arthritis and cardiovascular health but also pave the way for more

effective preventive and management strategies tailored to this specific patient population.

ACKNOWLEDGMENTS

We would like to acknowledge the assistance of the AI language model developed by Open AI, for its support in improving the grammar, coherence, and overall writing quality of this manuscript

AUTHORS' CONTRIBUTION

TS and AA: Concept and design, data acquisition, interpretation, drafting, final approval, and agree to be accountable for all aspects of the work. TS, AA, FH, RA, STM, MA, and JAS: Data acquisition, interpretation, drafting, final approval and agree to be accountable for all aspects of the work.

Disclaimer: None.

Conflict of interest: Authors declared no conflict of interest.

Source of funding: None.

Double blinded peer review history:
Submission complete: February 28, 2023
Review began: March 01, 2023
Revision received: January 08, 2024
Revision accepted: February 22, 2024

REFERENCES

1. Beukelman T, Patkar NM, Saag KG, Tolleson-Rinehart S, Cron RQ, DeWitt EM, et al. 2011 American College of Rheumatology recommendations for the treatment of juvenile idiopathic arthritis: initiation and safety monitoring of therapeutic agents for the treatment of arthritis and systemic features. Arthritis Care Res. 2011;63(4):465-82.
2. Turiel M, Sitia S, Atzeni F, Tomasoni L, Gianturco L, Giuffrida M, et al. The heart in rheumatoid arthritis. Autoimmun Rev. 2010;9(6):414-8.
3. Stenvinkel P, Ketteler M, Johnson RJ, Lindholm B, Pecoits-Filho R, Riella M, et al. IL-10, IL-6, and TNF-?: central factors in the altered cytokine network of uremia-the good, the bad, and the ugly. Kidney Int. 2005;67(4):1216-33.
4. Aviña-Zubieta JA, Choi HK, Sadatsafavi M, Etminan M, Esdaile JM, Lacaille D. Risk of cardiovascular mortality in patients with rheumatoid arthritis: a meta-analysis of observational studies. Arthritis Rheum. 2008;59(12):1690-7.
5. Lin CC, Yang CC, Wang CY, Tseng HC, Pan CS, Hsiao LD, et al. NADPH oxidase/ROS-dependent VCAM-1 induction on TNF-?-challenged human cardiac fibroblasts enhances monocyte adhesion. Front Pharmacol. 2016;6:310.
6. Goodson NJ, Symmons DP, Scott DG, Bunn D, Lunt M, Silman AJ. Baseline levels of C-reactive protein and prediction of death from cardiovascular disease in patients with inflammatory polyarthritis: a ten-year follow-up study of a primary care-based inception cohort. Arthritis Rheum. 2005;52(8):2293-9.
7. Lin YT, Wang CT, Gershwin ME, Chiang BL. The pathogenesis of oligoarticular/polyarticular vs systemic juvenile idiopathic arthritis. Autoimmunity Reviews. 2011;10(8):482-9.
8. Van Raemdonck K, Umar S, Szekanecz Z, Zomorrodi RK, Shahrara S. Impact of obesity on autoimmune arthritis and its cardiovascular complications. Autoimmunity Reviews. 2018;17(8):821-35.

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