SYSTEMIC JUVENILE IDIOPATHIC ARTHRITIS IN CHILDREN COMPLICATED BY AMYLOIDOSIS WITH SECONDARY NEPHROTIC SYNDROME

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ABSTRACT

A 7-year-old girl diagnosed with a case of juvenile idiopathic arthritis since the age of 1.5 years, generalized onset with systemic presentation is complicated by secondary nephrotic syndrome. Renal biopsy showing amyloidosis. The patient has a severe disease course complicated by severe flare-ups of disease symptoms and body swelling since the onset of the disease. The patient got NSAIDs, methotrexate, steroids, and TNF inhibitors with no or partial response. After diagnosis with secondary nephrotic syndrome due to secondary amyloidosis, the patient is started on intravenous tocilizumab. There is improvement in patient symptoms.

KEYWORDS: *Juvenile Idiopathic Arthritis, Secondary Nephrotic Syndrome, Amyloidosis, Tocilizumab*

INTRODUCTION

Juvenile idiopathic arthritis (JIA) is one of the most common diseases affecting the quality of life in children.[1] There is an unfavorable socio-economic impact due to the progressive course of the disease, not only in childhood but also in adulthood.[2] This disease is characterized by the progressive development of destructive and erosive arthritis in children up to 16 years of age, with the manifestation of joint swelling, deformities, and contracture. With prolonged involvement, there is muscle atrophy. There are also some patients with a variety of extra-articular lesions.[3] Eyes are commonly involved along with the heart and lungs.[4] Though kidneys are also involved, it is not extensively studied as there are changes in the kidney regardless of the duration of JIA, which determine the prognosis for these patients.[5] Renal pathology includes tubulointerstitial nephritis, glomerulonephritis, and secondary amyloidosis.[6] Secondary amyloidosis (AA-amyloidosis) involving kidneys is the most unfavorable lesion.[7] In the available literature, there is a description of 24 clinical cases of AA renal amyloidosis in children with JIA.[8] We are presenting the case of a 7-year-old girl diagnosed and managed as JIA having secondary renal amyloidosis after obtaining proper consent from her parents. Steroid-sensitive nephrotic syndrome can precede the development of juvenile idiopathic arthritis in children, with nephrosis often occuring before arthritis. Renal amyloidosis is an uncommon complication of juvenile idiopathic arthritis, and chlorambucil treatment improved the patient's condition and reduce proteinuria.

CASE REPORT

A 7-year-old girl, a known case of juvenile idiopathic arthritis since the age of 1.5 years presented with a history of body swelling for the last week. The patient is continuously taking treatment for JIA since the age of 1.5 years. Due to a progressive increase in patient symptoms and restriction of movements, the patient was being treated with steroids along with methotrexate. At the age of 6 years there was a flare up in symptoms and initially patient was managed on line of macrocyte activation syndrome (MAS) but work up not suggestive e of MAS. At presentation patient was started on etanercept for 3 months but there was no relief in symptoms and then patient was started on sub cutaneous tocilizumab. Despite getting s/c tocilizumab, there was no relief in patient symptoms. During the course patient got edema and workup showing the nephrotic syndrome. Urine detail report showing +++ protein, 24 hour urine protein 2730 mg, arum albumin 1.7 gm/dl, urea 18 mg/dl, creatinine 0.1 mg/dl, cholesterol 255 mg/dl. Patient was given two shots of infusion albumin for anasarca. Renal biopsy done which showed amyloidosis. Patient is started on intravenous tocilizumab and two shots of tocilizumab given. Patient condition improved both from JIA aspect and nephrotic syndrome symptoms also improved as

shown in figure of hand, foot and patient in figure 1,2 and 3 respectively.

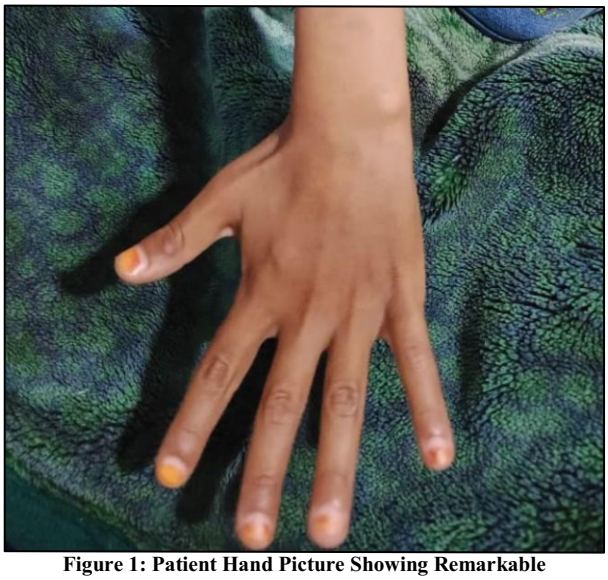

Figure 1: Patient Hand Picture Showing Remarkable Improvement in Swelling

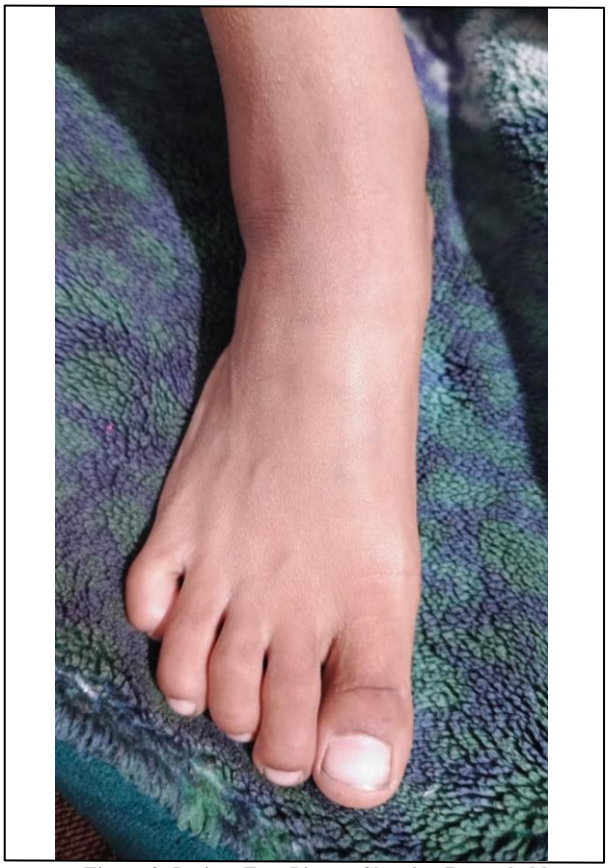

Figure 2: Patient Foot Picture Showing Remarkable Improvement In Edema

Figure 3: Patient Picture Showing Remarkable Improvement

DISCUSSION

Secondary amyloidosis is caused due to acute-phase reactant protein, serum amyloid A (A-SAA) in patients having chronic infection or inflammation or infection as JIA is the one of the common etiology in children.[8] The frequency of AA renal amyloidosis in children with JIA ranges from 0.8% to 2% as reported in literature from one of the neighboring country, but in Pakistan the prevalence and incidence in not known.[9] In literature the chronic use of glucocorticoids has been designated one of the risk factor for the development of AA-amyloidosis.[10] Though Disease-modifying antirheumatic drugs (DMARDs) are the main stay of treatment in children with JIA yet, targeted physiotherapy exercises may reduce pain and improve strength, gait, posture, and functional mobility.[2] The primary treatment of JIA in children is managed with NSAIDs, DMARDs), and Tumor necrosis factor (TNF)-alpha inhibitors as in our patient etanercept was given but there was not good response and then patient was given subcutaneous tocilizumab but still patient was symptomatic. Afterwards patient got body swelling and diagnosis as nephrotic syndrome. In literature, use of tocilizumab in management of JIA related secondary

amyloidosis is considered as effective and safe option.[11] And in our patient we have started patient on intravenous tocilizumab. Patient symptoms have improved and on follow up.

CONCLUSIONS

Though systemic JIA can present with nephrotic syndrome with secondary amyloidosis yet treatment options may include tocilizumab in patients with no response to other drugs.

CONFLICT OF INTEREST: None

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CONTRIBUTORS

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2. ***Bibi Aalia*** *- Drafting Manuscript; Critical Revision*