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Steroid Resistant Nephrotic Syndrome with Underlying Renal Amyloidosis Secondary to Systemic JIA: Rare Case Report

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Abstract **Case Report**

Nephrotic syndrome is mostly idiopathic but secondary causes are also not uncommon. In our case we had a steroid resistance case so on renal biopsy we found that it due to amyloidosis which was also attributed to systemic JIA. It showed response to biological agents.

Keywords: Renal Amyloidosis, systemic JIA, Nephrotic syndrome.

BACKGROUND AND AIM

Amyloidosis is a group of diseases characterized by the extracellular deposition of beta-sheet fibrils. Amyloidosis with renal involvement is a rare cause of nephrotic range proteinuria in children. Most common form of amyloidosis is reactive AA amyloidosis due to chronic inflammatory diseases. Here we present a case of Steroid resistant nephrotic syndrome secondary to renal amyloidosis.

CASE REPORT

12-year adolescent girl presented with complaints of generalised edema, fever, multiple joint pains, and facial rash for the last 1.5 yrs. Investigations revealed nephrotic range proteinuria, hypoalbuminemia, and hypercholesterolemia. She had already received prednisolone for the last 8 week but no improvement was observed, so diagnosis of steroid-resistant nephrotic syndrome was made. Further as a part of diagnostic evaluation, renal biopsy was done which revealed findings suggestive of renal amyloidosis. Workup for primary cause was done, and diagnosis of systemic onset JIA (sJIA) was made on basis of ILAR criteria. Treatment with Tocilizumab was started every 2 weeks, along with a tapering dose of steroids. After three months of follow-up, the child showed remission and was on tapering dose of steroids.

CONCLUSION

Nephrotic syndrome is a common illness presented to paediatricians. SRNS secondary to Renal Amyloidosis is a rare entity in children. Early diagnosis and prompt treatment is helpful for the prevention of serious complications.

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