AN ATYPICAL CASE OF ANTI GBM DISEASE

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Anti GBM disease is a very rare type of small vessel vasculitis. The incidence is around less than 1 per million per year. This disease is also known as “Good Pasteur Disease”. Usually it presents with rapidly progressive glomerulonephritis with or without lung haemorrhage. The pathognomonic hallmark of the disease is strong linear IgG deposition along the GBM along with positive anti GBM antibody. But when the circulating antibody is absent in the blood, with mild renal impairment it is termed as atypical anti GBM disease. Recently we have found a 26 years old gentleman who presented with leg swelling for 1.5 months along with decreased urine output. He was non diabetic, normotensive, there was no history of joint pain, rash, no offending medication intake, or coughing out of blood. His urine R/E report showed Alb++, RBC-plenty. After admission his serum creatinine was increasing rapidly. We have done renal biopsy and started treatment with I/V methylprednisolone followed by oral steroid. He had nephrotic range proteinuria. His auto antibody profile and HBsAg, Anti HCV was negative. We have done renal biopsy and it showed crescentic GN with strong linear deposition of IgG. His anti GBM Ab was negative. Thus we labelled the case as atypical anti GBM disease. Now patient is on RRT and we started therapeutic plasma exchange.

Keywords: Atypical case, Anti GBM disease.

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