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반복혈뇨가 있는 환아에서 IgG 사구체신염을 동반한 선천성 누두부 협착증 1예

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A Case of IgG Glomerulonephritis with Congenital Infundibular Stenosis

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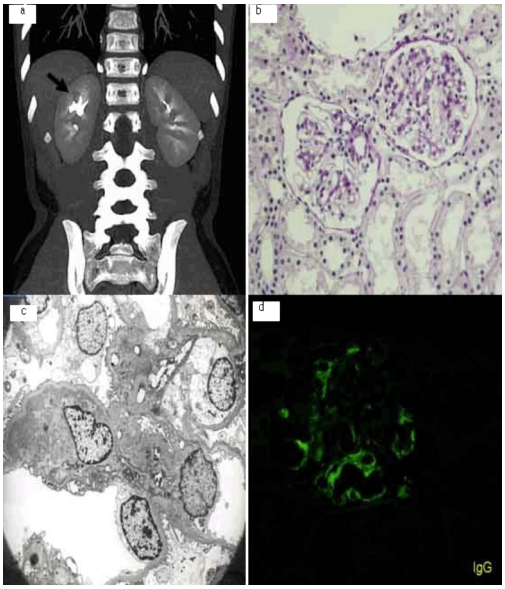
Introduction: IgG glomerulonephritis is a rare form of glomerulonephritis and was recently recognized as a distinct type of glomerulonephritis. Infundibular stenosis is an extremely rare condition characterized by dilation of one or more calyces proximal to a stenotic infundibulum. The clinical courses of IgG glomerulonephritis and infundibular stenosis vary from benign to end-stage renal disease. We report a case of pediatric IgG glomerulonephritis that was proven by renal biopsy in which congenital infundibular stenosis was also present. To our knowledge, this is the first report of a case of this nature.

Case: An 8-year-old boy was admitted because of recurrent gross and microscopic hematuria during the previous 6 weeks. Hematuria was confirmed by urinalysis, and renal imaging and an ultrasound-guided biopsy were performed for diagnostic purposes. Computed tomography showed focal dilatation of the upper polar calyx of the right kidney. Light microscopy revealed a mild increase in the size and cellularity of the mesangial matrix. Electron microscopy showed dense deposits in the mesangium and paramesangial lesions. Effacement of the foot process was significant. Immunofluorescence showed the presence of deposits of IgG (3+), IgM (1+), C3 (1-2+), C4 (1+), and C1q (1+), mainly in the mesangium (Fig. 1). The patient was given supportive treatment without medication and decompression and was discharged without hematuria. He was monitored closely thereafter and his renal function remained normal during the 40-month follow-up period.

Discussion: The patient described in this report demonstrated distinct primary GN with predominant IgG deposits in the absence of any other etiologic factor known to cause secondary IgG GN. He also had a less severe form of congenital infundibular stenosis. To our knowledge, there are no published reports that indicate that the simultaneous existence of both diseases results in ESRD or renal insufficiency. The two findings may have been a coincidence and not pathologic related to one and other. However, such cases should be closely monitored for potential decreases in renal function. Monitoring of renal function should be continued until adulthood even though proof of such effects in cases of coexistent IgG GN and infundibular stenosis is lacking. Although these diseases are very rare, pediatricians, radiologists, and nephrologists should be aware of their existence because they may be accompanied by unfavorable courses and prognoses.



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IgG glomerulonephritis, Infundibular stenosis