Situs inversus: An Uncommon Extrarenal Association of Alport's Syndrome

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Dear Sir,

Alport's syndrome is an inherited progressive kidney disease characterized by he-maturia and eventually renal failure often accompanied by hearing loss and ocular lesions such as lenticonus and retinal flecks. Electron microscopic studies of renal biopsy usually show alterations in glomerular basement membrane [1, 2]. A spectrum of extra-renal abnormalities have been reported in patients with Alport's syndrome such as thrombocytopathia, esophageal leiomyomatosis [3, 4], cryptorchidism and situs inversus. Only 3 cases of Alport's syndrome with situs inversus have been reported in the literature so far [5]. We now report the fourth case of Alport's syndrome with situs inversus.

The case is a 22-year-old man who developed jaundice at the age of 17, and while being studied for liver problems, he was found to have microscopic hematuria. Renal function gradually deteriorated and serum creatinine rose to 18.9mg/dl, necessitating hemodialysis at age 20. On clinical examination and plain radiography there was evidence of situs inversus in the form of dextro-cardia and abdominal viscerae on the opposite site. Ultrasonography of kidneys showed absence of right kidney with increased parenchymal echogenicity. The family history revealed that his father died at age 26 with renal failure and his sister is now undergoing hemodialysis. His paternal aunt has hematuria and proteinuria, and electron microscopic study of her renal biopsy showed thinning or splitting and splintering of the capillary basement membrane.

Ophthalmological examination of the patient and his sister showed decreased visual acuity, myopia and fine punctate lens opacities. His sister also had retinal pigmentary changes as bilateral fine yellow flecks in the midperiphery. Audiological examination of the patient showed unilateral low frequency hearing loss. His sister also showed bilateral trough-shape hearing loss compatible with the cochlear type.

Only 3 cases have been reported in the literature so far describing the coexistence syndrome and situs inversus in the same patient. Regarding our patient as the fourth case, situs inversus can now be considered a true but uncommon association of Alport's syndrome.

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