

일차성항인지질증후군에서 발생한 비허혈성 신증후군

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Nonischemic Nephrotic Syndrome Associated with Primary Antiphospholipid Syndrome

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Introduction : Renal involvement cases of antiphospholipid syndrome (APS) were mainly the result of vascular obstruction of renal vessels. The triad of renal pathologic features were: (1) fibrous intimal hyperplasia, (2) focal cortical atrophy resulting from ischemia, and (3) thrombotic microangiopathy. There was not routinely accepted an immune glomerulonephritis as one of primary antiphospholipid syndrome nephropathy. We present the immune glomerulonephritis of primary antiphospholipid syndrome.

Case : A 11 year old female child was admitted with generalized edema and severe visual disturbances with nearly blindness for 1 month. There was no specific family history. BP was 120/80 mmHg, On the physical examination, there was a eyelid swelling and pitting edema. There's no cranial nerve abnormality. On the fundus examination, both optic disc severe swelling and blunted disc margin with star shaped hard exudate on macula were noted. Serum protein and albumin were 4.4/2.1 g/dL. Serum cholesterol and triglyceride were 342/224 g/dL. CBC was as follows: WBC 8,360, Hb 11.7 g/dL and platelet 130,000/mm³. PTT was 50.6 sec (control 32.3 sec). Brain MRI was normal. A diagnosis of primary APS was made on the basis of antiphospholipid syndrome IgG/IgM (+/-), Cardiolipin IgG/IgM (+/+), ANA (-), anti-DNA Ab (-), anti-sm (-), anti-ENA (-), C3/C4 98.6/19.8 mg/dL. Kidney tissue showed diffuse proliferative glomerulonephritis with subepithelial electron dense deposits. She has been treated with coumadin, steroid and cyclophosphamide, but still remained proteinuria without edema. Her vision was much improved.

Conclusion : We suggest that there should be a need for concern about the possibility of the development of immune glomerulonephritis in primary antiphospholipid syndrome.

Key Words : 일차성항인지질증후군, 신증후군

Primary antiphospholipid syndrome, Nephrotic syndrome