Conflictos de interés

Los autores declaran que no tienen conflictos de interés potenciales relacionados con los contenidos de este artículo.

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Development of Focal Segmental Glomerulosclerosis in a Patient with Polycythemia Vera: can Polycythemia Vera be a cause of Focal Segmental Glomerulosclerosis?

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Dear Editor,

Polycythemia vera (PV) is a myeloproliferative disorder of unknown etiology. This condition is characterized by the abnormal proliferation of erythroid and myeloid series cells in the bone marrow.[1] Focal segmental glomerulosclerosis (FSGS) is a glomerular disease characterized by the presence of nephrotic syndrome, hypertension, and the progressive deterioration of the renal function. Etiology is usually unknown, but it may be seen in secondary conditions.[2-3] PV in association with FSGS is rare.[4] As far as we know, only eight cases have been reported in the literature.[4-10] In the report, we have presented a patient who development of FSGS associated with PV.

A 46-year-old male patient diagnosed with PV six years earlier was referred to the nephrology clinic due to the detection of proteinuria on routine controls. No important features were found on his history except for his use of the azathioprine for a month. Through a 24-hour urine analysis, 4g/day proteinuria was detected in the patient. The patient was admitted to the clinic. In physical examination is normal without arterial blood pressure of 140/90 and the spleen was 5cm palpable. Renal size and parenchyma were normal in abdominal ultrasonography. Laboratory tests results and examinations of glomerular disease have been showed at Table 1. Hence, causes of nephrotic syndrome were excluded. Renal biopsy was performed. In light microscope were shown 29 glomeruli. Global sclerosis and hyalinization were shown in five glomerule. There was intensive segmental sclerosis in more small segments of the other two to three glomeruli. The also remaining some glomeruli have mesengial cell proliferation and expansion. In Bowmans capsule of one to two glomeruli presence of synechiae noted. Tubulointerstitial area has been examined, focal interstitial mononuclear cell infiltration has been observed. In particular areas of inflammation have attenuation of some tubules epithelium and in the presence of eosinophilic material in the lumen characterized by atrophic changes were

observed. In vascular stuctures were normal except for a slight thickening of the wall. Glomerulosclerosis, segmental sclerotic areas and slight thickening of the glomerular basement membrane have been detected through the use of Trichome stain. Furthermore, amiloid staining and immunofluorescence study showed a negative. All above these findings were indicative of FSGS. In arterial blood pressure monitoring, stage 1 hypertension was determined. Perindopril, azathioprine, and ASA were prescribed and the patient was discharged.

FSGS is a clinical and pathological disorder involving primarily the glomerulus.[2-3] Progressive glomerular scarring is the most important feature in this disease. Early in the disease process, glomerulosclerosis is both focal, and segmental in nature. Furhermore, in later stages of the disease diffuse and global glomerulosclerosis develops. The loss of filtration barrier, depletion of podocytes and genetic susceptibility are the culprit factors in pathogenesis of FSGS. The condition can be idiopathic or occur secondary to obesity, intrarenal hemodynamic alterations, conditions with glomerulomegaly, the reduced number of nephron, and renal atheroembolic disease.[2,3] The tendency to thromboses may occur in PV which one of the chronic myeloproliferative disorders.[1] It has been suggested that the increase level of red blood cells, elevation of the platelet count, increase in tissue factor, polymorphonuclear leukocytes, coagulation reactions related to the platelet surface and the presence of microparticles were culprit factors.[11]

In the light of these data, we hypotezised that PV may cause of FSGS via recurrent thrombosis in microvascular level. Furthermore, it is well known that atheroembolic disease is a cause of FSGS. Thus, our case is important for present to develop of FSGS in the patient with PV. In the existing literature, a small number of cases of FSGS that are thought to be due to PV have been reported.[4-10] In addition, 3.6 % (only two PV) incidence of FSGS has been reported in patients with myeloproliferative disease.[10] It has been expressed in these case reports that hyperviscosity from increased hematocrit, hypoperfusion, predisposition to thrombosis related to elevated platelet counts and the continuation of these conditions in recurrent attacks may have a role on the development of FSGS.[9] The emergence of FSGS has been reported average three to seven years after the diagnosis of PV.[4-10] In the case of the subject of our study, considering that FSGS has been diagnosed with PV six years later is consistent with the literature. The PV was thought to be the possible reason for FSGS. Additionally, FSGS may occur by occlusions due to the long term recurrent microvascular thrombosis and this also could disorder to glomerular hemodynamics.

Consequently, the co-existence of PV and FSGS seems to be a cause-effect relationship rather than a random combination. Further studies will be needed to demonstrate for a better understanding of this association.

Table 1. Haematological, chemistrical, serological and urinalysis findings of patient.

Haematology	Blood chemistry and Serology		Urine analysis
WBC (mm³): 11600	ALP: 88 U/L	Na: 142mmol/L	Glucose (-)
RBC (mm³):	AST: 23 U/L	K: 5.06 mmol/L	Protein (++)
Htc (%): 52	ALT: 29 U/L	Uric acid: 4.8 mg/dl	O.B. (-)
Hb (g/dl): 16.3	LDH: 230 U/L	Anti-HCV (-)	Microscopic
PLT: 562000	Total Cholesterole:	HBsAg (-)	Examination:
	107 mg/dl		WBC: 3
	TG: 201 mg/dl	Anti-HIV (-)	RBC: 2
	TP: 6.7 g/dl	ANA (-)	24 hour urine
	Albumin: 4,2 g/dl	C3:1.2 g/L	protein: 4 gr/day
	Glucose: 88 mg/dl	C4:0.3 g/L	
	BUN: 18 mg/dl	RF (-)	
	Crea: 1.2 mg/dl	PTH: 64 pg/ml	

ALP: alkaline phosphatase; BUN: blood urea nitrogen; O.B.: occult blood; TP: total protein.

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Conflict of interest

The authors declare that there is no conflict of interest associated with this manuscript.

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Nefritis tubulointersticial y colangitis esclerosante asociadas a pancreatitis autoinmune

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Sr. Director:

La pancreatitis autoinmune (PA) es una forma de pancreatitis crónica causada por un proceso inflamatorio autoinmune con infiltración linfocítica y fibrosis que conducen a disfunción del órgano[1], relacionada con altos niveles de IgG4 y anticuerpos contra la anhidrasa carbónica tipo II[2,3]. Frecuentemente presenta manifestaciones extrapancreáticas, como la colangitis esclerosante y la nefritis tubulointersticial[4].

La colangitis esclerosante asociada a PA tiene hallazgos imagenológicos y presentación clínica similar a la colangitis esclerosante primaria (CEP), pero presenta una dramática respuesta a esteroides[5].

Describimos el caso de un paciente con episodios de colangitis y pancreatitis repetidas manejado como CEP sin respuesta, que desarrolló una nefritis tubulointersticial con hallazgos en biopsia renal sugestivos de proceso autoinmune, presentando resolución de las manifestaciones gastrointestinales y renales con esteroides.

CASO CLÍNICO

Varón de 37 años que consultó en marzo de 2006 por ictericia, fiebre y dolor abdominal; se pensó en episodio de colangitis, la colangiopancreatografía retrógrada endoscópica y p-ANCA tomados por sospecha de CEP fueron negativos, lo que hizo pensar en microlitiasis.

En mayo de 2006 se realizó esfinterotomía endoscópica. Ocho días después muestra nuevo episodio de colangitis. Se planteó posible colecistopatía acalculosa como explicación de colangitis recurrente; la gammagrafía con colecistoquinina fue compatible con el diagnóstico y se efectuó colecistectomía laparoscópica, pero a los 15 días presentó otro episodio de colangits.

Se retomó la sospecha diagnóstica de CEP, se hizo biopsia hepática que mostró colangitis aguda con mínimos focos de fibrosis. A principios de 2007 se realizó colangiorresonancia que demostró constricciones compatibles con CEP, sin posibilidad de intervención.

Recibió manejo como CEP, con ácido ursodeoxicólico y dosis bajas de antibióticos (ciprofloxacina), pero continuó presentando episodios de colangitis.

En octubre de 2007 consulta por fiebre y dolor abdominal, se inicia manejo con ciprofloxacina y se solicita tomografía de abdomen contrastada, previa creatinina, que informa 8,7 mg/dl. En mayo de 2007 la creatinina era de 1,2 mg/dl.

Se evalúa por Nefrología, se encuentra paciente con leve palidez como único hallazgo.

Exámenes: creatinina 7,6 mg/dl, nitrógeno ureico en sangre (BUN) 46, sodio y potasio normales, pH: 7,32, bicarbonato 16, Hb: 9,7 g/dl, uroanálisis con glucosuria (50 mg/dl) sin hiperglucemia.

La ecografía renal mostró riñones de tamaño normal, aumento en ecogenicidad bilateral.

Se diagnosticó insuficiencia renal aguda secundaria a nefritis tubulointersticial por consumo de quinolonas.

Al día siguiente, con el retiro del antibiótico e hidratación, la creatinina bajó a 5,5 md/dl y el BUN a 36 mg/dl. El complemento sérico fue normal, los anticuerpos antinucleares (ANA), la prueba serológica para la sífilis (VDRL) y para el virus de la inmunodeficiencia humana fueron negativos; proteinuria en 24 horas de 580 mg. Se le dio el alta con creatinina de 2,2 mg/dl.

Veinte días después regresa por fiebre, diarrea y edemas. Al ingreso presenta creatinina 15 mg/dl, potasio 5,8 mEq/L; citoquímico de orina: leucocituria, proteinuria (25 mg/dl), glucosuria (50 mg/dl), hematuria (eritrocitos 6 x ap). Examen físico sin hallazgos patológicos. Se consideró agudización de fallo renal previo; por la sospecha de nefritis tubulointersticial se inició manejo con prednisona y se realizó biopsia renal.

La biopsia renal reportó: nefritis tubulointersticial aguda, inmunofluorescencia: IgG: ++ (intersticio), IgA e IgM: +++ (intersticio), cadenas k y lambda: ausentes, C3: +++ periférico, M y cápsula de Bowman, ausencia de C1q. Interpretada como cambios histológicos compatibles con nefritis tubulointersti-