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Letters to the Editor - Case Report

Acute pancreatitis as initial manifestation in an adult patient with focal proliferative necrotizing purpura nephritis

Pancreatitis aguda como manifestación inicial en un paciente adulto con purpura y glomerulonefritis necrosante proliferativa focal

Dear Editor,

Henoch–Schönlein purpura (HSP) is a generalized vasculitis, which could cause a large variety of symptoms in different organs. Acute pancreatitis is an inflammatory disorder and may be life-threatening if it is severe. HSP could cause the acute pancreatitis rarely. It tends to develop in the first week of the illness after characteristic purpura, or develop later.[1] Exceptionally, acute pancreatitis developed during the course of HSPN as an initial presenting feature before the typical rash. Here we report an adult patient with HSPN presenting after the acute pancreatitis.

A 19-year-old man was admitted to our hospital with the chief complaint of "abdominal pain and abnormal urine test for 1 month, erythematosus purpura over legs for 5 days". He received the treatment for pancreatitis in the local hospital. However, the pancreatitis did not get remission. On admission of our hospital, the patient had a normal blood pressure and respiratory rate. There was a diffuse purpuric rash located over the legs. Abdominal examination showed tenderness in all quadrants with rebound tenderness. There was no hepatosplenomegaly, and bowel sounds were normal. Blood testing showed hemoglobin (101 g/l), white cell count (19.6×10^9 g/l), neutrophil (9.7×10^9 g/l). Erythrocyte sedimentation rate and C-reactive protein were significantly high at 29 mm/h and 100.3 mg/l, respectively. Serum testing showed glucose (3.3 mmol/l), urea (6.31 mmol/l), creatinine (52 umol/l), albumin (28.5 g/l), total cholesterol (4.1 mmol/l), and triglyceride (1.6 mmol/l). Serum amylase (332 U/l) and lipase (85.2 U/l) were increased. Urinary sediment examination showed microscopic hematuria. The proteinuria of 24 h is 2.48 g/2 L. Rheumatoid factor and complements (C3, C4) were within the normal range, and antinuclear antibodies, anti-DNA, antiphospholipids, antineutrophil cytoplasmic antibodies, and hepatitis viral markers were negative. Abdominal ultrasound showed a peritoneal effusion. Abdominal computed tomography (CT) scan noted an edematous pancreas (Fig. 1).

The result of renal biopsy was focal proliferative necrotizing purpura nephritis. Focal granular staining for IgA and C3 in the mesangium was shown using immunofluorescence staining. By light microscopy, glomeruli showed a mild hypercellularity in mesangial cells and matrix, focal endocapillary hypercellularity, eight crescents of 10 glomerulis, 1 necrosis of capillary loop. There is interstitial edema and inflammation composed mainly of mononuclear leukocytes and tubular epithelial injury (Fig. 2). The result of the light microscopy was confirmed by the electron microscopy.

The patient was diagnosed as purpura nephritis complicated by acute pancreatitis. He was started on therapy with methylprednisolone 40 mg/d intravenously for 5 days before the renal biopsy result. Afterwards, 0.5 g bolus of methylprednisolone was given intravenously for 3 days and after that prednisone orally (30 mg/day) and MMF orally (1.5 mg/day). Abdominal pain disappeared at the third day of the treatment and the pancreas was normal after two weeks' treatment. After 1-year follow-up, the patient recovered well with prednisone (10 mg/day) and MMF (0.25 mg/d), and renal function was normal, proteinuria was less than 150 mg/day, and abdominal CT scan was normal.

The association between pancretitis and Henoch–Schönlein Purpura Nephritis is rare. In our case, purpura nephritis complicated by acute pancreatitis was showed in this 19-year-old man. HSP is a rare cause of acute pancreatitis that can occur before or after the characteristic rash. In our case, the symptoms occurred before the rash. The

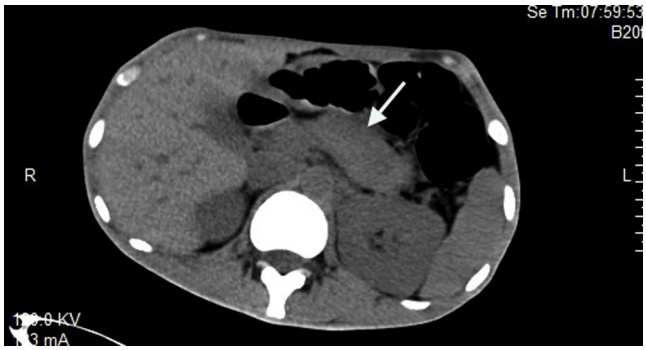

Fig. 1 – Edematous pancreas in abdominal CT scan.

pancreatitis tends to develop on the first day of the illness but may develop as late as day 75.[2] Acute pancreatitis is presumed to be caused by vasculitic involvement of the pancreas.

The clinical feature of HSP nephritis is quite variable. The pathology of this patient showed a mild hypercellularity in mesangial cells and matrix, focal endocapillary hypercellularity, eight crescents of ten glomerulis, 1 necrosis of capillary loop. In the literature, only two cases got renal biopsy of purpura nephritis with pancretitis. One case showed moderate-to-severe increase of mesangial matrix with crescent formation[3] and one case showed endocapillary proliferative glomerulonephritis.[2] There is no special feature in the pathology of those patients.

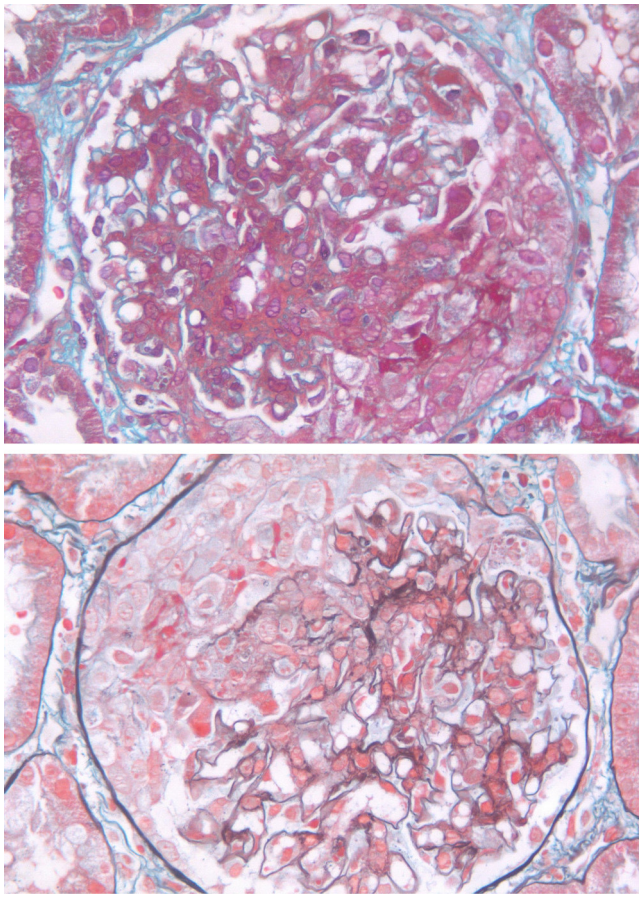

Fig. 2 – Crescents observed by light microscopy in Masson stained section (upper one, 400) and PAS stained section (lower one, 400).

Steroids have been used to treat HSP patients with pancretitis.[4–7] For most HSP related pancretitis without nephritis, parenteral nutrition and Nasogastric suction was used to treat the pancretitis,[8,9] and the outcome of pancretitis improved. In our case, the patient's pancretitis did not improve with the supportive treatment; however, the symptoms of pancretitis disappeared with the steroids usage. In the literature, two patients were diagnosed as HSP related pancretitis with nephritis. In the study by Frirui et al., kidney biopsy showed endocapillary proliferative glomerulonephritis and the patient was given 1 g bolus methylprednisolone intravenously for 3 days and after that prednisone orally (60 mg/day).[2] And in the study by Nie et al., as HSP nephritis (severe) was confirmed, intravenous methylprednisolone (500 mg/d) was given to the patient for 3 days followed by oral dehydrocortisone of 30 mg/d.[3] The two patients got complete remission in pancreas and kidney.

HSP is a rare and benign cause of acute pancreatitis. This complication could occur before the characteristic rash and shown as an initial manifestation of HSPN. Steroids could improve the outcome of the HSPN patients with pancreatitis.

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Respuesta a acetazolamida en paciente con calcinosis tumoral

Response to acetazolamide in a patient with tumoral calcinosis

Sr. Director:

La calcinosis tumoral (CT) es un raro trastorno del metabolismo del fósforo que se caracteriza por la formación de tumores periarticulares de fosfato de calcio[1]. La enfermedad es resultado de un defecto en la excreción renal de fósforo, debido a mutaciones en el factor de crecimiento de fibroblastos 23 (FGF23), Klotho y N-acetilgalactosaminiltransferasa-3 (GALNT3). La pérdida de función de FGF23 trae como resultado el aumento de la reabsorción tubular del fósforo, seguido del depósito en los tejidos subcutáneos[2].

Paciente varón de 23 años de edad, sin antecedentes importantes, inició su padecimiento a la edad de 18 años, con la presencia de una masa en la región dorsal y externa del glúteo y muslo derecho. El paciente informó del aumento progresivo del tamaño de la masa relacionado con actividad física, así como limitación de la función y dolor de la extremidad afectada. Seis meses antes de su ingreso a nuestra institución, se le realizó la escisión del tumor con las siguientes medidas: 18×7 cm del muslo derecho y 20×10 cm del glúteo derecho, con características amorfas.

A su ingreso presentó una masa sólida indolora en la región de escisión en la superficie exterior del muslo derecho, con dimensiones de $10 \times 4 \times 6$ cm. Los análisis de laboratorio reportaron niveles séricos normales de calcio, PA, creatinina, albúmina y PTH. La radiografía reveló una masa multinodular calcificada alrededor de la articulación de la cadera derecha.

La biopsia de la lesión reportó nódulos calcificados amorfos, algunos rodeados por proliferación de macrófagos y células gigantes tipo osteoclasto, separados por tejido fibroso denso, consistente con CT (fig. 1). Se realizó un seguimiento de 2 años, con acetazolamida (agosto de 2008 a octubre de 2011). La terapia demostró mejoría clínica y cese del crecimiento de las lesiones en el paciente. No se reportaron desordenes ácido-base durante su uso. Se realizó radiografía de control 7 años después del inicio del tratamiento, y es muy importante notar que no se observa incremento del tamaño de las lesiones (fig. 2).

La fisiopatología de la CT se centra en la anormalidad del metabolismo del fósforo[3]. La concentración sérica de fosfato se regula por la comunicación endocrina entre el esqueleto, el riñón y el intestino[4]. Los factores endocrinos implicados en el metabolismo del fósforo son la 1,25-hidroxivitamina D3, la hormona paratiroidea (PTH) y el FGF23. Este último sinergiza la acción de la PTH, reduciendo la expresión de los cotransportadores NaP(i)-IIa y NaP(i)-IIc en el borde en cepillo del túbulo proximal renal, aumentando la excreción renal de fósforo[5].

El FGF23 es una glicoproteína, formada por 251 aminoácidos con una región N-terminal y una C-terminal hecha de 71 aminoácidos[6]. Promueve la excreción de fósforo mediante la reducción de la expresión de los cotransportadores NaP(i) en el cepillo de la célula tubular proximal[3]. La unión de FGF23 a su receptor FGFR1c necesita ineludiblemente de Klotho para formar un heterodímero funcional[7]. El FGF23 es glucosilada por GALNT3[8]. Solo la proteína completa de FGF23 tiene actividad biológica. Cualquier alteración en estos puntos daría lugar a un aumento de la reabsorción tubular de fósforo, que es lo que ocurre en la CT[3].

La enfermedad se manifiesta con hiperfosfatemia y depósito masivo de calcio en tejido subcutáneo, siendo los principales síntomas que afectaban al paciente[3]. Esta enfermedad es más frecuente en mujeres y en afroamericanos, con un inicio de presentación en la infancia o la adolescencia temprana[2].

El tratamiento de CT se divide en la extirpación quirúrgica y el tratamiento médico. La extirpación quirúrgica de la lesión es un tratamiento bien documentado, sin embargo la recurrencia es común. Además, el paciente demostró recidiva tras su extirpación quirúrgica, debido probablemente a pobre circunscripción[9]. El tratamiento médico es preferible debido a la naturaleza metabólica de la enfermedad. Las terapias descritas incluyen la restricción de fosfato de la dieta, antiácidos, fármacos fosfatúricos y aglutinantes de fosfato. Sin embargo, la extirpación quirúrgica combinada con la privación de fosfato y el uso de acetazolamida, ha demostrado ser la terapia más eficaz[10].