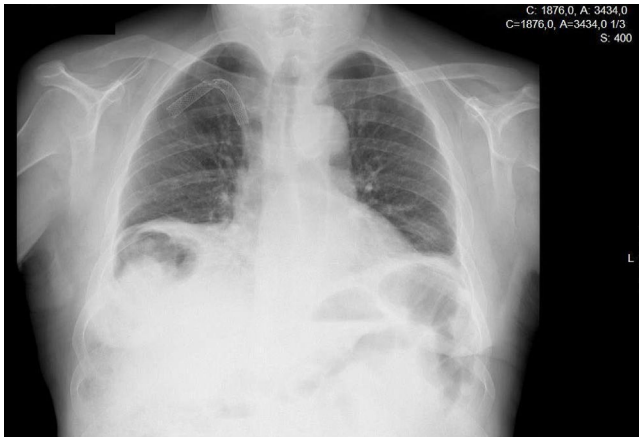

Figura 1 – Radiografía de tórax con síndrome de Chilaiditi.

obstrucción intestinal. Frecuentemente el cuadro se resuelve con tratamiento conservador, en caso de existir complicaciones, el tratamiento quirúrgico puede ser necesario llegando en ocasiones a requerir colectomía o colonopexia laparoscópica[4,5].

Tanto el signo como el síndrome de Chilaiditi son infrecuentes, siendo más frecuente en varones. Otros factores relacionados con un aumento en su frecuencia son el retraso mental y la enfermedad pulmonar obstructiva crónica, todos ellos presentes en este caso.

Consideramos que tanto el síndrome de Chilaiditi como la peritonitis esclerosante contribuyeron al cuadro clínico de suboclusión intestinal en nuestro paciente.

Entidades como el neumoperitoneo tienen expresiones radiográficas similares al síndrome de Chilaiditi (fig. 1), por ello en pacientes en diálisis peritoneal el diagnóstico puede pasar desapercibido más fácilmente. La realización de una TAC abdominal puede resultar de gran ayuda en estos casos[5,6].

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Segmental tuberous sclerosis in a patient with chronic kidney disease☆

Esclerosis tuberosa segmentaria en un paciente con enfermedad renal crónica

Dear Editor,

33-year-old woman presented with fatigue and renal failure. On physical examination, the patient had multiple non-traumatic periungual fibromas in both hands and feet (Fig. 1a). Shagreen patch was seen in the right lumbosacral region (Fig. 1b). On admission biochemical tests revealed; urea: 132 mg/dl, creatinine:4.7 mg/dl, potassium: 4.7 mmol/L, GFR

☆ This case was accepted as a poster presentation at Proceeding Book for ASN Kidney Week 2015, San Diego, USA.

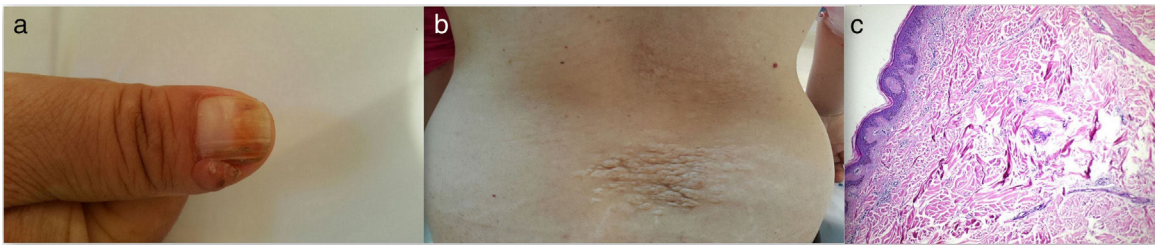

Fig. 1 – (A) Ungual fibroma. (B) Shagreen patch. (C) Shagreen patch. Biopsy specimen; collagen bundles in the dermis arranged in a haphazard manner (HEX40).

12 ml/min/1.73 m^2 (MDRD), pH 7.32, HCO_3: 18.4. Renal ultrasonography revealed bilateral renal cysts (17 and 5 mm at right kidney, 6 and 10 mm at left kidney) with normal size and parenchymal thickness. Histopathology of the Shagreen patch revealed dense collagen bundles in the dermis arranged in a haphazard manner (HEX40) (Fig. 1c). Due to these skin lesions and renal cysts the patient was diagnosed as TS (two major + 1 minor components). She denied any seizures in her past medical history. Renal biopsy was suggested but she did not accept the procedure yet and medical treatment was given. During the outpatient follow up creatinine level progressed up to 10 mg/dl. Due to low GFR and uremic symptoms hemodialysis was started and arteriovenous fistula was created.

TS complex is a multisystem, rare and genetic disorder of autosomal dominant inheritance which can involve different organs. It is caused by damage to the TSC1 (9q34) or TSC2 (16p13) genes coding respectively for hamartin and tuberin. Skin involvement is the commonest presentation of the disease. Recognizing specific dermatologic lesions, especially hypomelanotic macules (>3), Shagreen patch, forehead plaque, non-traumatic periungual fibromas, adenoma sebaceum, facial angiofibromas should remind the physicians TS. Renal involvement has the leading importance for mortality and morbidity. Kidneys are involved in almost 50–80% of the patients. The most frequent renal lesions in TS are angiomyolipomas and renal cysts. Bilateral and multiple characteristics of angiomyolipomas are primary radiological clues for suspicion and diagnosis. However in some rare cases these patients may present without frank renal lesions. These patients with renal involvement may also present with end stage renal disease in adult ages. TS is accepted as a glomerulocystic disease. They may occur in every part of the nephron. Renal cystic disease can also be microcystic, undetectable by imaging studies. The limited or obscure manifestations of TS with renal failure can be detected in nephrology practice.

Conflict of interest

The authors declare no conflict of interest.

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