

laboratory findings are leukocytosis, anemia, elevated acute phase reactant levels (erythrocyte sedimentation rate or C-reactive protein), microhematuria, proteinuria, glycosuria and/or pyuria. The ocular symptoms of uveitis include eye pain, red eye, loss of visual acuity and photophobia. The nephritis and uveitis do not normally present at the same time, a circumstance that further complicates the diagnosis. In 65% of the cases, nephritis is diagnosed first, in 21% it is uveitis that develops earlier, and in the remaining 15%, they appear simultaneously.¹ Mandeville et al.⁶ published a number of diagnostic criteria that require histological confirmation or extensive clinical evidence of acute tubulointerstitial nephritis and of uveitis. The kidney disease can have a good prognosis if treatment is begun early with systemic glucocorticoids. However, tubulointerstitial fibrosis and deterioration of the chronic renal function may develop if the disease goes untreated. For this reason, early diagnosis is essential. Recurrence of the uveitis is common (41%), but does not predict or imply recurrences of the kidney disease.⁷

The TINU syndrome is a condition in which the renal involvement can have a good prognosis with systemic glucocorticoid therapy. It is important that we know about it and include it in the differential diagnosis of uveitides and in that of acute tubulointerstitial nephritides. We consider multidisciplinary units to be increasingly necessary for cases like the one described here, as they would enable more precise diagnostic and therapeutic approaches.

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Bilateral Sudden Sensorineural Hearing Loss in a Patient With Microangiopathic Antiphospholipid Syndrome*



Anacusia súbita neurosensorial bilateral en un paciente con síndrome antifosfolípido primario microangiopático

To the Editor,

Sudden sensorineural hearing loss (SSHL) is defined as a sudden deterioration in auditory function of at least 30 dB; bilateral presentation (BSSHL) is extremely rare (<5% of all cases of SSHL).¹ The known etiological agents are viral infections, tympanic membrane perforation, vascular disorders and autoimmune diseases.² The autoimmune etiology was proposed by Ernst Lehnhardt in a patient with unilateral SSHL, who subsequently developed the same condition in the other ear.³ He postulated that the damage to the first ear led to the development of antibodies that affected the contralateral ear. The first associations of BSSHL with rheumatic diseases were reported in the 1980s in patients with systemic lupus erythematosus (SLE) and anticardiolipin antibodies (aCL).⁴ It was during the same period of time that Hughes pointed out the unusual presentations of thromboses, miscarriages and lupus anticoagulant in patients with SLE, an observation that would lay the foundations for the characterization of antiphospholipid syndrome (APS). Since then, the concept of APS has evolved considerably. In recent years, a variant with exclusively microangiopathic involvement (MAPS), in which virtually any organ can be affected, has been recognized.⁵

A 54-year-old man presented with BSSHL, headache and vertigo. He had had acute pancreatitis with secondary diabetes mellitus

4 years earlier and had a 7-year history of primary hypothyroidism. He underwent a neurotologic examination and, following audiometry, was diagnosed with bilateral neurosensory hearing loss, which was treated with transtympanic dexamethasone. A specialist in infectious diseases prescribed ganciclovir at a dose of 12 mg/kg/day for 10 days + prednisone at 50 mg/day for 4 weeks. Nerve conduction studies revealed short latency auditory evoked potentials with no response. Speech audiometry showed an absence of response to maximum intensities. The patient was referred to lip and face-reading therapy and to rheumatology because he had been found to have aCL. In the latter department, we reached a diagnosis of APS on the basis of high titers of IgM anti-β2 glycoprotein and aCL, a positive test for lupus anticoagulant and a prolonged activated partial thromboplastin time (aPTT). After immunological studies (Table 1), we ruled out the presence of lupus. In addition, in magnetic resonance imaging (MRI), we observed subcortical hyperintensity in bilateral frontal and parietal lobes, with normal vascular behavior and enhancement, findings that demonstrate microvascular involvement (Fig. 1). As the patient had never had large vessel thrombosis, we concluded that what he had was primary APS in its microangiopathic variant.

Table 1
Immunological Studies.

Test	Result
Antinuclear antibodies	80 homogeneous, 40 cytoplasmic
Anti-DNA antibodies	Negative
Anti-Sm antibodies	Negative
Lupus anticoagulant	1.69 (positive <1.2)
IgM anti-β2 glycoprotein	3350 (<8 U/mL)
IgG anti-β2 glycoprotein	6.8 (14.3 U/mL)
IgM anticardiolipin	>255 MPL (positive >15)
IgG anticardiolipin	46.3 GPL (positive >20.1)
Complement C3	296.7 mg/dL (90–180)
Complement C4	29.8 mg/dL (10–40)

GPL, IgG phospholipid units; MPL, IgM phospholipid units.

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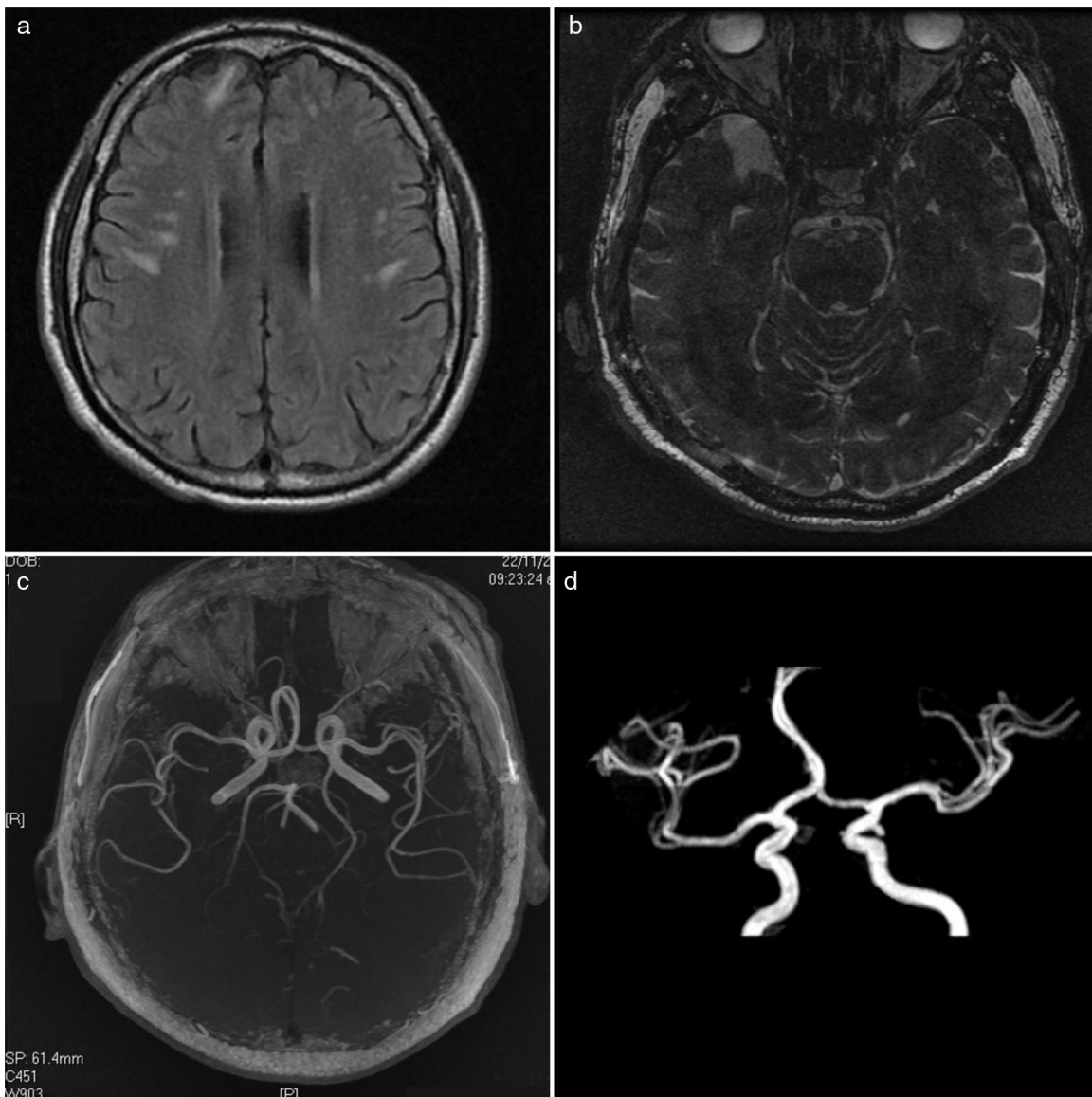


Fig. 1. (a) T1-weighted magnetic resonance imaging sequence showing a hyperintense area of microvascular damage in white matter of bilateral frontal and parietal lobes; (b) inversion recovery-weighted magnetic resonance imaging sequence showing lesions in right temporal lobe; (c) magnetic resonance angiography showing anterior cerebral circulation with no large vessel involvement; (d) magnetic resonance angiography showing normal circle of Willis.

We report a representative case of MAPS, diagnosed on the basis of the high titers of IgM anti- β 2 glycoprotein and aCL, the presence of lupus anticoagulant, prolonged aPTT and microvascular signs. Central nervous system involvement was made evident by the typical microvascular changes on MRI, characterized by lesions predominantly in the white matter because of the greater vulnerability of that region to ischemia.⁶ Bilateral SSSL is a very rare finding and, although its association with aCL is well established, the pathogenic mechanisms remain a mystery. It has been suggested that antiphospholipid antibodies (aPL) activate the endothelium of the cochlear blood vessels and that this overregulation produces local microthrombi and ischemia in the inner ear.⁷ With respect to the pancreatic manifestations, once other nosological entities have been ruled out, it is reasonable to consider that they may be related to MAPS. In this respect, the first case of pancreatic compromise in APS was described by Bird et al.⁸ and, since then, several cases have been reported. Autopsies show chronic inflammation and thrombosis in pancreatic arteries in the absence of vasculitis.⁹ Concerning the presence of hypothyroidism, the clinical significance of aPL in

thyroid autoimmune disease is still a subject of controversy. Current evidence suggests that the production of anti-thyroglobulin and anti-microsomal antibodies is accompanied by aPL synthesis as an epiphenomenon.¹⁰

In conclusion, BSSHL is a medical emergency with an ominous prognosis. There is no consensus with respect to the treatment, but it is highly recommendable that anticoagulation therapy begin immediately. Immunosuppressive therapy has not been found to be useful in the reported cases.

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Pelvic Osteoid Osteoma Simulating Sacroiliitis[☆]



Osteoma osteoide pélvico simulando sacroileitis

To the Editor,

The course of sacroiliitis is characterized by inflammatory pain in the lumbosacral and gluteal region. When the condition is bilateral, it is most commonly caused by spondyloarthritis. Sometimes only 1 side is affected and, in this case, it is necessary to rule out other less common causes, such as infections or tumors, among others.

We report the case of a 49-year-old man who presented with a 1-year history of pain in right buttock. He had previously been examined in orthopedics and rehabilitation, and brought with him a technetium bone scintigraphy that showed diffuse hyperactivity in right sacroiliac joint compatible with sacroiliitis.

The patient complained of pain in right buttock that waked him up during the night and was relieved by nonsteroidal anti-inflammatory drugs (NSAID). He had no history of traumatic injury, fever or arthritis, and reported nothing during work-up that suggested spondyloarthritis or recent infections. The physical examination was completely normal, with negative results in sacroiliac pain provocation tests (flexion, abduction, external rotation [FABER], distraction, compression and Laguerre tests), and the joint showed no peripheral or axial functional limitations. The neurological examination revealed no evidence of disease. The results of the laboratory tests, including the acute phase reactant levels, were normal. With respect to imaging studies, as there were no evident bone changes on plain radiography of the pelvis, the patient underwent magnetic resonance imaging (MRI) of the sacroiliac joints. There was a hypointense area on T1-weighted sequences (Fig. 1) and a hyperintense area in short tau inversion recovery (STIR) images (Fig. 2) on the border of the iliac side of the right sacroiliac joint, with an isointense central zone on T1-weighted sequences (Fig. 1), which was hypointense on the STIR sequences (Fig. 2). These findings pointed toward a definitive diagnosis of osteoid osteoma, which was confirmed by computed tomography (CT). The patient was treated immediately by means of CT-guided radiofrequency ablation and his symptoms disappeared.

Osteoid osteoma is a benign bone tumor that occurs in the femur or tibia in 50%–60% of the cases; between 7% and 10% are located in the spine.¹ Pelvic osteoid osteoma, like that of our patient, is less common. It occurs more frequently in males between the ages of 10 and 30 years. A typical symptom is nocturnal pain, which is

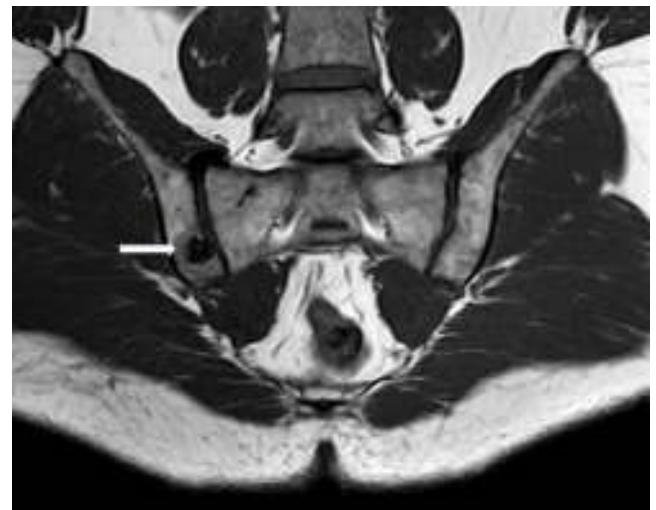


Fig. 1. Semicoronal plane of sacroiliac joint in a T1-weighted image, showing a hypointense area on the border of the iliac side of the right sacroiliac joint, with an isointense central zone.

relieved by NSAID.² The pattern of nighttime pain associated with this type of tumor can lead to its being mistaken, as in the case we report, for certain inflammatory rheumatic diseases, especially when located at certain sites, a circumstance that can result in a delay in the diagnosis. Although the osteoid osteoma is not often included in the differential diagnosis of sacroiliac pain, we should take it into account when the pain is relieved by NSAID, and in

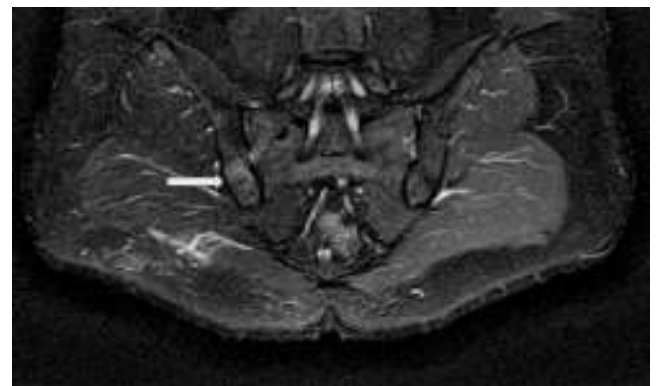


Fig. 2. Semicoronal plane of sacroiliac joint in a STIR sequence, showing a hyperintense area on the border of the iliac side of the right sacroiliac joint, with a hypointense central zone.

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