disease is described in the CAT scan of the case. Treatment included transplantectomy and wide-spectrum antibiotics. There was histological evidence of injuries consistent with EPN in the patient. Two microorganisms commonly reported to be isolated in the literature, *K. pneumoniae* and *K. oxytoca*, were found in cultures of the pathological specimen of this case.[6]

Companion diagnostics must be made with the presence of renal abscesses, xanthogranulomatous pyelonephritis, and renal tuberculosis.[7]

Even though our patient was not diabetic, the history of immunosuppression due to multiple previous renal transplants may become a risk factor associated with EPN.

This kind of clinical cases should be reported to further investigate their epidemiology and clarify the pathogenic aspects guiding the selection of optimal treatment for future cases.[6–9]

Conflicts of interest

The authors have no conflicts of interest to declare.

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Lupus anticoagulant-hypoprothrombinemia syndrome: A rare association in systemic lupus erythematosus☆

Síndrome de anticoagulante lúpico-hipoprotrombinemia: una extraña asociación en el lupus eritematoso sistémico

To the Editor,

Lupus anticoagulant-hypoprothrombinaemia syndrome (LAHS) is a disorder characterised by the acquired deficit of coagulation factor II (prothrombin) together with the presence of lupus anticoagulant. It is an extremely rare syndrome (less than 100 cases described in the literature),[1] in which there is a predisposition to bleeding, unlike antiphospholipid syndrome (APS), which is characterised by an increased risk of thrombosis.

The first case was described by Rapaport et al.[2] in 1960, but it was not until more than 20 years later that the study be Bajaj et al.[3] demonstrated the presence of anti-prothrombin antibodies. Although these antibodies do not impede prothrombin

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activity, they lead to secondary hypoprothrombinaemia due to rapid clearance of the antigen–antibody complexes from the circulation.

The most common treatment of LAHS consists of steroid therapy together with other immunosuppressors (cyclophosphamide, azathioprine, or rituximab). The aim of treatment if to reduce the risk of bleeding and eliminate prothrombin inhibitor.[4]

We present the clinical case of a 37-year-old Bulgarian man, with a history of lupus nephropathy diagnosed in his country in 2004. Despite receiving treatment, he had a poor clinical course, requiring renal replacement therapy 7 years after diagnosis. Two years later, he attended our hospital to continue haemodialysis. He was assessed by the rheumatology team and the haematology team for chronic thrombocytopenia, for which he received immunosuppressive treatment with steroids, immunoglobulins, and rituximab, as well as thrombopoietin-receptor agonists (eltrombopag), with a poor response. He also had APS and was positive for anti-cardiolipin antibodies and anti-β2-microglobulin antibodies. This presented with several thrombotic events (thrombosis of several vascular access devices) and a cerebrovascular event in 2014, for which he had been started on anticoagulant treatment.

The patient was admitted to our department for an episode of general malaise and low-grade fever in the context of a possible central venous catheter bacteraemia. He started broad-spectrum antibiotic treatment with a clear clinical improvement, and it was decided to stop anticoagulation temporarily to allow a change of venous catheter. During this period of withheld anticoagulation, he experienced a transient ischaemic attack which presented as right-sided paraesthesia and reduced power, with expressive aphasia. It was therefore decided to re-start anticoagulation with heparin sodium. Surprisingly, brain magnetic resonance angiography showed, along with old ischaemic lesions, a significant subdural haematoma at the left cerebral convexity with mass effect in the parieto-occipital region (Fig. 1). Between the neurosurgery and neurology teams, it was decided to stop anticoagulation and wait and see how he progressed clinically and radiologically. Given the presence of thrombotic and haemorrhagic events, a new coagulation study was requested. This showed lupus anticoagulant, elevated levels of anti-cardiolipin antibodies and anti-β2-microglobulin antibodies, and low coagulation factor II (prothrombin) activity, which corrected with mixing, confirming the presence of lupus anticoagulant-hypoprothrombinaemia syndrome (Table 1).

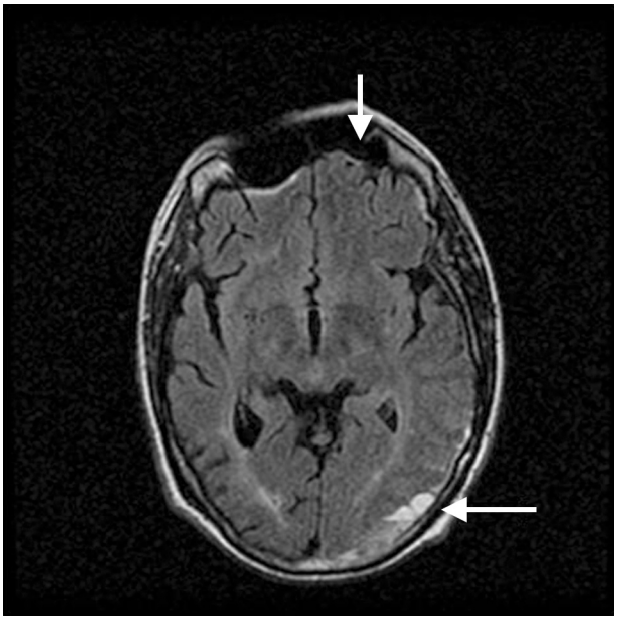

Fig. 1 – Magnetic resonance-angiography image of ischaemic lesions (frontal atrophy, arrow) and haemorrhage (left parieto-occipital region, arrow).

Table 1 – Clotting factor activity with and without correction for lupus anticoagulant.

Factor	Factor activity (%)	Corrected factor activity (SynthAFax®) (%)	Mixed factor activity (%)
Factor II	25	–	70
Factor V	102	–	–
Factor VII	136	–	–
Factor X	114	–	–
Factor VIII	16	83	–
Factor IX	6	96	–
Factor XI	8	94	–
Factor XII	7	98	–

Combined treatment was started with anticoagulation and immunosuppression with low dose steroids, mycophenolic acid, and rituximab 6-monthly.

The association of lupus anticoagulant with hypoprothrombinaemia is a rare syndrome, infrequently described. It is a syndrome that is more common in children and young adults, and is commonly associated with viral infections and auto-immune diseases (above all systemic lupus erythematosus), although cases have also been described in association with drugs or tumours.[1] LAHS is usually self-limiting when associated with viral infection, whereas in those associated with auto-immune diseases, relapse is common despite treatment.

It is clinically characterised by haemorrhagic diathesis, with epistaxes and ecchymoses being the most common types of bleed, although cases have been described of haematuria, gastrointestinal bleeding, and intracranial haemorrhage, amongst others[5] Several cases have been described of LAHS with associated thrombosis, in some cases multiple thromboses. However, those cases were mainly in the context of starting treatment against prothrombin inhibitor.

Treatment of LAHS is based on immunosuppression to avoid haemorrhagic events and to try to eliminate factor II-inhibitor.[4] An increase in thrombotic events has been described in patients with LAHS, as immunosuppression reduces inhibitor levels, but not lupus anticoagulant levels.[5] There are no guidelines indicating what the best treatment is for LAHS, most treatment being based on corticoids with another immunosuppressor.

In conclusion, in patients with lupus nephropathy with both thrombotic and haemorrhagic processes, lupus anticoagulant-hypoprothrombinaemia syndrome must be suspected. Diagnosis and treatment-based on immunosuppression to control the clinical manifestations of the disease are essential.

Conflicts of interest

The authors declare that they have no potential conflicts of interest related to the contents of this article.

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Barakat syndrome or HDR syndrome: Another association of kidney disease and deafness☆

Síndrome hipoparathyroidism, deafness and renal displasia o síndrome de Barakat otra asociación de sordera y nefropatía

Dear Editor,

If a patient has kidney disease and deafness, we think on Alport syndrome, a widespread entity. However, this is not always the case. One of the recent issues of the journal Nefrología included an excellent review of kidney disease in the context of mitochondrial diseases and how nephrologists should suspect this diseases in a nephropathy (tubulopathy or glomerular injury, manifested by kidney failure and proteinuria) which is accompanied by hearing loss or sensorineural deafness.[1] In this letter, we present a case of Barakat syndrome or hypoparathyroidism, deafness and renal dysplasia (HDR) syndrome, another disease that should be included in the differential diagnosis of kidney disease and hereditary deafness.[2,3]

A 32-year-old patient was admitted to the obstetrics department owing to oedema. She was 36 weeks pregnant and the laboratory tests revealed a plasma creatinine at 1.4 mg/dl and 6 grams of proteinuria in a 24-h urine collection. Medical history included familial hypoparathyroidism in chronic treatment with vitamin D and calcium carbonate, bilateral sensorineural deafness and left kidney agenesis. With this history, in 2007, she was diagnosed with HDR syndrome. A genetic study demonstrated the presence of the c.431 mutation in the GATA3 gene (gene for the transcription factor GATA3, located in the short arm of chromosome 10). Both the patient and her mother were heterozygous for this mutation. The rest of the family (father, sister and maternal aunt) did not have any clinical manifestations of the syndrome; nevertheless, a molecular genetic study ruled out the presence of this mutation. Prior to pregnancy, the patient had been examined in the urology department as she was a single-kidney patient. She had undergone laboratory testing that showed mild kidney failure with a serum Cr of 1.3 mg/dl and proteinuria at 2.8 g/day.

Since she did not have HTN, the presence of pre-eclampsia was ruled out, and she was diagnosed with worsening of renal function in a patient with chronic kidney disease (CKD) in pregnancy.

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