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http://dx.doi.org/10.1016/j.nefroe.2017.05.011

Pulmonary haemorrhage in a patient with IgA nefropathy☆

Hemorragia pulmonar en paciente con nefropatía IgA

Dear Editor,

Henoch-Schönlein purpura (HSP) is a leukocytoclastic vasculitis affecting small-vessels that is rarely seen in adults. Lung involvement is extremely rare, with an unpredictable clinical evolution and high rates of mortality.[1,2]

We report the case of a 69-year-old male patient with IgA nephropathy and a bilateral alveolar haemorrhage in the context of HSP. As for his personal history, he suffered from hypertension and atrial fibrillation, wore a mechanical prosthesis and was undergoing treatment with oral anticoagulants. The patient was admitted due to fever, dyspnoea, oedemas, coughing and expectoration. On admission, a deterioration of kidney function was observed (Cr 1.67 mg/dl; GFR 41 ml/min) as well as leukocytosis with a left shift. Following diuretic and antibiotic treatment, an improvement in kidney function occurred (Cr 0.9 mg/dl, GFR 82 ml/min) and the leukocytosis decreased. A few days later, the patient presented again deterioration of kidney function (Cr 2.1 mg/dl), anaemia (Hb 7.5 g/dl) along with purpuric lesions of the lower limbs. The tests performed showed: normal albumin, cholesterol and lactate dehydrogenase; an ESR of 52 mm, urine element and sediment with +++proteins, ++++Hb, >100 red blood cells/field, 24-hour urine protein of 1870 mg; IgA 437.00 mg/dl, normal remaining immunoglobulins and complement, antinuclear antibodies, anti-neutrophil cytoplasmic antibodies, anti-glomerular basement membrane antibodies and negative viral serology testing. Haptoglobins < 25, blood smear without schistocytes, positive direct Coombs test. Urological ultrasound with no pathological findings.

In light of suspected vasculitis, treatment was initiated with three boli of methylprednisolone 500 mg on consecutive days, followed by prednisone at 1 mg/kg every 24 hours. 72 hours later, symptoms of haemoptysis and anaemia appeared; a chest X-ray and chest CT scan were performed, showing findings suggestive of a bilateral alveolar haemorrhage and the presence of intraparenchymal pulmonary haematoma (Figs. 1 and 2). Treatment was initiated with a bolus of 500 mg/m^2 cyclophosphamide and immunoglobulins at 2 g/kg, spread across five doses. Five days later, frank haemoptysis, anaemia and significant respiratory effort occurred, prompting a transfer to ICU, where orotracheal intubation and mechanical ventilation were performed. The patient also required aspiration due to massive bleeding.

During the patient's stay in the ICU, six sessions of plasmapheresis were performed on alternate days, and treatment with corticosteroids was maintained. A clinical and analytical improvement was seen over the subsequent days and

☆ Please cite this article as: Belmar Vega L, Fernández-Díaz C, Palmou Fontana N, Rodrigo Calabia E, Martin Penagos L, Arias Rodríguez M, et al. Hemorragia pulmonar en paciente con nefropatía IgA. Nefrologia. 2017;37:347–349.

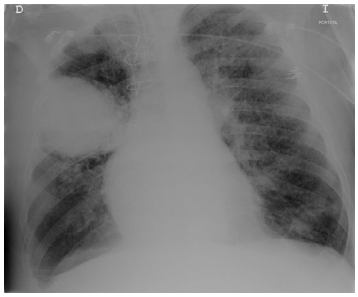

Fig. 1 – Alveolar-interstitial infiltrates secondary to intrapulmonary haemorrhage and haematoma.

extubation was performed 13 days later, with normalised kidney function and no new bleeding episodes. Finally, a kidney biopsy was performed in which 15 glomeruli were observed, four of which were sclerosed while the others showed the presence of diffuse segmental lesions in the form of endocapillary and mesangial proliferation and glomerular necrotic lesions. Interstitium with mild fibrosis and patchy inflammatory infiltrate. Positive immunofluorescence in the face of granular mesangial IgA (++), IgM (++) and C3 (++). After three months in hospital, the patient was discharged on a monthly immunoglobulin treatment and a decreasing corticosteroid regimen. Due to the risk of superinfection, combined treatment with other immunosuppressants was postponed until the pulmonary haematoma had resolved.

At the last hospital visit, from a clinical point of view the patient presented without dyspnoea, and clear improvements were seen in his lung X-ray and laboratory tests: creatinine 1.2 mg/dl, GFR 61 ml/min, urine with minimal microscopic haematuria (15–20 red blood cells/field) with a urine protein:creatinine ratio of 238.6 mg/g, and no anaemia or leukocytosis.

HSP is a form of leukocytoclastic vasculitis that affects the small vessels and, on rare occasions, the medium vessels. Its symptoms are characterised by non-thrombocytopenic palpable purpura (100% of cases), arthralgia in the lower limbs (82%), gastrointestinal involvement (50–75%) and renal involvement (20–50%). Musculoskeletal, neurological, pulmonary, cardiac and ocular conditions have been described less frequently.[1,2] The incidence in adults is around 1.3 cases per 100,000.[3]

The EULAR/PRINTO/PRES diagnostic criteria (2008) for HSP include, as a mandatory criterion, the existence of non-thrombocytopenic palpable purpura, predominantly in the lower limbs and, in case of atypical distribution, proven IgA deposition in the biopsy. In addition, at least one of the following characteristics is required: diffuse abdominal pain, biopsy with IgA deposition, arthritis or arthralgia and renal involvement (haematuria or proteinuria).[4]

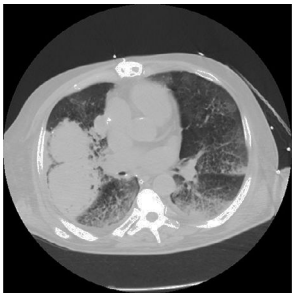

Fig. 2 – Intrapulmonary haematoma, areas with ground-glass opacity and crazy-paving pattern due to pulmonary haemorrhage.

Renal involvement usually manifests in the form of mild glomerulonephritis, with microscopic haematuria and proteinuria, and a biopsy showing IgA nephropathy.[5,6]

Lung involvement in HSP is extremely rare and includes, above all, haemorrhage or interstitial disease. Diffuse alveolar haemorrhage, which may present immediately following the HSP diagnosis, or even several years later, has an unpredictable clinical evolution and is associated with high rates of mortality.[7,8]

Chest X-rays are usually non-specific and computed tomography is required to confirm the findings. The severity of the situation warrants the use of intensive immunosuppressive therapy[9]; treatment combined with glucocorticoids and cyclophosphamide, azathioprine, methotrexate or rituximab is common. Plasmapheresis has also been noted as a therapeutic option.

The most common aetiologies that coexist with alveolar haemorrhage and glomerulonephritis are anti-neutrophil cytoplasmic antibody-associated vasculitis and anti-glomerular basement membrane antibody disease. Although uncommon, HSP should be considered among the differential diagnoses in a patient with pulmonary haemorrhage and renal involvement.

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http://dx.doi.org/10.1016/j.nefroe.2017.05.013

Hypertensive pulmonary edema related to desmopressin acetate

Edema pulmonar hipertensivo relacionado con el acetato de desmopresina

Dear Editor,

Desmopressin acetate, known as DDVAP (1-deamino-8-D-arginine vasopressin), increases factors VIII and vWF, shortens activated partial thrombin time and bleeding time. Desmopressin is the best treatment in emergency situation, increases the release of the von Willebrand factor from the endothelium in uremic bleeding.[1] Potential side effects of desmopressin include headache, nausea, malaise, hypotension, facial flushing, tachycardia, dizziness and hyponatremia.[2] However hypertension weren't defined before, we present two cases of hypertensive pulmonary edema after the infusion of desmopressin prior to ultrasound (USG) guided percutaneous kidney biopsy.

A 49 year-old female with end-stage renal failure secondary to unknown etiology received a living-donor transplant 3 years ago, was scheduled to have a transplant biopsy due to deteriorating graft function. Blood pressure was 160/90 mmHg, pulse was 72 beats/min, and respiratory rate was 14 breaths/min.

She was given an infusion of 15 μg of intravenous DDAVP over 20 min in preparation for biopsy. Biopsy performed without complications. After 40 min from the infusion; the patient suffers from dyspnea. Her blood pressure elevated to 220/140 mmHg. On examination there was crepitation on the both lungs on every area. SPO2 was 70 on the pulse oximeter.

Respiratory rate was elevated to 40. Immediately she was given furosemide and nitroglycerin infusion with oxygen. Her SPO2 levels elevated after the treatment. She was monitoring all day and her blood pressure fall to 140/80 after 8 h.

A 42 year-old male with crescentic IgA nephropathy, who was diagnosed 7 months ago, was treated with pulse steroid and cyclophosphamide for induction. On examination, the patient was relieve, body temperature was 36.6 C, blood pressure was 150/90 mmHg, pulse was 72 beats/min, and respiratory rate was 14 breaths/min.

He was given an infusion of 15 μg of intravenous DDAVP over 20 min in preparation for biopsy. Biopsy performed without complications. After 2 h from the infusion; the patient suffers from dyspnea. His blood pressure elevated to 230/120 mmHg. He was treated with oxygen, furosemide and nitroglycerin. His blood pressure was 150/90 mmHg after 8 h of intense therapy. His oxygen saturation had risen to 98 from 75 on room air.

Both of cases didn't have any history of coronary heart disease or heart failure but they have hypervolemia due to renal failure.

Hergesell et al. reported that risks in order to minimize, to adopt adequate biopsy technique is not only important, but also high-risk patients, especially uncontrolled blood pressure, with a clotting disorder, or has emphasized that it is necessary to exclude those unwilling to cooperate.[3] Mannucci et al. reported that prohibited for use in who have arterial disease. DDAVP related myocardial infarction and cerebrovascular disease has been reported to case presentations, can't be used in patients with known cardiovascular disease.[4]

DDAVP was an analog of vasopressin, may provoke symptomatic hyponatremia because of water retention. According to the U.S. Food and Drug Administration Agency, the half-life of DDAVP in patients with severe renal impairment can be extend by 9 h. Therefore, patients should be advised to restrict fluid intake from 1 h before to 9 h after administration of DDAVP. Blood sodium control is proposed following use in hyponatremic patient.[5,6]

There are no publications that specify the use of DDAVP in hypervolemic patient. In our cases DDAVP effected like vasopressin and increased water retention in the hypervolemic patients. In patients without structural heart disease DDAVP may have a role that could lead to hypertensive pulmonary edema. In our patients; increased blood pressure responded diuretic and vasodilator treatment.