

Funding

None.

Conflict of interest

The authors declare that they have no conflict of interest.

Acknowledgements

None.

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2173-5743/

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Response to: Madelung deformity[☆]**En respuesta a: Deformidad de Madelung**

To the Editor,

We read the publication by Ly-Pen and Andreu¹ in *REUMATOLOGÍA CLÍNICA* with great attention. They reported the case of a 39-year-old woman who had been diagnosed with bilateral carpal tunnel syndrome secondary to Madelung's deformity. She received a local injection of triamcinolone, which improved the symptoms. We would like to express our experience in the pediatric rheumatology unit of our hospital.

Madelung's deformity is a dysplasia of the distal radial physis. Its early closure is accompanied by a progressive deformity with dorsal dislocation of the ulna and palmar carpal displacement and, thus, supination and rotation are limited. It affects girls between the ages of 8 and 12 years, and is generally bilateral. This disorder is associated with a number of syndromes such as Leri-Weill dyschondrosteosis, Hurler syndrome, Turner syndrome, chondrodysplasia and Ollier disease.

We report the case of a 14-year-old girl who in October 2015 had been diagnosed with Leri-Weill syndrome. The disease was detected incidentally in a radiograph carried out due to a traumatic injury. She had mutations in the Short Stature Homeobox-containing (*SHOX*) gene and in pseudoautosomal region 1 (*PAR1*) in 5'. She was referred to pediatric rheumatology by her endocrinologist as she complained of mechanical pain in both carpi that had increased progressively in the preceding months and had a poor response to ibuprofen. The results of physical examination were consistent with bilateral Madelung's deformity, with pain on dorsal flexion of both carpi, with no



Fig. 1.



Fig. 2.

[☆] Please cite this article as: Muñoz Calonge MA, Murias Loza SM, Itza N. En respuesta a: Deformidad de Madelung. *Reumatol Clin*. 2018;14:60–61.



Fig. 3.

limitation (Figs. 1–3). There was no evidence of paresthesias or other neurological symptoms. Under sedation, the patient received the injection of 20 mg of triamcinolone in each carpi. In a check-up 1 month later, she reported a clear improvement, mentioning the nearly complete disappearance of the pain within 24–48 h after the injection, and an improvement in mobility.

We consider that, like the authors of the letter we comment here, local corticosteroid injection is a valid option in cases of carpal

pain secondary to Madelung's deformity during the pediatric age, even in the absence of symptoms of carpal tunnel syndrome. Moreover, as the skeleton of the child is still growing, it is necessary to postpone surgical treatment. Thus, in our opinion, local injection is an interesting conservative option given the potential relief of the symptoms, with a low probability of complications. Nevertheless, we have not found publications dealing with this therapeutic approach, nor of its medium- and long-term efficacy.

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2173-5743/

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Diaphragmatic Paralysis in Eosinophilic Granulomatosis With Polyangiitis[☆]



Parálisis diafragmática en granulomatosis eosinofílica con poliangiitis

To the Editor,

Eosinophilic granulomatosis with polyangiitis is an autoimmune disease that affects the small vessels. Its cause is unknown and a delay in the diagnosis is associated with a worse prognosis. Patients can be present with some degree of respiratory insufficiency due to asthma, pneumonia, alveolar hemorrhage and thromboembolism, as well as cardiovascular or neuromuscular problems.¹ There have been occasional reports of cases of unilateral diaphragmatic paralysis, with a variable impact on respiratory function.^{2,3} We present the case of a woman with eosinophilic granulomatosis with polyangiitis who also had hypercapnic respiratory failure associated with a polyneuropathy manifested by diaphragmatic paralysis and brachial plexopathy.

The patient was a woman who gave her age as 49 years old. She had been diagnosed as having asthma 4 years earlier, and had been admitted to the hospital several times with severe exacerbations, despite high doses of inhaled glucocorticoids. The last time she had been admitted she had required mechanical ventilation to resolve her asthma episode. Supplemental oxygen was delivered by facemask. Arterial blood gas analysis recorded a pH of 7.31, arterial carbon dioxide partial pressure (PaCO₂) of 62 mmHg, arterial oxygen partial pressure (PaO₂) of 79 mmHg, bicarbonate (HCO₃⁻) of 21.1 mEq/L and oxygen (O₂) saturation of 88%. Physical exploration disclosed an urticaria-like rash on her extremities and, particularly,

her left hand showed the sign of the “preacher's hand” (Fig. 1A). Complete blood count revealed severe eosinophilia with a level of 6000 cells/μL. Treatment included high doses of systemic glucocorticoids, aminophylline and nebulizers. Fortunately, the mechanical ventilator was withdrawn a few days later and she was finally extubated. However, radiological monitoring showed elevation of her left diaphragm. A fluoroscopic-guided study confirmed paralysis of that portion of the diaphragm (Fig. 1B and C). Computed tomography of the cervical spine and thorax ruled out myelopathy and nerve root compression. Electromyography of the left arm was consistent with brachial plexopathy and severe axonal injury of the proximal and distal ulnar nerve. A specimen obtained in a skin biopsy of the erythematous lesions revealed the presence of extravascular eosinophils. Nevertheless, perinuclear and cytoplasmic antineutrophil cytoplasmic antibodies (ANCA) were negative. A few weeks after hospital discharge, she recovered diaphragmatic mobility. However, there was no improvement in the ulnar neuropathy, but there was a notable decrease in the recurrence of exacerbations of the asthma.

Whereas the possible mechanisms that can produce respiratory insufficiency in a patient with eosinophilic granulomatosis with polyangiitis are well known, diaphragmatic paralysis has not been considered among them, even as a contributing factor. In the literature, we found a total of 5 cases of unilateral diaphragmatic paralysis in patients with this vasculitis. The mechanism underlying its production is similar to that described for other neuropathies secondary to autoimmune inflammation that involves the *vasa nervorum* resulting in axonal ischemia in the phrenic nerve.^{2,3} Moreover, involvement of the phrenic nerve has also been reported in other vasculitides.^{4,5} Idiopathic brachial plexopathy or Parsonage–Turner syndrome is a rare condition described in certain clinical situations, including giant cell arteritis and polyarteritis nodosa, in which the phrenic nerve can also be affected.⁶ Nevertheless, it is a painful syndrome that can be accompanied by important sensory abnormalities, a complication that was not observed in our patient.

[☆] Please cite this article as: Flores-Franco RA, Castillo-Soltero J, Pacheco-Tena CF. Parálisis diafragmática en granulomatosis eosinofílica con poliangiitis. *Reumatol Clin.* 2018;14:61–62.