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Acquired Madelung's Deformity in Rheumatoid Factor-positive Polyarticular Juvenile Idiopathic Arthritis^{*}



Deformidad de Madelung adquirida en la artritis idiopática juvenil poliarticular con factor reumatoide positivo

Dear Editor:

Juvenile Idiopathic Arthritis (JIA) is a heterogeneous group of diseases that is characterized by presenting arthritis of unknown etiology before 16 years of age and includes the systemic, oligoarticular and polyarticular with positive and negative rheumatoid factor (RF) types.¹ Persistent joint inflammation in these diseases may eventually lead to premature skeletal maturation and this, in time, originate a discrepancy in extremities, small vertebral bodies, deviation of the tibio-astragalus joint and Madelung's deformity of the carpus.²

We report the case of a 34-year-old male diagnosed with RF positive polyarticular JIA at 15 months of age, based on symmetric polyarthritis affecting elbows, wrists, metacarpophalangeal and proximal interphalangeal joints on the hands, as well as the knees, ankles and metatarsophalangeal joints and subcutaneous nodules, positive for RF (353 IU/ml) and elevated acute phase reactants (erythrocyte sedimentation rate of 45 mm/h, C-reactive protein 6.5 mg/l). He had no other relevant family or personal history. He was treated with nonsteroidal antiinflammatory drugs, systemic and intra-articular corticosteroids, gold salts and methotrexate but, despite this, the disease remained active with persistent synovitis of both wrists, developing bilateral subluxation at that level and limitation of dorsiflexion and supination. Radiographs of both wrists showed shortening of the medial distal radius, exaggeration of the radial tilt and proximal migration of the bones of the first row of the carpus, adopting a V shape between the radius and ulna, consistent with Madelung's deformity (Fig. 1). The patient also developed "trigger fingers" in both hands and bilateral hallux valgus. At age 24, he began treatment with etanercept, achieving control of disease activity. Currently no synovitis of the wrists is

present, but the limited movement persists, though it does not prevent him from carrying out his usual activities.

Madelung's deformity is a rare congenital disorder characterized by asymmetric carpal growth and curvature of the distal radial physis, resulting in a decrease in grip force of the hand. The common mechanism in Madelung's deformity is a premature closure of the epiphyseal growth cartilage in the medial and anterior portion of the distal palmar radius. This results in a shortening of the radius and, therefore, an apparent ulnar overgrowth. It is frequently associated with genetic syndromes such as Leri-Weill dyschondrosteosis and Turner's syndrome.³ Alterations similar to those produced in congenital Madelung's deformity have been described in other disorders, including trauma, tumors, infections, endocrine disorders and generalized skeletal dysplasias.^{3–5} The deformity may also occur in patients with the acquired form related to JIA because early skeletal maturation induced by joint inflammation can cause early closure of the carpal physis.^{2,6} There have been 2 cases of severe Madelung's deformity in patients with RF negative polyarticular JIA who also had Turner's syndrome,⁷ but in our case there was no evidence of another associated genetic syndrome.

Among the clinical and radiological characteristics of Madelung's deformity, dorsal and medial curvature of the distal radius, an increase in the inclination of the joint surface of the distal radius, the triangulation of the carpus with proximal migration and volar migration of the semilunar bone, as well as the prominent ulnar head, are common.⁸ This deformity is typically present bilaterally and manifested before 20 years of age. Patients have a limited pronation and supination of the affected carpus.^{3,5} Severe cases develop progressive osteoarthritis and instability of the distal radioulnar joint, radiocarpal osteoarthritis and may complicate with rupture of the extensor tendons of the fingers.³

There are no specific guidelines for the treatment of this deformity and it is initially conservative, postponing surgical correction until skeletal maturity is achieved. Several techniques have been described for the correction of Madelung's deformity, but no clear evidence exists that supports the specific use of any of them. Common indications for surgery are pain, limitation in mobility and esthetic concerns.³

In conclusion, acquired Madelung's deformity may occur as a complication of JIA, worsening the functional capacity even more in these patients, making it important to consider it in the patient's clinical and radiological evaluation.

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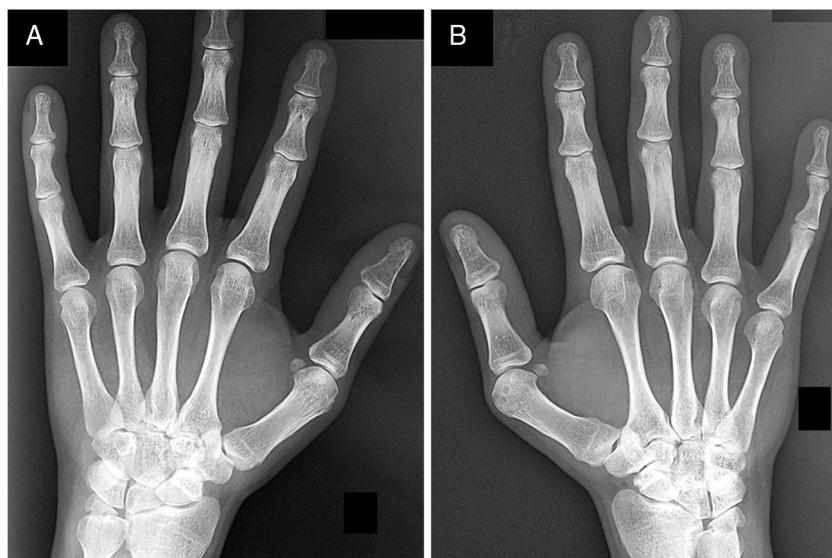


Fig. 1. Anteroposterior X-rays of the left (A) and right carpus (B) showing bilateral Madelung's deformity.

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Atypical Cogan's Syndrome Associated With Sudden Deafness and Glucocorticoid Response*



Síndrome de Cogan atípico asociado a sordera brusca y respuesta a los glucocorticoides

Dear Editor,

Cogan's syndrome (CS), described by David G. Cogan in 1945,¹ is a disease of unknown etiology, but probably has an autoimmune basis. It is characterized by the association of non-syphilitic interstitial keratitis and audiovestibular symptoms (similar to Ménière's syndrome), occurring in a period of less than 2 years. It occurs predominantly in young adult Caucasians. There is no predominance of gender.² When the eye or audiovestibular manifestations are different from what is seen, or the interval between them is more than 2 years, it is called atypic CS.³ It may be associated with other systemic manifestations such as aortitis and vasculitis.⁴ A patient with atypical CS is presented.

The patient is a 44-year-old male, hypertensive, with axial psoriatic arthritis since age 20 and anterior nongranulomatous

uveitis developed in the last 4 years. He attended consultation derived from Otorrhinolaryngology for bilateral Ménière's syndrome resistant to standard therapy, of 3 years of evolution. Audiometry revealed cophosis stood in the left ear and moderate hearing loss in the right ear (RE) at 60 dB (the patient wore a hearing aid). Physical examination showed psoriasis and redness in the extremities of both ears, which were rubbery to the touch. No peripheral arthritis was found but he had axial involvement (BASMI: 6). No other data were found upon examination. Laboratory tests showed: ESR of 49 mm in the 1st hour and CRP 25 mg/L. CBC, serum biochemistry and urinalysis were normal. Autoimmunity: rheumatoid factor, antineutrophil cytoplasmic antibodies, antinuclear antibodies and anti-CCP antibody were all negative. HLA B27 antigen was positive. C3 and C4 were normal. Radiographs of the chest, hands and feet were normal. The sacroiliac X-rays showed stage IV sacroiliitis. Electrocardiogram, echocardiography and computed tomography with intravenous contrast were normal. Atypical CS was diagnosed associated with relapsing polychondritis (no biopsy was performed) and psoriatic arthritis. Methotrexate (MTX) was started orally at doses of 12.5 mg/week. At 2 months he presented sudden hearing loss in the RE compared to baseline. The bilateral otoscopy was normal. Audiometry revealed a serious 60 dB drop in treble in the RE conversational frequencies. ESR and CRP were 86 mm and 45.30 mg/L, respectively. Treatment was initiated with boluses of methylprednisolone 1000 mg every 24 h for 3 days, showing a good response, recovering

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