Letters to the Editor

**Jaccoud's Arthropathy Associated With Reynolds Syndrome**

**Artrópata de Jaccoud asociada a síndrome de Reynolds**

To the Editor:

Jaccoud arthropathy (JA) is a recurrent form of arthritis that initially produces reversible deformities of the hands and feet (less frequently knees, shoulders and wrists) and can lead to erosions that are different from those in rheumatoid arthritis (RA). Its association with autoimmune diseases has been described, particularly systemic lupus erythematosus (SLE), but also with systemic sclerosis (SS). Primary biliary cirrhosis (PBC) is a chronic cholestatic disease of autoimmune origin, characterized by the presence of anti-mitochondrial antibodies (AMA) which is the most common liver disorder SS (especially the limited cutaneous form), with an estimated prevalence of 3%–50%. This association is known as Reynolds (RS) syndrome and its first description was in 1971. Although arthritis occurs in 4%–42% of patients with PBC, JA development has not been previously described in this single entity or in RS.

We present the case of an 80-year-old woman, diagnosed at 57 with seronegative RA based on symmetric polyarthritis affecting metacarpophalangeal (MCP) and proximal interphalangeal (PIP) of the hands, wrists, hips and knees. She underwent treatment with gold salts, which were suspended after one year due to impaired cholestatic liver function, with a biopsy compatible with PBC. Since then she has received varying doses of corticosteroids, NSAIDs on demand and ursodeoxycholic acid 600 mg/day. The liver function remained stable but she developed multiple joint deformities; as a result of steroids, she developed osteoporosis with inferior pubic ramus fracture and osteonecrosis of the left hip. From the age of 68 she began presenting Raynaud’s phenomenon in the hands and feet without digital ulcers, xerostomia and xerophthalmia.

In March 2010, she was referred to our center for back pain secondary to a sacral fracture and vertebral collapse. Examination revealed nasal sharpening, microstomia, facial telangiectasias and distal sclerodactyly, subluxation and ulnar deviation of MCP predominantly on the right, ‘gooseneck’ deformity and ‘Z’ deformity of the thumbs (Fig. 1A), and hallux valgus and subluxation of metatarsophalangeal joints, with no signs of active synovitis. Cardio-pulmonary auscultation was normal. Laboratory tests revealed slightly elevated liver enzymes (alkaline phosphatase 144 U/l, gamma glutamyl transferase 93 U/l), C-reactive protein 6.62 mg/l, erythrocyte sedimentation rate 55 mm/h, antinuclear antibody 1/320 with centromere antibodies and positive AMA, but the results for nuclear extractable antigen antibodies, cyclic citrullinated-peptide (anti-CCP), and rheumatoid factor (RF), complement and immunoglobulins were negative or normal. Radiology confirmed the hand deformities described and demonstrated the absence of erosions; calcification of the triangular ligament of the right carpus was also observed (Fig. 1B), and capillaroscopic study revealed the presence of dilated and tortuous loops without avascular areas. The diagnosis of SS limited to the skin associated with PBC and JA was established, being discharged with prednisone 5 mg/day and calcium and vitamin D, and adding, in a later visit, alendronate 70 mg/week, with adequate fracture recovery and good pain control.

Joint involvement in SS appears in 46%–97% of cases and may be the initial manifestation prior to Raynaud’s phenomenon in 12%–65%. It presents as joint pain, arthritis, morning stiffness and functional limitation secondary to flexion contractures, relating both to joint inflammation and mechanical restriction secondary to periarticular fibrosis. Radiographic changes are usually mild.

**Fig. 1.** Jaccoud arthropathy on the hands, with ulnar deviation of the MCP, ‘gooseneck’ deformity and ‘Z’ thumb deviation with these deformities being more pronounced on the right hand (A). X-ray of the hands in which, in addition to the abovementioned deformities we find juxtaarticular osteopenia, chondrocalcinosis of the right carpus and absence of erosions or degenerative signs (B).

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but some patients develop erosive arthritis similar to RA, with it associating with calcinosis and acroosteolysis. However, the development of JA is quite uncommon, with only 4 cases reported to date, so it is considered that it may correspond more to the coexistence of idiopathic JA than a manifestation of SS. It has been suggested that JA in SLE associated with inflammatory activity due to prolonged or recurrent low degree swelling of the synovial membrane and joint capsule, and ligamentous laxity cause an imbalance of muscle forces. However, in cases of associated JA and SS there is no clinical or imaging inflammation evident, so other mechanisms could be involved, including pericapsular and tendon fibrosis. A symmetrical erosive arthritis of small joints, often RF positive, has been described in PBC, which can be indistinguishable from RA. Up to 31% of PBC cases develop arthritis, characterized by a non-deforming asymmetric affection (although cases have been reported with deforming/erosive arthritis), negative for RF and anti-CCP, and recently it has been shown that it has a special histopathology regarding synovial infiltration predominantly by B lymphocytes and plasma cells. However, JA is not described in these patients. Pyrophosphate deposition disease has also been associated with JA, so that may have contributed to its development in our patient because there was evidence of chondrocalcinosis of the right wrist, but not in previous episodes of arthritis at that level. In conclusion, the development of JA in the RS seems to be rather incidental, because it is not properly a joint manifestation of any of the entities that make up this syndrome.

References


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We included, from January to December 2010, adult patients with clinical manifestations of RA and no previous evaluation by a rheumatologist or DMARD treatment. Patients were sent to different family physicians or from the internal medicine specialist to the rheumatologist of a regional General Hospital of the Instituto Mexicano Seguro Social (IMSS). We defined RA based on the 1987 criteria proposed by the American College of Rheumatology. We evaluated the following timelines: time from onset of symptoms to the visit to the family physician; time from disease onset to the first visit to the rheumatologist and time since disease onset and DMARD treatment onset.

Mean age±SD of the 98 patients was 38±9 years; 85% were female; 49 had an RA disease progression of 1–6 months, the diagnosis established by the family physician was RA in 79% of patients and the mean number±SD of visits the patient received before referral to rheumatologist was 6.6±5.8. In 19, 33, 23 and 24% of patients, the onset of DMARD treatment was 1–3, 4–6, 7–12 and >13 months after RA symptom onset, respectively. The mean time for the patient to receive medical attention was 2.9 months, and for referral to Rheumatology from primary care was 6.6 months. For DMARD treatment onset it was 9.9 months.

Our results show that only in 19% of patients was DMARD started in the first three months after disease onset and the delay in the prescription of DMARD was mainly due to the delay in referral from family medicine to the rheumatologist. Studies performed in the past 2 decades which evaluate the onset of DMARD treatment in patients with early RA, performed in the US, Spain, Canada, the United Kingdom, the Middle East, and in European countries, show that the mean time since the onset of disease and the onset of DMARD treatment ranges from 6 to 18 months, similar to our findings (mean 11 months). This indicates that the diagnosis of RA after