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Jaccoud's arthropathy

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Jaccoud's arthropathy (JA) is a condition characterised clinically by 'reversible' joint deformities such as swan neck, thumb subluxation, ulnar deviation, 'boutonniere' and hallux valgus, along with an absence of articular erosions on a plain radiograph. JA was initially described in patients with rheumatic fever (RF), but as this disorder has become rare the main clinical entity associated to JA at present is systemic lupus erythematosus (SLE). JA has also been described in other connective tissue diseases, infections and neoplasia. In general, its prevalence in either SLE or RF is around 5%. The etiopathogenic mechanisms of JA are not known, but some authors have suggested an association with hypermobility syndrome. Several studies have attempted to identify an association of different antibodies with JA in SLE patients, but their findings do not allow for the drawing of any definite conclusions. Newer imaging techniques such as magnetic resonance and high-performance ultrasonography have revealed the presence of small erosions in joints of a few patients with JA. Presently, the therapy for JA is conservative and based on the use of non-hormonal anti-inflammatory drugs, low doses of corticosteroids, methotrexate and antimalarials. The role of surgery through either the realignment of soft tissue around the joint – or more aggressive procedures such as arthrodesis, silastic implant and arthroplasty – needs to be proven.

More than a century ago François-Sigismond Jaccoud described a deforming arthropathy in a patient who had suffered several attacks of rheumatic fever (RF) [1]. This articular manifestation was then later named 'Jaccoud's arthropathy' (JA) [2,3]. Besides its classical description in RF [4–16] in

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which its prevalence is of 4.2% [10], such arthropathy has also been described in diffuse connective tissue diseases, particularly systemic lupus erythematosus (SLE) [17–29], infections and neoplasia.

As opposed to what happened in the past when the prevalence of RF was high, presently the majority of the cases of JA are associated to SLE. In this disorder its prevalence is around 5% [30]. However, in a previous study, 'swan neck' deformity was observed in 38% of the SLE patients. This finding can be attributed to a bias in the selection of these patients in a tertiary rheumatologic attendance centre [31]. However, as there are no definitive diagnosis criteria for JA, it is not surprising that its prevalence varies in different series. In other conditions associated to JA, the epidemiology is not known, and as it is a rare phenomenon it appears in the literature as case reports. Thus, JA was observed in Sjögren's syndrome [14,32] scleroderma [33,34], dermatomyositis [34], psoriatic arthritis [35], vasculitis [36–43], ankylosing spondylitis [44], mixed conjunctive tissue disease [2,45], pyrophosphate deposition disease [46], neoplasia [47,48], hypermobility syndrome [49], chronic pulmonary diseases [50], inflammatory intestinal disease [51], Caroli's disease [52], Borreliosis [53], human immunodeficiency virus (HIV) infection [54], mycosis fungoids [55], sarcoidosis [56], gangrenous pyoderma [57], KID syndrome (keratitis, ichthyosis and deafness) [58], chronic eczema [59] and angioimmunoblastic lymphadenopathy [60]. Curiously, JA has also been described without any associated diseases, a form referred to as 'idiopathic' or in the elderly population as 'senescent' [61–63].

Since the clinical features of JA may mimic those of rheumatoid arthritis (RA), it is very important to recognise the potential for misdiagnosis, which may lead to inadequate management.

The main topics to be addressed in the present review are clinical settings, diagnostic issues, imaging, management and perspectives for basic and clinical research.

Clinical and laboratory aspects of Jaccoud's arthropathy

The classical clinical manifestation of JA is the presence of 'reversible' joint deformities, mainly in hands, but also observed in other joints, such as the feet [11,23,64–66], knees [18,67] and shoulders [68]. It may be symptomatic or painless and may even occur without any previous history of arthritis [4,22,59,69,70]. In this regard the report by Levin [4] is well illustrated in the following statement: "The patient had extremely mild manifestations of rheumatic fever and had had two episodes of chorea. Later rheumatic heart disease with pure mitral stenosis was diagnosed. Never were there symptoms related to any of the joints, and deformity of the hands developed so gradually that the patient was not fully aware that it had occurred until she was an adult. "

The joint deformities most frequently seen in JA are ulnar deviation, swan neck, 'z'-thumb, 'boutonniere' and hallux valgus, which, apart from their reversible character, are quite similar to those seen in RA. These deformities vary in intensity and it is not rare for them to be present without being noted by the patient or his/her physician. On the other hand, particularly in older well-established deformities, there may be limitation of function of the involved joints, probably secondary to the local residual fibrosis leading to contractures. In patients seen in this stage, the misdiagnosis of RA is common. It should be emphasised that although 'reversible' and sometimes painless, JA is associated with considerable loss of quality of life [30]. Fig. 1(A) demonstrates a classical JA case in an SLE patient, illustrating the reversibility of the deformities in hands (B). Rarely, diffuse subcutaneous swelling in the hands can be observed (Fig. 2) [31].

In 1950, Bywaters observed for the first time that patients with RF and JA had more valvular heart disease (VHD) [71]. Curiously, some cases of JA have been described in patients with hypocomplementemic urticarial vasculitis and concomitant non-rheumatic valvular disease, probably mediated by immune complexes [36,41]. Recently, it was demonstrated that in SLE there is also an association between JA and VHD [72].

Clinically significant tenosynovitis has rarely been described in JA probably because other features of the disease overshadow the symptoms related to the tendon sheaths, but in some patients crepitus may be detected [73]. A very intriguing point is a supposed association of JA with a predisposition for tendon rupture, at least in SLE patients. In a recent systematic review of the cases of tendon rupture in SLE patients, both were observed in a total of 55 published cases. The most frequently observed rupture sites were the patellar and Achilles' tendons. Sixteen patients concomitantly had JA (29%) suggesting that such association has been underestimated [74].