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LETTER TO THE EDITOR

Jaccoud's arthropathy: proper classification criteria and treatment are still needed

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Dear Editor,

A deforming arthropathy was first described in patients with rheumatic fever (RF) by François-Sigismond Jaccoud more than a century ago [1], and it has been widely known in the literature as Jaccoud's arthropathy (JA). Presently, the majority of the cases of JA are seen in systemic lupus erythematosus (SLE) [2]. Either in RF or in lupus, its prevalence is around 5 %. Such arthropathy has also been described in other connective tissue diseases, neoplasias and infections [3].

JA is characterized clinically by "reversible" joint deformities such as swan neck, thumb subluxation, ulnar deviation and "boutonniere." However, the term "reversible" seems to be inappropriate because once the deformity is installed it lasts forever. Likewise, although the "reducible" nature of the arthropathy is demonstrated in the majority of the cases, sometimes the longstanding process of fibrosis in soft tissues of the joints can lead to a limitation of their mobility, making the diagnosis of JA still more difficult. Although mainly observed in hands, JA has also been seen in other sites such as feet, knees and shoulders [4]. The classical imaging feature of JA is the absence of articular erosions on a plain radiograph.

The mechanisms responsible for the development of JA are not entirely known, but it is recognized that the deformities are a result of soft tissue abnormalities including laxity of ligaments and joint capsule and secondary deviation of the tendon from its axis with the contribution of muscular imbalance. Synovitis may contribute to the process, but it is not as aggressive as in rheumatoid arthritis (RA). Some studies have suggested an association of JA with hypermobility syndrome, and others have attempted to establish an association with different antibodies in SLE patients, but their findings did not lead to any definite conclusion.

A limitation on the study of JA is the lack of definite diagnostic or classification criteria. Previous attempts to classify JA were made based on the presence of "reversible deformities" and absence of erosions on X-rays and rheumatoid factor negativity [5] or as "any deviation of the metacarpus finger axes assessed by a goniometer" [6]. Spronk et al. [7] developed a diagnostic "index" which allowed for the presence of different deformities and attributing JA a score of over five points. None of these sets of criteria has been universally accepted.

Furthermore, to make the definition of JA more complicated, it has also been observed in individuals without any associated disease, a form referred to as "idiopathic" or in the elderly population as "senescent" [8, 9]. In fact, it is not rare to find typical "swan neck" deformity in normal population as an individual phenotypical feature. Sometimes, other members of the same family may have such deformities. In these cases generally, there is no other type of joint deformity.

Thus, we understand the definition of JA is more appropriate if the criteria below are fulfilled:

1. Typical joint deformities such as swan neck, thumb subluxation, ulnar deviation, "boutonniere," *genu recurvatum*, *hallux valgus* and flat feet, which are correctable in a passive position.
2. Presence or history of articular inflammation in the deformed joints, regardless of its intensity or etiology (RF, SLE, etc.).

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3. Absence of similar deformities in other healthy members of the same family.
4. No erosion on plain radiographs regardless of the finding of erosions on magnetic resonance or high-performance ultra sound exam.

Even more frustrating than the absence of classification criteria is the lack of an effective therapeutical approach for patients with JA. The joint deformities can be severe enough to lead to a considerable loss of joint function as well as quality of life.

There is no guarantee that the conservative strategy based on the use of non-hormonal anti-inflammatory, low doses corticosteroids, methotrexate and antimalarial drugs will be able to avoid the development of deformities. The benefit of physical therapy as well as the use of orthotic devices needs to be proven. Although there are a few reports on surgical procedures to correct JA, their indication, the best modality and when to indicate them are not known.

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Conflict of interest None.

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