## Lusorian artery simulating rheumatic diseases

Sirs,

Our patient (LF) was first admitted to our clinic at 13 years 10 months of age for right hand oedema and articular pain. At 11 years 4 months of age, he first complained of hand tumefaction (mainly the right hand) associated with pallor, pain (more intense with prehension) and functional impairment that started suddenly in the morning and then regressed. During the following months new transient episodes occurred and disappeared after 24-48 hours. Different diagnoses were made (transient arthritis, episodic arthritis, algodystrophy) and the patient sporadically received NSAIDs, with transient resolution of symptoms.

On our first observation, the patient's appearance was that of a normally developed adolescent, with a negative clinical examination and apyrexia. All blood and serum tests such as rheumatoid factors, antinuclear antibodies, C3, C4, HLA-B27, AGA, EMA, PRIST, RAST, LDH, CK, fT3, fT4, TSH, C1q were negative, except for a slight increase in the ASO titer (343 IU/ml) and anti-deoxyribonuclease B (588.5 IU/ ml). Moreover, Toxoplasma, CMV, EBV, Yersinia and Borrelia antibodies were absent.

During hospitalization, the child suddenly presented right hand oedema, pallor, pain and functional impairment that regressed spontaneously after about 48 hours (Fig. 1). The patient history (sudden symptom onset and spontaneous disappearance within 24-48 hours,etc.), clinical symptoms (transient oedema of the right hand and functional impairment), and negative in-flammatory parameters suggested a different etiology from connective tissue disease. In performing the differential diagnosis, vascular malformation was considered. This etiological hypothesis was confirmed by angioMR, which showed the presence of aberrant right subclavian artery (ARSA) originating in the aortic isthmus and running anteriorly to the vertebrae and posteriorly to the oesophagus and trachea, and car-otid arteries originating in the aortic arch and connected to it through a short common trunk.

In addition, capillaroscopy showed the presence of diffuse anomalies secondary to right hand neoangiogenesis (1), and lymphoscintigraphy showed a significantly slower superficial lymphatic drainage of the right hand with tentative compensation by the deep lymphatic vessels.

We diagnosed Lusorian Artery (LA), consisting of a mechanical obstruction to lymphatic flow in the right subclavian artery. In our case, the clinical manifestation was characterized by periodic exacerbations, especially when the child assumed incorrect postures. Symptoms disappeared after microsurgical correction consisting of multiple lymphatic-venous anastomoses at the third medium arm (2-3). Follow-up over 3 years showed no relapse of oedema.

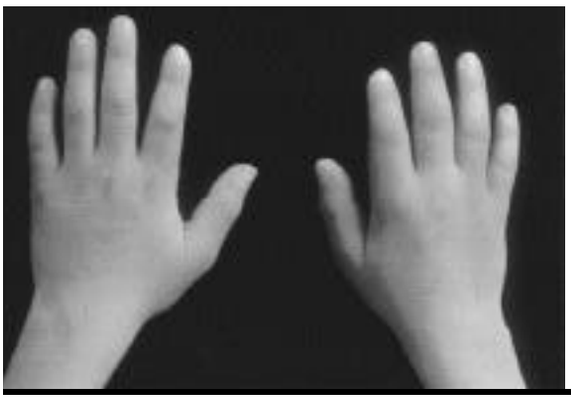

**Fig. 1.** Photograph of the patient's hands, in which diffuse swelling distal to the right joint is evident.

LA is the most frequent congenital aortic arch anomaly, although rarely observed (4), and is characterized by ARSA running posteriorly to the trachea and oesophagus in 78% of patients, between the oesophagus and trachea in 18%, and slantwise anteriorly to the trachea in 4% (5, 6). It was first described by Hunauld in 1735 (7), The incidence is 0.6 – 0.8% in the general population, and about 60-70% of patients remain asymptomatic during their lifetime (8).

ARSA is to be suspected in case of dysphagia, dyspnea or ischemic symptoms in the upper limbs not ascribable to other diseases. Other less frequent symptoms include chest pain, cough, wheezing (4,5), and vertigo. In infancy, the symptoms are correlated with compression of the mediastinal structures such as the trachea and oesophagus or with congenital cardiac anomalies that can be associated with ARSA. Our patient was first admitted to our observation for suspected rheumatic disease and in particular juvenile idiopathic arthritis, which was excluded by the presence of the transient symptoms of pain,oedema and functional impairment of right hand and wrist, frequent spontaneous resolution and the regular recurrence of the clinical picture. In addition, blood tests as well as instrumental and clinical examinations (negative swab) suggested against the diagnosis of acute articular rheumatism (AAR). A differential diagnosis with septic arthritis was also considered, but normal inflammatory indexes and apyrexia excluded this hypothesis; in addition, the absence of Toxoplasma, CMV, EBV, Yersinia,and Borrelia antibodies excluded the diagnosis of post-infectious arthritis.

Only instrumental examinations such as angio-MR, capillaroscopy and lymphoscintigraphy were able to demonstrate the presence of ARSA, the true cause of the patient's symptoms. Our case shows that in suspected rheumatic diseases the differential diagnosis with congenital vascular anomalies is important.

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