Letters to the Editor

A boy with bizarre hands mimicking an inflammatory chronic disease

Sirs,

A 14-year-old boy was brought to our attention in December 2004 with the diagnosis of polyarticular juvenile idiopathic arthritis (JIA), due to the apparent swelling of metacarpophalangeal joints (MCP) with ulnar deviation of both hands. Family medical history was negative for neurological, metabolic and autoimmune diseases. Pregnancy and delivery had been uneventful, and his psychomotor development was normal. At birth, both feet were reported as slightly extrarotated, but this mild clubfoot improved over one year with orthopaedic shoes. Plain x-rays had ruled out skeletal dysplasia. Conversely, already at birth, both hands showed a mild ulnar deviation that progressively worsened. During the previous two years, joint swelling of involved fingers was recorded. Notwithstanding some clinical findings similar to those of JIA, the boy did not complain of morning stiffness, pain or limited motion. He was a soccer player and did not report any significant problem. On examination, he was in good clinical condition and there was no swelling, nor any functional limitation in any of the joints. The ulnar drift was associated with flexion deformity at the MCP joint (Fig. 1) The patient did not have contracture nor webbing of the thumb, no hypoplasia of the forearm and hand muscle, nor were the digital extensors hypoplastic. Routine blood tests, including rheumatoid factor autoantibodies and thyroid function, were all normal. Lysomal storage disorder was excluded through extensive tests for metabolic diseases. Plain hand x-rays showed ulnar deviation of both hands in absence of localized osteopenia, reduction of articular space, and of bone erosions. Hand and wrist magnetic resonance with gadolinium excluded synovial enhancement, and bone scintigraphy did not show bone uptake at any site. JIA was excluded (1), the boy was seen by the hand orthopaedic surgeon and the diagnosis of congenital ulnar drift (CUD) was made (2, 3). The patient underwent surgery, and the abnormal subcutaneous bands were resected (4). The ulnarly dislocated extensor tendons were relocated by first dissecting them transversally and then the proximal stump of the extensor tendon was fixed to the base of the first phalanx (5, 6), followed by a successful recovery of hand appearance.

The syndrome of CUD is a rare condition that encompasses multiple malformations involving the hands in a bizarre aspect, consisting of an ulnar deviation of the MCP (9). The thumb is characteristically adducted with flexion of the MCP joint and hyperextension of the interphalangeal joint. This thumb-in-palm deformity is typical of CUD, even though it might mimic an inflammatory chronic process leading to finger alterations (10). A defect in embryological development is hypothesized (8). The anomaly may also be inherited as an autosomal dominant trait, but in the family of our patient, no other members were reported with this abnormality.

The differential diagnosis of JIA is wide and encompasses inherited and acquired conditions. Many chromosomal and congenital disorders can present with bone and joint abnormalities. However, a good history and physical examination may discriminate between malformative and inflammatory disorders. Ulnar drift of the fingers can be present in adult rheumatoid arthritis, as well as in children with polyarticular JIA, especially in teenagers with rheumatoid-factor positive onset. However, our patient did not have any of the clinical, laboratory or radiological features of JIA. Additionally, the appearance in our patient of bone and joint deformities shortly after birth was more suggestive of a congenital abnormality, rather than an acquired, inflammatory disease. Our patient had isolated CUD syndrome since no other associations were detected. This case underlines the possibility of rare, inherited disorders in the diagnostic work-up of a child with musculoskeletal signs and symptoms starting at an early age that may be erroneously diagnosed as chronic inflammatory disease.

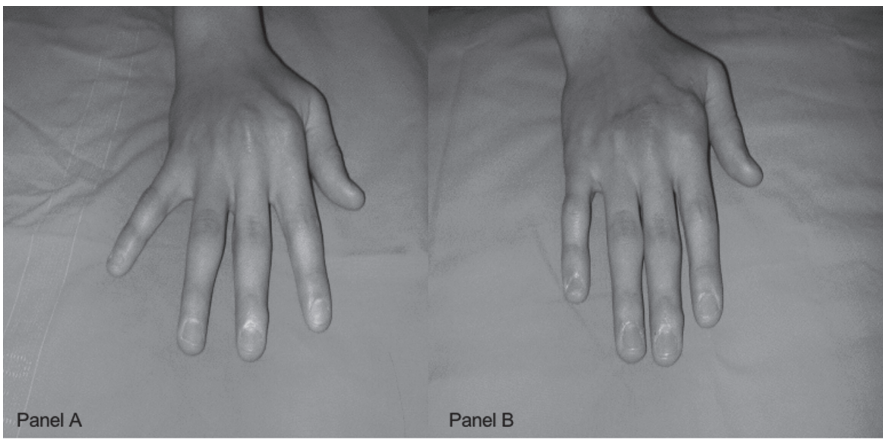

Fig. 1. Panel A: Ulnar deviation of right hand's metacarpophalangeal joints (MCP). The thumb is adducted with flexion of the MCP joint and hyperextension of the interphalangeal joint.
Panel B: Right hand after surgery.

F. Falcini
R. Cimaz[1]
L. Ricci
S. Fanner[2]
M. De Martino
M. Ceruso[2]

Department of Paediatrics, Rheumatology unit and [2]Hand Surgery, University of Florence; [1]Universitè Claude Bernard, Lyon, France.

Address correspondence to:
Fernanda Falcini, MD, Departmento di Pediatria, U.O. di Reumatologia, Università di Firenze, Via Pico della Mirandola 24, 50132 Firenze, Italy.
E-mail: falcini@unifi.it

Competing interests: none declared.

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