Letters to the Editor

Peritonitis and cervical arthritis as presenting manifestations of systemic juvenile idiopathic arthritis

Sirs,

Early involvement of the cervical spine is uncommon and peritonitis is a rare extra-articular presentation in systemic-onset juvenile idiopathic arthritis (sJIA) (1-3).

We describe a previously healthy 4-year-old boy with C1-C2 arthritis and peritonitis presenting as the initial manifestations of sJIA. He was admitted following 2 days of fever, lethargy, vomiting, abdominal pain and neck stiffness. A blood count showed 20100 leucocytes/ml (74% neutrophils), the C-reactive protein was 2.8 mg/dl. Cerebral spinal fluid analysis excluded meningitis. Twelve hours post admission he appeared clinically septic, developed abdominal signs suggestive of peritonitis and C-reactive protein increased to 10 mg/dl. Antibiotics (ampicilin, gentamicin and metronidazole) were started. Exploratory laparotomy revealed a normal appendix, mild ascites (not analysed), and multiple lymph nodes with non-specific reactive hyperplasia on histology. Over the following days he maintained high fevers, generalized abdominal pain and neck stiffness. He initiated mucousy and bloody diarrhoea. Mild hepatosplenomegaly and axillary limphadenopathy were noticed. Ultrasound showed moderate ascites and pleural effusion. Blood, peritoneal and stool cultures were negative. Ceftriaxone was started and he was sent to our tertiary care centre. On day 13, a non-itchy, evanescent and faint macular salmon-pink rash was observed, particularly in the legs. At this moment the haemoglobin was 8.9 g/dl, ferritin levels were raised (>15000 ng/ml; normal < 370) and erythrocyte sedimentation rate (ESR) was 47 mm/h. Antinuclear antibodies and rheumatoid factor were negative. Due to continued neck stiffness and decreased neck movements, cervical bone radiography and bone scintigraphy were carried out, both normal, and cervical MRI showed a small effusion and discrete enlargement of the two apophyseal joints between C1 and C2 (Fig. 1). Echocardiography revealed mild pericardial thickening and a small pericardial effusion with normal cardiac function. On day 20, prednisone (2 mg/kg/day) and acetylsalicylic acid (80 mg/kg/day) were started and antibiotics stopped. Despite maintaining an evening peak of pyrexia (38ºC) until day 49, clinical and laboratorial improvements were evident, including normal mobilization of the neck after day 36. Therapy was gradually tapered after 12 days of apyrexia and acetylsalicylic acid was stopped 2 months later. Three months after presentation he developed a morning right lower limb limp, from which he improved. At the seventh month of disease (on 10 mg prednisone on alternate days) he relapsed with fever, macular salmon-rash, right hip arthritis and neck and hand tenderness. He presented leucocytosis (18730/ml, 94% neutrophils) and a high ESR (81 mm/h). Methotrexate was started (15 mg/m^2 once a week, orally) and prednisone increased to 15 mg/day, resulting in a good outcome.

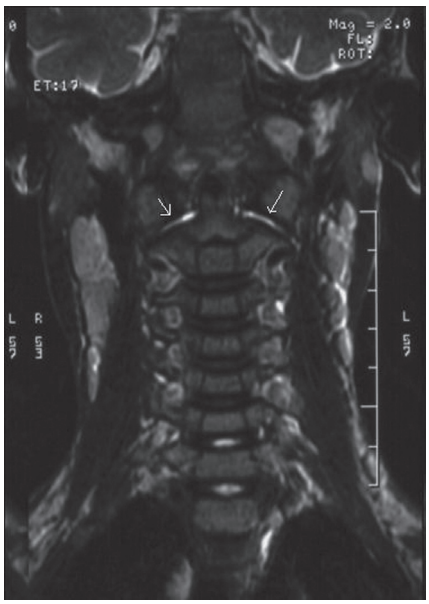

Fig. 1. Cervical MRI: arrows showing bilateral effusion and enlargement of the apophyseal joints between C1 and C2

Remission was achieved two years after presentation. Prednisone and methotrexate were respectively stopped 2 months and 2 years later. Five years following the initial presentation he is symptom-free and leads a normal life.

Diffuse abdominal pain suggestive of serositis is present in about 10% of children with sJIA (1), but reports in the medical literature of peritoneal involvement are rare (2, 4, 5). Although only a single joint may be affected, usually, multiple joints are involved simultaneously in sJIA (2). Atlantoaxial compromise is a rare finding at the early stages of the disease or as the single joint implicated (6-9). In our patient, who presented with fever, neck stiffness and signs of peritoneal inflammation, meningitis and peritonitis secondary to appendicitis were excluded. The associated evanescent macular salmon-pink rash and cervical spine involvement suggested sJIA. Diagnosis was confirmed by the outcome with a relapse.

Over 21 years we observed 264 cases of JIA, of whom 3 had monoarticular C1-C2 involvement lasting for several months (2 oligoarthritis previous to this case).

We are not aware of any other published cases of sJIA who presented with C1-C2 arthritis and peritonitis simultaneously .

A. VAZ, *MD*
C. VEIGA, *MD*
P. ESTANQUEIRO, *MD*
M. SALGADO, *MD*

Department of Pediatric Rheumatology, Hospital Pediátrico de Coimbra, Portugal.

Address correspondence to:
Dr. Ana Vaz, Avenida Bissaya Barreto, Hospital Pediátrico de Coimbra, Consulta de Reumatologia, 3000-076 Coimbra, Portugal.
E-mail: apvj76@gmail.com

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