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## Letter to the Editor

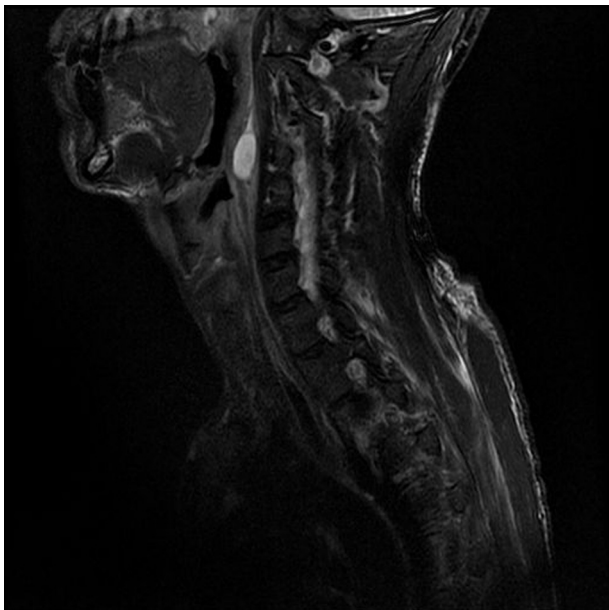
### Adult-onset Still's disease presenting as retropharyngeal abscess



A 33-year-old man, with no notable medical history, was admitted to hospital with a 10-day history of neck pain and fever, associated with upper dysphagia and odynophagia. Clinical examination demonstrated fever of 39°C and blood pressure of 140/70 mmHg. The laboratory work-up showed an inflammatory syndrome (CRP: 140 mg/L), leukocytosis with 17,000 leukocytes/mm<sup>3</sup> and 14,000 granulocytes/mm<sup>3</sup>, hepatic cytolysis (ALAT: 160 IU/mL and ASAT: 120 IU/mL) with no apparent site of infection (urine cultures and blood cultures remained sterile), negative Rickettsia, Epstein–Barr virus, HIV, Coxsackie, adenovirus, and cytomegalovirus serologies, negative immunological work-up (antinuclear antibody, rheumatoid factor and antineutrophil cytoplasmic antibody), and normal chest X-ray. Neck MRI demonstrated retropharyngeal abscess (Fig. 1). Aspiration of the abscess revealed pus, for which direct examination and culture were negative. The patient was treated with amoxicillin and clavulanic acid together with surgical drainage of the abscess, and the subsequent course was marked by a transient, non-pruritic macular rash, arthritis of the ankles and knees, hepatosplenomegaly and cervical lymphadenopathy. Serum ferritin was elevated to 1500 µg/L, and glycosylated ferritin was markedly decreased to 12%. The

diagnosis of adult-onset Still's disease (AOSD) was established on the basis of Yamaguchi's criteria (Table 1) [1]. Administration of a bolus of methylprednisolone (240 mg) for three days, followed by prednisone 1 mg/kg/day allowed resolution of the clinical signs and correction of laboratory parameters.

AOSD is a systemic disease characterized by persistent fever, rash, arthralgia or arthritis, sore throat, hepatomegaly or splenomegaly and leukocytosis with granulocyte predominance, hepatic cytolysis and high serum ferritin and very low glycosylated fraction. The diagnosis is based on Yamaguchi's criteria [1] after excluding infectious, haematological or autoimmune diseases. Retropharyngeal abscess is rare in adults and is usually observed in a context of trauma [2] or immunodepression [3]. In the case reported here, retropharyngeal abscess led to the diagnosis of AOSD; this association is exceptional, as only one case has been reported in the literature [4]. The diagnosis of retropharyngeal abscess is difficult in this setting due to the non-specific clinical features (infectious syndrome, dysphagia and odynophagia) corresponding to the features of AOSD. Odynophagia, which is observed in about 2/3 of patients with AOSD, may be the first sign of the disease, but can also occur during subsequent flare-ups and contributes to the diagnosis of AOSD [5]. The diagnosis of retropharyngeal abscess must be considered in the case of absence of improvement or deterioration of clinical features during



**Fig. 1.** Neck MRI demonstrating retropharyngeal abscess in T2-weighted sequence, with cyst showing a high-intensity signal.

**Table 1**

Yamaguchi's criteria for the diagnosis of adult-onset Still's disease.

#### Major criteria

1. Fever of at least 39°C for at least one week
2. Arthralgias for at least two weeks
3. Typical rash (a)
4. Leukocytosis (at least 10,000/mm<sup>3</sup>) with at least 80% of granulocytes

#### Minor criteria

1. Sore throat
2. Lymphadenopathy (b) and/or splenomegaly (c)
3. Abnormal liver function tests (d)
4. Negative tests for antinuclear antibody and rheumatoid factor (e)

#### Exclusion criteria

- I. Infections (especially sepsis and infectious mononucleosis)
- II. Neoplasms (especially lymphomas)
- III. Systemic diseases (especially PAN and RA with extra-articular signs)

At least five criteria are required, with at least two major criteria in the absence of any exclusion criteria

- (a). Non-pruritic salmon-coloured macular or maculopapular rash, usually transient (at the time of fever spikes)
- (b). Recent and significant lymph node enlargement
- (c). Splenomegaly confirmed by palpation or ultrasound
- (d). Elevation of transaminases and/or LDH related to the disease, excluding drug toxicity or any other cause
- (e). Negativity of the usual tests performed to detect IgM rheumatoid factor and antinuclear antibody by immunofluorescence
- (f). Each criterion can only be taken into account in the absence of another explanation

LDH: lactic dehydrogenase; PAN: polyarteritis nodosa; RA: rheumatoid arthritis.

treatment of AOSD (in our patient, retropharyngeal abscess was the first sign of AOSD in the absence of corticosteroid therapy). Computed tomography and MRI are highly contributive to the diagnosis [2]. The micro-organisms most commonly isolated are *Streptococcus pyogenes*, *Staphylococcus aureus* and *Haemophilus influenzae*, but all cultures remained negative in our patient. Antibiotic therapy alone may be insufficient and most authors recommend surgical drainage in addition to antibiotics [2].

#### Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

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