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Case Report

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Multi-System Inflammatory Syndrome in Children (MIS-C) with Retropharyngeal Involvement Mimicking Abscess: the Therapeutic Role of Anakinra

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Abstract

Multi-System Inflammatory Syndrome in Children (MIS-C) is a new diagnostic entity that appeared with the Covid-19 pandemic, and that only concerns the pediatric age. Early diagnosis and early immunomodulatory treatment lead most of the time to a complete resolution of the pathology. We present a case of MIS-C, whose main clinical feature was neck pain due to inflammation of retropharyngeal tissues. The use of anakinra was critical for healing.

Keywords: Anakinra; Covid-19; MIS-C; Pediatric

Case Report

A seven-years-old girl was admitted to our pediatric unit because of persistent fever for nine days, she was not responsive to broad-spectrum oral antibiotic therapy, associated with headache, sore throat and neck pain. Physical examination showed a pharyngitis with a medial shift of the left tonsil, left lateral cervical lymphadenopathy and painful neck stiffness. There were no signs of meningeal irritation, conjunctival injection or rash, cardiothoracic objectivity was normal. Initial blood tests revealed an elevated C-reactive protein (CRP) level (47,6 mg/L), white blood cell count in range (5640/uL) with a mild lymphopenia (1190/uL), normal hemoglobin and platelets count, liver and kidney function tests in range, normal coagulation tests and troponin levels. Intra-abdominal and thoracic infectious foci were excluded (normal abdominal ultrasound and chest X-ray). Intravenous antibiotic therapy with Ceftriaxone and Clindamycin was started. A Magnetic Resonance Imaging (MRI) was performed showing inflammation of the parapharyngeal and retropharyngeal tissues, with uncertain presence of an abscess (Figure 1).

After two days of parenteral antibiotic therapy the patient was still feverish and the clinical condition further deteriorated, with inappetence, oliguria, worsening of neck pain with significant stiffness, plus occurrence of bilateral non-secretive conjunctival injection, mild cheilitis, and abdominal pain. The presence of persistent fever unresponsive to antibiotic therapy, mucocutaneous inflammation, lymphadenopathy, high level of CRP, associated with previous Covid-19 infection detected, led to the diagnosis of Multi-System Inflammatory Syndrome in Children (MIS-C). Therefore, the patient was treated with intravenous immunoglobulin (IVIG) in combination with methylprednisolone 2 mg/kg and low dose acetylsalicylic acid. In spite of an initial improvement, three days after administration of IVIG with ongoing steroid therapy, there was a recurrence of conjunctivitis, painful neck stiffness, headache and fever, therefore a therapy with anakinra (4 mg/kg/day intravenous) was started, in combination with a second dose of

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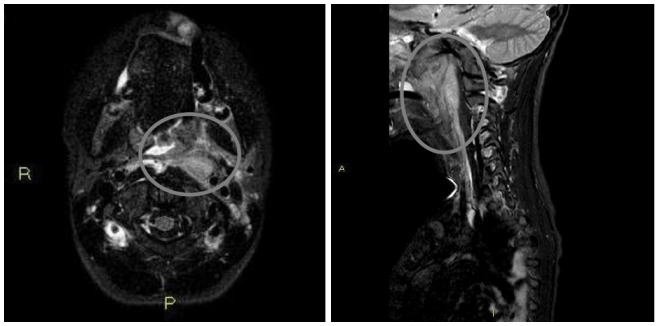

Figure 1: MRI imaging of interested parapharyngeal and retropharyngeal tissues.

IVIG, carrying on methylprednisolone. This therapy resulted in a swift clinical improvement, fever regression and CRP negativization. No heart involvement was observed on follow-up.

Discussion

Retropharyngeal edema is a rare presentation of Kawasaki Disease (KD) [1]. At MRI image, an edema and an abscess have features in common and this can lead to a clinical misjudgment. Nevertheless, an abscess usually is characterized by central liquefaction [2,3]. In literature, a few MIS-C cases present symptoms like neck pain, trismus, voice change, drooling, dysphagia, odynophagia or rarely stridor and respiratory distress with radiological findings similar to retropharyngeal abscess [4]. Both in KD and MIS-C, an important involvement of cervical lymphatic tissue can be observed, de facto KD is also called "mucocutaneous lymph node syndrome". In addition to superficial cervical lymph nodes deep lymphatic tissues can also be affected, mimicking a retropharyngeal/peritonsillar abscess [5,6]. In these cases, the international consensus guidelines on MIS-C recommend a multistep anti-inflammatory treatment [7]. First-line therapy is IVIG (dose of 2 mg/kg) plus low-to-moderate–dose glucocorticoids (1–2 mg/kg/day). In patients who do not achieve clinical improvement within 24-48 hours, an intensified therapy is recommended with either high-dose (10–30 mg/kg/day) glucocorticoids, anakinra (recombinant human IL-1 receptor antagonist, dose of >4 mg/kg/day and often 5–10 mg/kg/day), or infliximab (10 mg/kg IV x 1 dose) [7]. In more severe patients with organ dysfunction anakinra can be considered as first line therapy [7]. In the reported case, the use of anakinra in combination with a second dose of IVIG led to immediate clinical improvement, until complete healing.

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The authors have no conflicts of interest relevant to this article.

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