A case report of adult-onset Still’s disease of a patient presenting with the symptoms of intraparotid lymphadenopathy

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Abstract

Adult-onset Still’s disease (ASOD) is rare in adults. We herein report a 21-year-old female patient with swelling on right cheek, fever, sore throat, and myalgia for two months. The radiologic, pathologic, laboratory and serologic investigations led to no specific diagnosis. Our case fulfilled 3 major (fever, arthralgia, and leukocytosis) and 4 minor (pharyngitis, lymphadenopathy in the neck and parotid gland, abnormal liver function test and negative for Rheumatoid arthritis) criteria for the diagnosis of ASOD according to Yamaguchi et al. This was a rare case presenting with intraparotid lymph nodes and diagnosed with ASOD. After corticosteroid treatment, all symptoms, liver function tests and inflammatory parameters regressed to normal ranges.

Keywords: Still’s disease, Intraparotidal lymphadenopathy, Pharyngitis, Fever

Introduction

Adult-onset Still’s disease (AOSD) is a rare inflammatory disorder. The initial symptoms of the disease include arthritis, daily high fever, and skin rash [1]. The etiology of AOSD, which was first described by George F., is still unknown [2]; however, it is thought to be related to bacterial infections such as yersinia enterocolitica, mycoplasma pneumoniae [3] and viral infection, such as rubella [4]. The diagnosis is made by exclusion of infection, malignancy, and rheumatologic disorders. Elevated ferritin levels are considered particularly valuable for the diagnosis of the disease.
Case presentation

A 21-year-old female patient presented to the ear, nose, and throat (ENT) department with complaints of swelling on her right cheek, sore throat, myalgia, arthralgia, and fever for 2 months. She had no other symptoms. The patient’s medical history included tonsillitis episodes (2-3 times per year) and no autoimmune and chronic diseases in her family medical history.

Physical examination revealed pharyngo-tonsillitis and swelling on her right cheek. Her blood pressure was 130/75 mmHg, her heart rate was 90 beats per minute (bpm), there were multiple lymph nodes in her neck and her body temperature was 39.0°C (102.2 °F). Two grams of ampicillin/sulbactam was administered per day for five days, but no change was observed in the symptoms.

The neck ultrasound and MRI scan showed intraparotid lymph nodes (27x14 mm) and numerous lymph nodes at levels 2, 3 and 5 (Figure 1).

Figure 1: MRI showing intraparotid lymph node

A lymph node biopsy was performed on the neck for exclusion of lymphoma and tuberculosis, which revealed no signs of malignancy, tuberculosis, and other diseases, except nonspecific reactive paracortical hyperplasia (RPH).

Laboratory test results were as follows: White blood cell count: 24.120/mm³ (normal range: 4-10x10³/mm³), neutrophil ratio: 84.9%, C-Reactive Protein level: 20 mg/dl (normal range is 0-0.5 mg/dl), ferritin: 3720.39 ng/ml (normal range is 4.63-204), AST: 254 U/L (normal range is 5-34 U/L), ALT: 236 U/L (normal range is 0-33 U/L), and urine and stool culture were normal (Table 1).

Table 1: Laboratory test results

<table>
<thead>
<tr>
<th>Test</th>
<th>Result</th>
<th>Unit</th>
<th>Normal Range</th>
</tr>
</thead>
<tbody>
<tr>
<td>White Blood Cell</td>
<td>24.120</td>
<td>%/uL</td>
<td>4000:10000</td>
</tr>
<tr>
<td>Neutrophils</td>
<td>20</td>
<td>mg/dl</td>
<td>-</td>
</tr>
<tr>
<td>C-Reactive Protein</td>
<td>4.63-204</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ferritin</td>
<td>254</td>
<td>U/L</td>
<td>5-34</td>
</tr>
<tr>
<td>ALT</td>
<td>236</td>
<td>U/L</td>
<td>0-33</td>
</tr>
<tr>
<td>Urine Examination</td>
<td>Normal</td>
<td>Normal</td>
<td></td>
</tr>
<tr>
<td>Culture</td>
<td>Normal</td>
<td>Normal</td>
<td></td>
</tr>
</tbody>
</table>

Among immunologic tests, CCP (anti-cyclic citrullinated peptide) was negative for Rheumatoid arthritis, just as lupus anticoagulant test (LAC), anticardiolipin IgG-IgM, and complement C3-C4 and anti dsDNA (ELISA) sensitivity were normal.

The serologic test results showed that Anti-HIV, mono test, Brucella agglutination test, Gruber-Widal test for Salmonella were negative, Parvovirus B19 Ig G-M were normal, and Weil-Felix test for rickettsia, CMV IgM and Toxoplasma IgG-IgM were negative.

Discussion

ENT doctors must be careful while diagnosing AOSD as there are no pathognomonic signs of this disease. Yamaguchi et al. [5] reported the diagnostic criteria of AOSD as follows:

- Major criteria:
  1. Fever >39 °C, lasting one week or more
  2. Arthralgia two weeks or more
  3. Skin rash
  4. Leukocytosis with at least 80% granulocytes

- Minor criteria:
  1. Sore throat
  2. Lymphadenopathy
  3. Hepatomegaly or splenomegaly
  4. Abnormal liver function tests
  5. Negative results for antinuclear antibody and Rheumatoid factor

In our case, we eliminated lymph node malignancy, infections, and other rheumatic disorders. The case presented with 3 major criteria, namely, fever, arthralgia, and leukocytosis with 84.9% neutrophils and 4 minor criteria, including pharyngitis, lymphadenopathy in the neck and parotid gland, abnormal liver function tests (AST 254 U/L, ALT 236 U/L), and negative Anti CCP results. Ferritin level was also remarkably high (3720.39 ng/ml). This was a rare case presenting with intraparotid lymph nodes and diagnosed with AOSD. We administered 80 mg methylprednisolone per day for one week, followed by 10 mg methylprednisolone per day for one month. All symptoms (swelling on the cheek, sore throat, myalgia, arthralgia, and fever), as well as liver function tests and inflammatory parameters returned to normal. No complaints or relapse of AOSD symptoms has been observed since corticosteroid treatment.

Conclusion

Adult-onset Still’s Disease (AOSD) is rare and should be considered in patients presenting with fever, lymphadenopathy in the neck, sore throat, arthralgia, and myalgia.

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References


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