^{18}F-FDG PET-CT in adult-onset Still's disease

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DESCRIPTION

A 36-year-old Japanese woman presented with a sore throat, arthralgia, rash and fever. Physical examination revealed enlarged cervical lymph nodes with tenderness. Laboratory tests revealed a white blood cell count of 32.3×10^9/L with 98% neutrophils, a haemoglobin level of 125 g/L, a platelet count of $40.9\times10^4/\mu$L and a C reactive protein level of 15.0 mg/dL. The serum ferritin and soluble interleukin-2 receptor concentration levels were 6436 ng/mL and 947 U/mL, respectively. Individual culture results, antinuclear antibody and rheumatoid factors were all negative. CT showed generalised lymphadenopathy and hepatosplenomegaly. Positron emission tomography–CT (PET-CT) revealed increased uptake of ^{18}F-fluorodeoxyglucose (FDG) in the cervical and axillary lymph nodes, diffuse bone marrow and spleen (figure 1A,B). We performed bone marrow aspiration and a cervical lymph node biopsy to detect the presence of malignant lymphoma; however, no findings showed any signs of the disease. The lymph node histology showed reactive lymphoid tissue. We diagnosed the patient with adult-onset Still's disease (AOSD) based on Yamaguchi's criteria[1] and markedly high level of ferritin and treated her with prednisolone and methotrexate. Her symptoms improved, and her C reactive protein level normalised.

AOSD is a systemic inflammatory disease of unknown aetiology and pathogenesis characterised by fever, rash, systemic organ involvement and arthralgia. There are no specific serological markers or imaging techniques for AOSD diagnosis. Therefore, several classification criteria are currently used for AOSD diagnosis, such as Yamaguchi's,[1] Cush's[2] or Fautrel's.[3] Before reaching AOSD diagnosis, it is essential to rule out other diseases, especially malignant lymphoma as both diseases sometimes show similar clinical presentations.[4]

PET-CT can detect and diagnose many malignancies, and indications for PET-CT have increased rapidly. Some reports on patients with AOSD showed FDG accumulation in the spleen, bone marrow, lymph nodes, skin, joints, parotid glands or serous effusions.[5–7] Although the definitive diagnosis cannot be made solely based on PET-CT findings in a case with suspected AOSD, these findings are useful to support the diagnosis and evaluation of disease activity.[5–7] However, in some cases, the imaging findings of PET-CT in AOSD are similar to those observed in malignant lymphoma, which makes differential diagnosis difficult. Maximum bone marrow uptake in AOSD is reportedly lower than that in malignant lymphoma[5]; however, this trend has not been confirmed. In such cases, a biopsy may be indispensable for the diagnosis.

Figure 1 Positron emission tomography–CT demonstrated an increased ^{18}F-fluorodeoxyglucose uptake among the cervical and axillary lymph nodes (A) as well as diffuse bone marrow (A, B) and spleen (A, B).

Although PET-CT cannot directly determine the diagnosis of AOSD, it plays important roles in

Learning points

- Adult-onset Still's disease (AOSD) is diagnosed by classification criteria and by the elimination of other diseases, especially malignant lymphoma.
- The characteristic findings of positron emission tomography–CT (PET-CT) in patients with AOSD show increased ^{18}F-fluorodeoxyglucose accumulation in the spleen, bone marrow and lymph nodes.
- PET-CT cannot directly determine the diagnosis of AOSD; however, PET-CT plays important roles in evaluating the involved lesions and guiding the biopsy of the involved organs.

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evaluating the involved lesions and guiding the biopsy of the lymph nodes, bone marrow or other tissues.

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