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EDİTÖRE MEKTUP / LETTER TO THE EDITOR

Castleman's disease mimicking acute rheumatic fever

Akut romatizmal ateşi taklit eden Castleman hastalığı

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To the Editor;

Castleman's disease was first reported by Castleman et al. in 1954 and identified as an uncommon lymphoproliferative disorder[1]. It may present with many systemic findings such as fever, weight loss, arthralgia, hepatosplenomegaly, skin rashes, and high sedimentation. Due to the wide variety of symptoms, it is important to keep CD in mind in differential diagnosis. In this article, we presented a case who was mistakenly diagnosed with acute rheumatic fever.

A 6-year-old girl presented with fever and joint pain. There was a history of tonsillitis and antibiotic use 2 weeks ago. On physical examination, she had arthralgia in both knee joints and a 2/6 systolic murmur in the mitral valve area. In blood tests performed, erythrocyte sedimentation rate (ESR)= 59 mm/hour, C-reactive protein (CRP)= 95mg/dL, anti-streptolysin O (ASO)= 1000 Todd/unit was found. Complete blood count (CBC), serum electrolytes were normal. Mild mitral regurgitation was detected on echocardiography. She was hospitalized with a diagnosis of acute rheumatic fever according to modified Jones criteria (There were two major and one minor criteria in addition to the supporting findings for high-risk countries). Benzathine penicillin and aspirin treatment was initiated at the appropriate dose.

No atypical cells were found in the peripheral blood smear. Other serological tests were performed for differential diagnosis. Brucella agglutination test, HBs Ag, anti-CMV, anti-EBV, anti-HCV, anti-HAV, anti-HIV IgG and IgM tests were negative. Serum complement levels, rheumatologic and autoimmune disease markers were normal.

On the 7th day of admission, some examinations were repeated due to the findings of the patient still not improving. Repeated abdominal USG revealed a 29x26 mm hypoechoic solid mass under the pacreatic head. Magnetic resonance imaging revealed a 36x30 mm annular, well-circumscribed mass. (Fig 1) The mass was surgically excised. A nodular mass of 3.8 cm in greatest dimension was excised. Cut surface of the lesion was solid and yellowish. H&E stained sections of the lesion revealed a lymph node with lymphoid folicule hyperplasia of which were characterised with regresive, vascularised, hypocellular germinal centers and enlarged mantle zones. Enlarged secondary follicule organisation was evident focally. Interfolicular vascular proliferation and plamacytosis was observed. Immunohistochemically plasma cells were politypical for kappa and lambda light chains. Immunohistochemistry for HHV8 was performed and found to be negative. (Fig 2)

The patient was discharged and followed up after the clinical findings improved in a few days after surgery. Castleman's disease (CD), also known as angiofollicular lymph node hyperplasia, who defined a 40-year-old man with a mediastinal mass characterized histologically by lymph node hyperplasia and follicles with small, hyalinized foci[1]. In addition, salivary glands, lung, pancreas, larynx, parotid gland, meninges and even limb muscles can

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be involved lymphatically in the body. In our case, the affected region was inferior to the pancreatic head. Because of its diverse manifestations and ability to affect any body region, CD is a great mimic of both benign and malignant abnormalities in the neck, chest, abdomen, and pelvis[2].

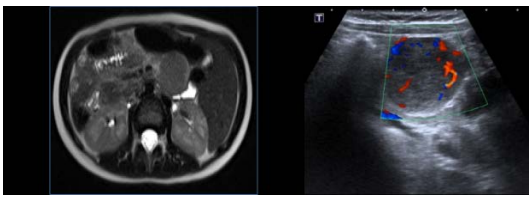

Figure1.USG ve MRI.

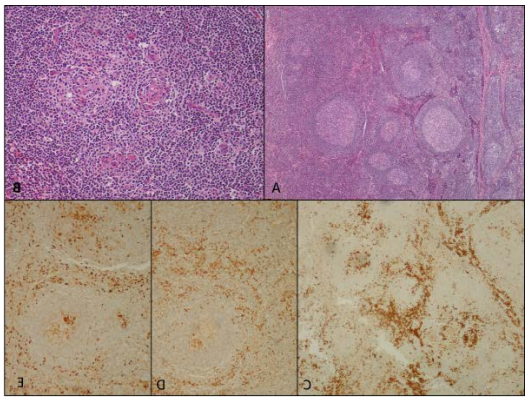

Figure 2. Pathology

In addition, although CD is not a cancerous disease, it may accompany malignant cases such as Kaposi's sarcoma, non-Hodgkin lymphoma, POEMS (Polyneuropathy, Organomegaly, Endocrinopathy, M protein, and Skin changes) syndrome, paraneoplastic pemphigus, and plasma cell dyscrasias[3].

Keller et al., classified CD histologically as a hyaline vascular and plasma cell variant, but rarely, features of the two types can coexist[4]. Most of the patients with localized (unicentric) disease are of the hyaline vascular type, and most of the patients with multicentric disease are of the plasma cell type. HHV-8 positive cases show different histological features (plasmablastic variant)[5]. In our case, although the disease showed unicentric type involvement, histologically less common plasma cell variant was observed.

The pathogenesis of CD has not been fully elucidated, but the role of IL-6 (Interleukin-6) in unicentric disease and IL-6 and HHV-8 in multicentric disease has been well described[6]. IL-6 play major role in the pathogenesis by stimulating a widespread inflammatory response that results in systemic manifestations. The increase of IL-6 stimulates the production of various acute phase reactants in the liver. This condition causes constitutional symptoms and laboratory abnormalities such as anemia, high inflammatory markers, hypergammaglobulinemia, hypoalbuminemia, etc. In our case, there was an increase in acute phase reactants that persisted despite anti-inflammatory therapy until surgical excision was achieved.

The definitive diagnosis of CD is usually made by excisional biopsy and histopathological evaluation. Especially in hyaline vascular type CD, there is a risk of bleeding due to its vascularity in case of biopsy with radiographic image. Therefore, surgical intervention is recommended[7]. In our case, since the mass was located retroperitoneally, the diagnosis was made by surgical lymph node excision with laparotomy.

In terms of imaging, generally well-circumscribed soft tissue mass is seen on CT and calcification is not common. The hyaline vascular type shows more contrast due to being a hypervascular tumor. Arteries feeding the vascular tumor, such as the bronchial, internal mammarial or intercostal, can be demonstrated by angiography[8]. Enlarged lymph nodes are seen as solid on MRI imaging. Compared to muscles, the signal is isointense or hyperintense in T1-weighted series, and slightly hyperintense in T2-weighted series[9]. The mass in our case was observed as round, well-circumscribed on MRI.

Multicentric systemic CD is usually symptomatic, and there is no consensus on its treatment yet. The most common treatment combination is surgical excision, corticosteroid and chemotherapy. However, the prognosis of the multicentric form is poor despite treatment. The localized form is often asymptomatic and can be detected in routine radiographs. Depending on the location and size of the mass, it may show compression findings such as pain and dyspnea. Surgical excision is a curative treatment[10]. In our case, there was unicentric lymph node involvement, rapid clinical improvement was observed, and no recurrence developed during follow-up.

Castleman's disease can mimic many different

diseases because it can have a wide variety of symptoms. In our case, we tried to treat it as acute rheumatic fever disease, which is quite common in our country. Castleman's disease should be considered in the differential diagnosis of many diseases, especially due to its constitutional symptoms.

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