Erdheim-Chester Disease with Chest Wall Involvement

Sir,

Erdheim-Chester Disease (ECD) is a rare systemic disease characterised by multifocal infiltration of histiocytes in bones with or without involvement of multiple organs. Clinical presentations of ECD vary from bone involvement to life-threatening multisystemic disease. Its pathogenesis is still unclear, but it is characterised by clonal expansion of histiocytes. Symptoms and signs of ECD depend on the location and extent of such involvement. The underlying cause of ECD is unclear. It is common between the ages of 40-60 years. Here, we present a patient who was diagnosed with ECD, as a result of chest wall involvement.

In the physical examination of a 41-year male patient with a complaint of chest pain admitted to our polyclinic, a mass on the chest wall was found. An expansion was detected in the right 4th rib of the patient in chest computed tomography (CT) (Figure 1). The patient was operated and the lesion was excised 4 cm wide proximally and distally. Histopathological examination showed distinctive infiltration of ECD. The patient was discharged on the third postoperative day without any complication.

ECD described by Jakob Erdheim and William Chester in 1930, is a rare inflammatory disease with unknown etiology, characterised by organ infiltration of non-Langerhans foamy histiocytes (CD68 +, CD1a- negative sign) (Figure 2).[1,2] Depending on the distribution of lesions, the clinical manifestations of EDC are varied. The disease primarily involves long bones (especially the distal femur, proximal tibia and fibula), but can involve all organs and tissues. According to the involved organ, it has a spectrum of presentations ranging from asymptomatic to life-threatening illness. Bone involvement is present in 96% of patients but symptomatic in 50%. More than 50% of cases have non-skeletal internal organ involvement.[3] Correspondingly, exophthalmos, papilledema, xanthelesma and papulonodular skin lesions, diabetes insipidus, severe pulmonary diseases, renal insufficiency, retroperitoneal involvement, cardiomyopathy, and disorders due to central nervous system involvement (pyramidal disorders, congnitive impairment, headache, *etc*.) are seen.[3] Treatment should be multidisciplinary depending on clinical involvement.

Patients with chest tumor usually present with palpable mass. Asymptomatic patients are generally diagnosed on imaging studies during examinations, usually for unrelated reasons. Based on the growth of the tumor, the symptoms develop due to invasion of adjacent structures. If invasion occurs in the neurological structures, paresthesiae and loss of strength occur. Symptoms such as fever, fatigue, weight loss may be seen. Chest X-ray, CT chest and magnetic resonance imaging (MRI) are used in the diagnosis. The exact diagnosis is made by histopathologic examination. There are no universally accepted guidelines for the diagnosis and treatment of ECD. Chemotherapy and/or radiotherapy is performed according to the pathological diagnosis after surgical resection. ECD is diagnosed upon a thorough clinical evaluation, a detailed patient history of symptoms, and high clinical surpicion in addition to specialised tests.

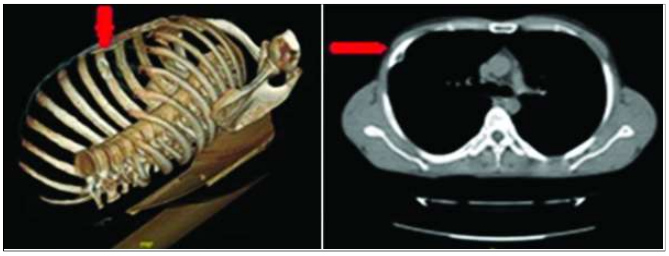

Figure 1: Preoperative 3D-CT and 2D-CT.

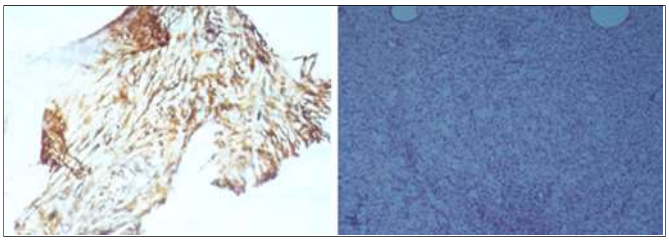

Figure 2: Typical histological finding; SMA infiltration and foamy histiocytes.

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