Mediastinal lipomatosis: as a rare complication of systemic steroids in an adolescent with lupus nephritis

To the Editor,

Mediastinal lipomatosis (ML) is a rare condition caused by excessive fat accumulation in the mediastinum (1). It is frequently related to endogenous or iatrogenic Cushing syndrome or obesity (2,3). Patients with mediastinal lipomatosis may be asymptomatic or may present with clinical findings which are caused by excessive fat accumulation and may lead to severe compression findings. The most common radiologic finding is bilateral enlargement in the mediastinum on postero-anterior chest graphy and can be confused with mediastinal masses (1). The definite diagnosis is made by computarized tomography (CT) or magnetic resonance imaging (MR) (4,5). Steroid related ML generally has a good prognosis and improves when the dose of steroid is decreased or steroid treatment is discontinued. However, patients with severe compression findings who need surgical intervention have been reported in the literature (5,6). We wanted to draw attention to a rare complication of systemic steroid use with this case.

A 17-year-old female patient who was diagnosed as systemic lupus erythematosus (SLE) and whose renal biopsy was compatible with stage IV diffuse membranoproliferative glomerulonephritis had received multiple intravenous high dose methylprednisolone, intravenous cyclophosphamide and oral prednisolone and azathioprine/mycophenolate mofetil (MMF) treatments because of active disease. When she presented for follow-up visit, she was using 40 mg prednisolone and 1000 mg MMF. On physical examination, she appeared fallen and pale. Moon face, buffalo hump and pretibial edema were present. Her blood pressure was found to be 150/100 mmHg, her pulse rate was found to be 86/min and her respiratory rate was found to be 20/min. She had no peripheral lymphadenopathy or organomegaly. A 2/6 systolic murmur was heard at the mesocardiac area. Other system examinations were found to be normal. Laboratory tests revealed a deep anemia (hemoglobin: 6.5 g/dL, haptoglobin: <28.5 mg/dL), hyoalbuminemia (serum albumin: 2.6 g/dL), proteinuria (urinary protein: 2.1 g/day) and renal dysfunction (serum creatinine: 2.2 mg/dL). On antero-posterior chest graphy bilateral mediastinal enlargement was observed (Figure 1). Echocardiography revealed pericardial effusion (6 mm) and hypertension secondary to left ventricular hypertrophy findings. Thoracic CT (without contrast) was performed with a suspicious of mediastinal mass and widespread fat accumulation was found (Figure 2). No mediastinal adenomegaly or mass was observed. Steroid-related ML was considered in the patient who had a history of long-term use of high dose steroid and the dose of steroid was reduced up to 10 mg, but steroid treatment could not be discontinued, since the disease had an active course. On the

Figure 1. Bilateral enlargement with regular borders in the upper mediastinum on antero-posterior chest graphy
follow-up graphy taken 4 weeks later enlargement in the mediastinum was observed to be regressed.

Mediastinal lipomatosis is a rare complication of systemic steroids and was described by Koerner and Sun (1) in 1966 for the first time. As far as we know, steroid-related ML has been reported only in one pediatric subject until the present time (7). While some patients have no clinical findings, some others may have severe compression findings related to airways, vena cava superior, heart and other organs (6,8,9,10). Our patient had no clinical finding suggesting compression of ML. However, echocardiography of our patient revealed pericardial effusion. When the cases in the literature were evaluated, it was found that Hsu et al. (11) reported a case of pericardial effusion related to ML. However, we think pericardial effusion can not be explained with ML alone in our patient who was being followed up with a diagnosis of SLE and who had hypoalbuminemia.

The diagnosis of mediastinal lipomatosis is generally made by observation of mediastinal enlargement on chest graphy and demonstration of intensity changes related to fat on CT or MR imaging. In our patient, mediastinal enlargement was observed initially and CT was performed considering secondary malignancy because of a history of long-term and intensive immunosuppressive treatment. A diagnosis of ML was made by demonstrating widespread fat mass on CT. ML was thought to be related to steroid in our patient who was known to have used long-term steroid and who had clinical findings related to steroid use (moon face and buffalo hump). Regression of mediastinal enlargement with reduction of steroid dose supported the diagnosis.

Conclusively, ML is a rare complication which should be considered in the differential diagnosis in patients who have a history of systemic steroid usage and mediastinal enlargement.

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