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Antiphospholipid syndrome and Langerhans cell histiocytosis: exceptional association

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The antiphospholipid syndrome (APLS) is a thrombotic disorder characterised by recurrent arterial or venous thrombosis and/or pregnancy complications of fetal loss, pre-eclampsia, or eclampsia in the presence of antiphospholipid antibodies (1). The syndrome can be primary or secondary to several conditions such as lupus erythematous, malignancies, infectious diseases, drugs or even to other autoimmune diseases (2, 3). However, the association between APLS and Langerhans cell histiocytosis (LCH) has not been reported in the literature. The aim of our work is to report an exceptional case of APLS in a 37 year old man with concomitant pulmonary LCH.

Case report

A 32 year old man was admitted to hospital with dyspnea. He was a chronic smoker since the last 15 years. He had non previous medical problems. Two days before admission, he presented chest pain and dyspnea. Physical examination showed tachypnea with respiratory rate of 38 cycles per minute, a finger clubbing, heart rate of 115 beats per minute and blood pressure was 110/60 mmHg. The cutaneous and neurological examination was normal. Arterial blood gas objectified a PaO2: 57 mmHg, PaCO2: 27 mmHg, SaO2: 91, 7%. White blood cell count was 6.5 x 109 /l and hemoglobin level 147 g/l. Renal and liver function were normal. Electrocardiography showed signs of acute right heart failure. Chest X-ray showed predominantly micronodular shadows distributed over both lungs, some of them showing cavitations (Fig.1). Sputum analysis did not show any pyogenic organisms or acid-fast bacilli and no malignant cells were evident. HRCT chest showed thrombosis of the right pulmonary artery, dilated right heart cavities and multiple thin-walled cysts mainly in the upper and mid-zones (Fig.2, 3). All these findings were typical of pulmonary Langerhans histiocytosis X associated with pulmonary embolism. Bronchoalveolar lavage has set evidence a rate of CD1a cells to 5%. Lung biopsy was advised but it was deferred because of the excessive surgical risk entailed. Prednisone (1 mg/kg/day) was initiated because of dyspnea and the poor respiratory function, associated with anticoagulant treatment. Three months later, the patient was admitted for acute respiratory failure. Physical examination showed edema of inferior members. Arterial blood gas analysis showed hypoxemia: PaO2= 50, 5 mmHg, PaCO2=30, 9 mmHg and oxygen saturation 85%. Laboratory findings revealed a normal WBC count (5800 GB/ mm3) and normal CRP level (1 mg/ ml). Chest X-ray has not shown a pneumothorax or pleural effusion. HRCT chest showed lower right segmental lobar pulmonary embolism. An etiologic assessment was then performed. Tests for antinuclear antibodies (ANA), anti-dsDNA were negative. The levels of complements (C3) and (C4) and the activated partial thromboplastin time (aPTT) were normal. The levels of antithrombin-3 and Protein C and S were in the normal ranges. Tests for factor-V-Leiden and MTHFR mutations were normal. Lupus anticoagulant was negative. The level of anticardiolipin (ACA) IgG was 30 UGPL (N < 10), indicating positivity, with another positive sample at an interval greater than 12 weeks. Prolonged anticoagulant treatment is then indicated to the association of antiphospholipid syndrome and recurrent pulmonary embolism. For his pulmonary LCH, patient received 6 months corticosteroids associated with oxygen therapy at home but without improvement of his respiratory condition. Cytotoxics treatments were not advised because of the absence of other sites of disease. Currently, he is candidate for heart-lung transplantation.

Figure 1 : Chest X-ray PA view showing micronodular shadows distributed in the all zones of lung

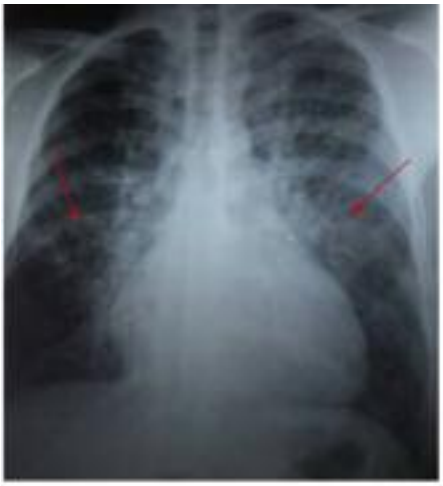


Figure 2 : An axial section of a CT scan of the chest revealed a multiple irregular thin-walled cystic with varying size and form in the upper zones.

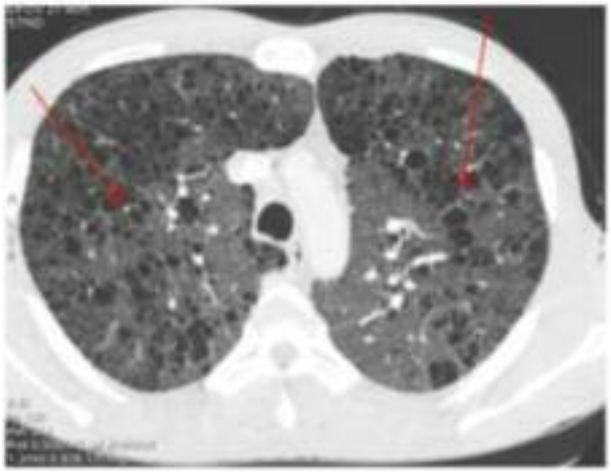
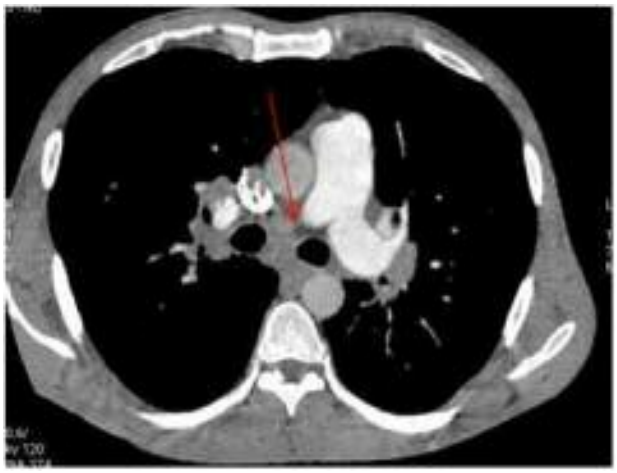


Figure 3 : An axial section of a CT scan of the chest showing thrombosis of the right pulmonary artery.



Conclusion

Antiphospholipid syndrome (APLS) is usually primary, but can be associated with a wide range of other conditions such as systemic lupus erythematosus, Sjögren's syndrome, mixed connective tissue disease (MCTD), idiopathic inflammatory myopathies, infectious diseases, malignancies, and drug-induced conditions. However, the association between APLS and pulmonary LCH has not been described in the literature.

The pathology of pulmonary LCH is poorly understood, but granulomas rich in LCs, eosinophils, macrophages and lymphocytes develop in and destroy distal bronchioles as a result of a cell-mediated immune response. APLS also has an immunological basis related to antiphospholipid antibodies and/or phospholipid binding proteins. In addition, APLS and LCH are multisystem diseases which may affect multiple organs.

Thus, the questions to be answered are whether it is a coincidental association or the two diseases are secondary to similar pathogenic mechanisms and if this association worsens the prognosis of LCH.

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Small cell carcinoma of the ovary of the hypercalcemia type In children

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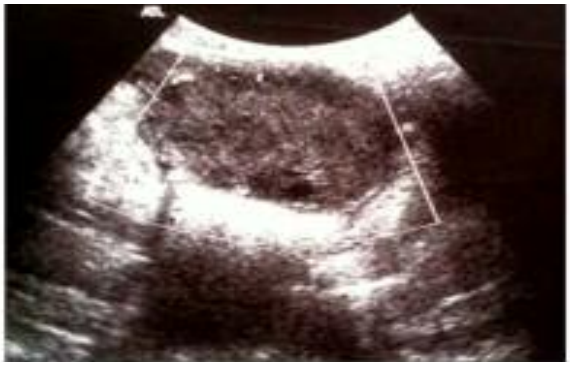
Small cell carcinoma of the ovary of the hypercalcemia type (SCCOHT) is uncommon and aggressive ovarian tumor that primarily affects young women and rarely observed in premenstrual adolescents. Histologically, the typical pattern is diffuse follicle-like sheets of small, closely packed cells with scant cytoplasm. As in epithelial ovarian carcinomas, CA-125 could be a useful marker (1-3). Hypercalcaemia is present at the time of diagnosis in more than 60% of cases with SCCOHT (2-4). We report a case of this unusual tumor whose diagnosis had not been suspected before surgery and we point out its evolution and its management.

Case report

A 10-year-old girl was admitted to our department for acute abdominal pain. She had had an intermittent and isolated pelvic pain for 6 months. A history of weight loss could not be confirmed. On examination, the pelvis was tender without fever or any palpable mass. No features of precocious puberty were noted. Ultrasound examination showed a 6-cm solid right ovarian mass and ascites (figure 1).

Figure 1 : Ultrasound examination showed a 6-cm solid right ovarian mass.



An emergency laparotomy was performed revealing a necrotic right ovary due to torsion of an ovarian solid tumor. The left ovary looked normal and neither ascites nor adhesion was detected in the abdominal cavity. A right salpingo-oophorectomy was therefore performed. Histological and immunophenotypical studies concluded that it was a small-cell carcinoma of the ovary of the hypercalcemic type (figure 2). Subsequent laboratory tests revealed hypercalcemia (2,8 mmol/l). HCG, -FP and serum carcinoembryonic antigen (CA) 125 were normal. Chest x-ray, radionuclide bone scan and cerebral CT scan were normal. The postoperative course was uneventful. In view of the highly malignant form of the tumor, the patient was given 6 courses of chemotherapy based on vinblastine, bleomycin and cisplatin. Seven years after the initial surgery, the patient was free of recurrent disease (normal calcium level, negative staging including chest radiography and abdominal ultrasound).