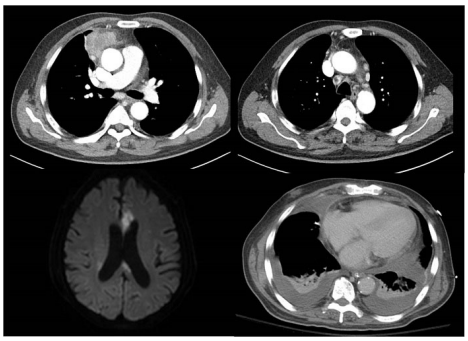
■ Sat-381 ■

A Case of Multicentric Castleman Disease Diagnosed After Resection of Concurrent Thymic Carcinoma

울산대학교 의과대학 서울아산병원 혈액내과

*박준홍, 박한승

Multicentric Castleman disease (MCD) is a group of rare lymphoproliferative disorders of plasma cells that present with systemic manifestations such as fever, lymphadenopathies, pleural, pericardial effusions, and ascites. We report a rare case of MCD diagnosed after resection of concurrent thymic carcinoma that presented with recurrent cerebral infarction. Previously healthy 67-year-old male visited our emergency department for generalized abdominal pain, fever, and malaise. His complete blood count and chemistry profile revealed no specific findings other than elevated C-reactive protein and alkaline phosphatase level. Chest CT scan revealed a 5.5-cm anterior mediastinal mass and multiple lymphadenopathies. A few days later, the patient had a sudden onset of right hemiparesis due to multifocal acute cerebral infarct, followed by rapid progression of acute kidney injury, peripheral edema, ascites, and progressive thrombocytopenia. Hemodialysis was required for adequate volume control. In suspicion of paraneoplastic syndrome, surgical removal of the thymic mass was performed. After the surgical resection, however, the patient did not recover consciousness due to recurrent cerebral infarction, and thrombocytopenia was further aggravated to 24,000/μL. MCD was suspected by frozen section, and glucocorticoid treatment (methylprednisolone IV 60mg/day) was given for five days, which resulted in no significant improvement. The thymic mass revealed thymic carcinoma(B3); however, the lymph node specimens were diagnosed with MCD with plasma cell variant, with negative serum and urine immunofixation. Treatment using anti-IL-6 antibody Siltuximab was started, which was given every three weeks. Multiorgan manifestations gradually improved, and by the third cycle of Siltuximab treatment, the patient showed marked alleviation of thrombocytopenia, ascites, fever, azotemia, and neurologic symptoms. The patient eventually weaned off from hemodialysis and platelet transfusion. This unique case featured thymic mass, multiple lymph node enlargement, unexplained thrombocytopenia, fever, azotemia, and ascites. Siltuximab treatment was highly effective for this case of MCD.



■ Sat-382 ■

A rare case of adult T-cell leukemia/lymphoma without lymphadenopathy or hepatosplenomegaly

인하대병원 의과대학 내과학교실

*조상용, 임주한, 이문희, 조진현

Introduction: Adult T-cell leukemia/lymphoma (ATLL) is uncommon lymphoid neoplasm that occurs on patients with human T-lymphotropic virus, type I (HTLV-I) infection. The clinical features of ATLL include presence of lymphadenopathy, hepatosplenomegaly, atypical lymphocytosis, hypercalcemia, lytic bone lesions, and skin lesions. Especially in acute variant, which is the most common and highly aggressive subtype of ATLL, lymphadenopathy is seen in almost all cases. Here, we report a case of acute type ATLL patient without presence of lymphadenopathy or hepatosplenomegaly. **Case:** A 47-year-old woman was admitted due to fever. She had no skin lesion or palpable mass on physical examination. Laboratory evaluation revealed atypical lymphocytosis (65,660/uL), normal hemoglobin (14.3g/dL), normal platelet counts (294,000/uL), elevated lactate dehydrogenase (502 IU/L) and hypercalcemia (14.5mg/dL). No lymphadenopathy, lytic bone lesion or hepatosplenomegaly was detected on chest and abdominopelvic computed tomography scans. Positron emission tomography showed no abnormal FDG uptake on whole body. Peripheral blood smear showed medium sized lymphocytes with multi lobulated nuclei which were similar morphology to the so-called “flower cells” (Figure A). Bone marrow study also showed abnormal nucleated lymphoid cells which were positive for CD3, CD4 and negative for CD2, CD8(Figure B). Eventually, anti-HTLV-I antibody was detected in her blood and the diagnosis of ATLL was confirmed. Despite of treatment with anthracycline based chemotherapy (CHOP, hyper-CVAD), she died of CMV pneumonia 6 months after diagnosis. **Conclusion:** We reported a case of ATLL in a 47-year-old Korean woman with atypical clinical feature. This case reminds clinicians to be aware that ATLL could occur without presence of lymphadenopathy or hepatosplenomegaly

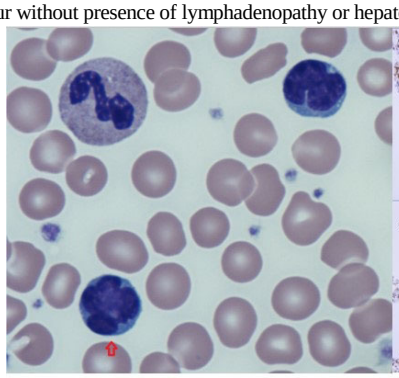


Figure A. Blood cell morphology, Flower cell

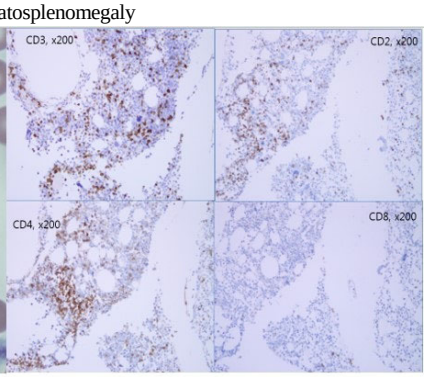


Figure B. Bone marrow Biopsy