

## CASE REPORT ΕΝΔΙΑΦΕΡΟΥΣΑ ΠΕΡΙΠΤΩΣΗ

# Spontaneous tumor lysis syndrome in a patient with angioimmunoblastic lymphoma A rare oncological emergency

Angioimmunoblastic T-cell lymphoma (AITL) is a rare form of non-Hodgkin lymphoma characterized by an aggressive clinical course with fever, generalized lymphadenopathy, hepatosplenomegaly, anemia, hypergammaglobulinemia and auto-immune-like manifestations. The case is presented of a 60-year-old male patient with AITL who eventually died due to spontaneous tumor lysis syndrome (STL). Vigorous fluid administration with rasburicase and even renal replacement therapy may prove to be life-saving in this clinical setting. STL must be included in the differential diagnosis of acute kidney injury among patients with angioimmunoblastic lymphoma. It may be lethal, as in this patient.

Angioimmunoblastic T-cell lymphoma (AITL) is a rare form of non-Hodgkin lymphoma accounting for approximately 2% of non-Hodgkin lymphomas and up to 20% of peripheral T-cell lymphomas.<sup>1,2</sup> It is characterized by an aggressive clinical course with fever, generalized lymphadenopathy, hepatosplenomegaly, anemia, hypergammaglobulinemia and auto-immune-like manifestations.<sup>2,3</sup> The case is presented of a 60-year-old male patient who presented with fever of unknown origin, was diagnosed as having AITL and eventually died due to spontaneous tumor lysis syndrome (TLS).

### CASE PRESENTATION

A 60-year-old male patient presented with a fever of 38 °C without rigors, but with a skin rash, fatigue, and cervical, maxillary, epitrochlear and inguinal lymphadenopathy. His past medical history was unremarkable except for smoking. Abdominal and thoracic computed tomography (CT) revealed hepatosplenomegaly and mesothoracic and intra-abdominal lymphadenopathy. Excision

biopsy of a right inguinal lymph node showed histopathology and immunochemistry compatible with AITL. While awaiting the biopsy results the patient developed acute kidney injury (AKI) with marked elevation of the serum levels of potassium (7.46 mmol/L), uric acid (16.9 mg/dL), phosphorus (8.82 g/dL) and creatinine (8.81 mg/dL). Hypergammaglobulinemia (gamma globulin 7.18 g/dL) and anemia (Ht: 32.5%) were detected, and the white blood cell count (WBC) was  $48,360 \times 10^3/\mu\text{L}$ . The patient died despite intravenous (iv) administration of rasburicase and fluid and renal replacement therapy.

### DISCUSSION

AITL is a lymphoid malignancy, which usually displays an aggressive clinical course and poor response to conventional chemotherapy. Studies found an overall 5-year survival of 40%.<sup>4-6</sup> A standard approach for treating patients with AITL has not yet been established due to the lack of a large series of patients because of the rarity of this clinical entity. The treatment options include steroids, immuno-

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Αυτόματο σύνδρομο  
λύσης όγκου σε ασθενή με  
αγγειοανοσοβλαστικό λέμφωμα

Περίληψη στο τέλος του άρθρου

### Key words

Acute kidney injury  
Angioimmunoblastic lymphoma  
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modulators, alemtuzumab, bortezomib, rituximab, single-agent cytotoxic drugs and combination chemotherapy. Among these, anthracycline-based chemotherapy has been recommended as first line therapy, while more intensive chemotherapeutic regimens, consolidation with autologous stem-cell transplantation, and even allogeneic stem-cell transplantation have been documented to be effective.<sup>7-10</sup> There is little evidence from randomized controlled trials, however, that these forms of treatment have improved the survival of patients with AITL.

Several prognostic indices, including age (>60 years), raised WBC, anemia, thrombocytopenia, mediastinal lymphadenopathy and increased IgA, are suggested to be associated with poor prognosis and overall survival.<sup>11</sup> Of these, this patient had an elevated WBC ( $48,360 \times 10^3/\mu\text{L}$ ), anemia (Ht: 32.5%), thrombocytopenia ( $134 \times 10^3/\mu\text{L}$ ), mediastinal lymphadenopathy and high IgA (728 mg/dL).

On the other hand, TLS is the most common oncological emergency, resulting from the process of the rapid lysis of tumor cells and leading to AKI, arrhythmia and death. Central to its pathogenesis is the rapid accumulation of uric acid derived from the breakdown of nucleic acids, which leads to kidney failure by various mechanisms. Kidney failure then limits the clearance of potassium, phosphorus, and uric

acid resulting in hyperkalemia, hyperphosphatemia, and hypocalcemia. Although the rapid release of electrolytes from intracellular stores to the extracellular space may have fatal consequences, homeostatic mechanisms can usually compensate for these shifts, provided that kidney function remains robust. AKI is therefore central to the development of TLS; it is often caused by acute uric acid nephropathy (from metabolism of liberated nucleic acids), but it may also be mediated by uric acid-independent mechanisms, including parenchymal and tubular deposition of calcium-phosphate salts.<sup>12</sup>

TLS is typically seen during the initial chemotherapeutic treatment of hematological malignancies,<sup>12</sup> but spontaneous TLS, occurring without any treatment, is being increasingly recognized worldwide, especially in leukemias and Burkitt's lymphoma.<sup>13-15</sup>

To our knowledge, the occurrence of spontaneous TLS has not been previously described in angioimmunoblastic lymphoma. Spontaneous TLS should be included in the differential diagnosis of AKI among patients with angioimmunoblastic lymphoma. Rigorous administration of iv fluids, together with rasburicase, and renal replacement therapy as salvage therapy may possibly prove to be life-saving in these circumstances.

## ΠΕΡΙΛΗΨΗ

### Αυτόματο σύνδρομο λύσης όγκου σε ασθενή με αγγειοανοσοβλαστικό λέμφωμα

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Το αγγειοανοσοβλαστικό λέμφωμα είναι μια σπάνια μορφή non-Hodgkin λεμφώματος, που χαρακτηρίζεται από επιθετική κλινική πορεία με πυρετό, γενικευμένη λεμφαδενοπάθεια, ηπατοσπληνομεγαλία, αναιμία, υπεργαμμασφαιριναιμία και αυτοάνοσες εκδηλώσεις. Στο παρόν άρθρο περιγράφεται η περίπτωση άνδρα ασθενούς, ηλικίας 60 ετών, ο οποίος διαγνώστηκε με αγγειοανοσοβλαστικό λέμφωμα και που τελικά απεβίωσε λόγω αυτόματου συνδρόμου λύσης όγκου. Η επιθετική ενυδάτωση σε συνδυασμό με τη χορήγηση ρασπουρικής και ακόμη και η αιμοκάθαρση, μπορεί να αποβούν σωτήριες. Το αυτόματο σύνδρομο λύσης όγκου πρέπει να περιλαμβάνεται στη διαφορική διάγνωση σε περιπτώσεις ασθενών με αγγειοανοσοβλαστικό λέμφωμα και οξεία νεφρική βλάβη και μπορεί να είναι θανατηφόρο, όπως στην περίπτωση του ασθενούς μας.

**Λέξεις ευρητηρίου:** Αγγειοανοσοβλαστικό λέμφωμα, Αυτόματο σύνδρομο λύσης όγκου, Οξεία νεφρική βλάβη

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