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Case Report

The Constant Mimic Disease: Angioimmunoblastic T-cell Lymphoma, A Mimic for DRESS Syndrome

Álex García Tellado[1], Pablo Alija Piret[1], Mercedes De la Fuente Vázquez[1], Carlos Manuel Bercebal Vega[2], Santiago Montes Moreno[2], María Teresa Valiente[1] and Nuria Puente Ruiz[3*]

[1]Department of Clinical Internal Medicine, Marqués de Valdecilla University Hospital, Spain

[2]Department of Anatomical Pathology, Marqués de Valdecilla University Hospital, Spain

[3]Department of Clinical Internal Medicine, Marqués de Valdecilla University Hospital, Lopez-Albo post residency program Marqués de Valdecilla University Hospital, Spain

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Corresponding author:
Nuria Puente Ruiz,
Department of Clinical Internal Medicine, Marqués de Valdecilla University Hospital. Lopez-Albo post residency program Marqués de Valdecilla University Hospital, Santander. Spain
Email: nuriapuenteruiz@scsalud.es

ABSTRACT

Introduction/Aims/Background: Angioimmunoblastic T-cell lymphoma (AITL) is an uncommon and aggressive lymphoma which is usually presented in elderly patients. The etiology of AITL is unknown, with a prevalence around 1-2% of non-Hodgkin lymphomas. It may present a broad clinical spectrum of symptoms. Fewer than 8 cases of AITL presenting a DRESS like-syndrome as first clinical features, have been reported worldwide. DRESS syndrome, a T-cell mediated hypersensitivity reaction with maculopapular exanthema, fever, internal organ involvement, and generalized lymphadenopathy, poses a challenging differential diagnosis to AITCL.

Case description: We present a rare case of a 71-year-old man admitted to the hospital University Marqués de Valdecilla in Santander, Spain, in July 2023. We diagnosed the Angioimmunoblastic T-cell lymphoma based on clinical, analytical, and histological findings.

Results: We report a case of AITL that mimicked DRESS syndrome, and it was only distinguishable by histological findings.

Discussion: This patient suffered from a rare and extensive systemic affection with characteristic primary lesions in the form erythroderma that responded to corticosteroids. It is known that exposure to a drug, mostly antibiotics, has been chronologically associated with the onset of the disease in at least one third of the cases, being the DRESS syndrome crucial to be dismissed.

Conclusion: AITL is a heterogenous disease that must be considered in the differential diagnosis of a patient with fever, generalized lymphadenopathy, eosinophilia, and maculopapular exanthema.

Patient consent: We have the patient's consent to publish the patients' photographs presented in the text below.

Learning points:

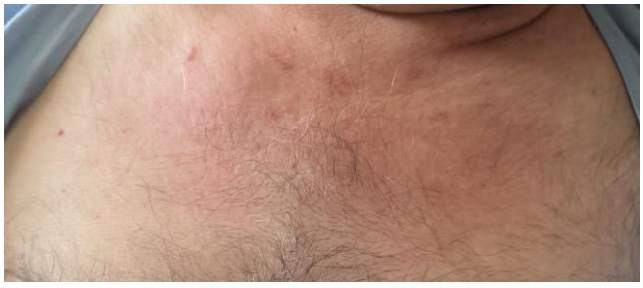
- Angioimmunoblastic T-cell lymphoma is a heterogenous disease that has to be considered in the differential diagnosis of a patient with fever, generalized lymphadenopathy, eosinophilia, and maculopapular exanthema.

- It is known that exposure to a drug, mostly antibiotics, has been chronologically associated with the onset of the disease in at least one third of the cases.
- DRESS syndrome, a T-cell mediated hypersensitivity reaction with maculopapular exanthema, fever, internal organ involvement, and generalized lymphadenopathy, poses a challenging differential diagnosis to AITCL. It is crucial a complete anamnesis and physical examination to establish a correct suspicion of this mimicking syndrome.

CASE REPORT

A 71-year-old man native from Spain was admitted from the emergency department after developing over the last 2 weeks a maculopapular exanthema on the chest, superior members and back. His medical history included a recent diagnosis of a drug induced cutaneous reaction by NSAIDs, that was followed by the Allergology department, as well as an antiphospholipid syndrome and a classic migraine both with ambulatory control. His usual treatment was a recently exposed to NSAID, as well as a chronical treatment with an angiotensin II antagonist.

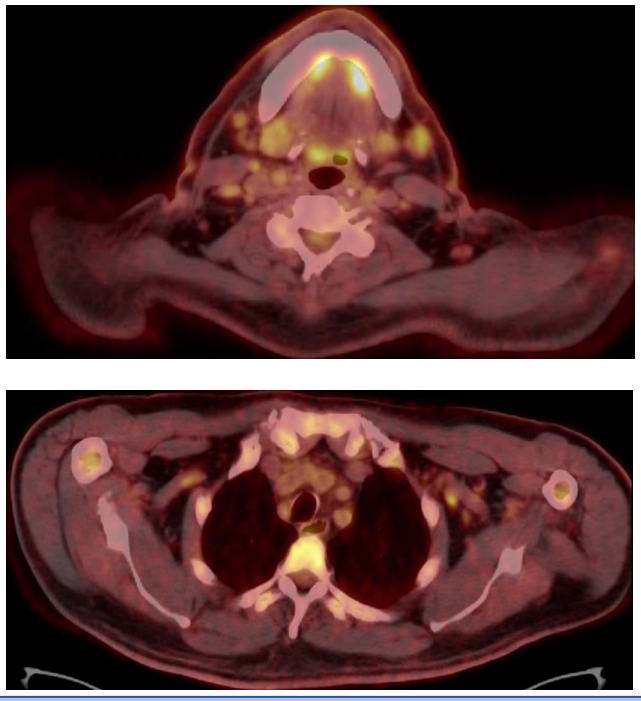
At the physical examination he presented a painless cutaneous rash consisting in a pruriginous erythroderma with non-desquamative primary lesions. The lesions were rapidly progressing, with a distribution mostly in chest, back and shoulders, having a complete resolution during the hospital admission **(Panel A)**. We also notice the presence of supraclavicular and axillary adenopathy's, unnoticed for the patient, with B symptoms consisting in a loss ponder of 6 Kilos as well as a profuse nocturnal sweating in the last 6 months.



Panel A. Chest. Pruriginous and rapidly progresive erytroderma with non-desquamative primary lesions.

Laboratory tests showed minimal elevation of acute phase reactants (Protein C reactive 2.3 mg/dl and a sedimentation rate of 10mm). We conducted a complete microbiological study (including VHH6) and carried out some autoimmune markers (including ANAs, ANCAs and ENA screening), with all the results negative. A proteinogram found a remarkable oligoclonal pattern with an elevated Beta-2-Microglobulin (9.5mg/dl).

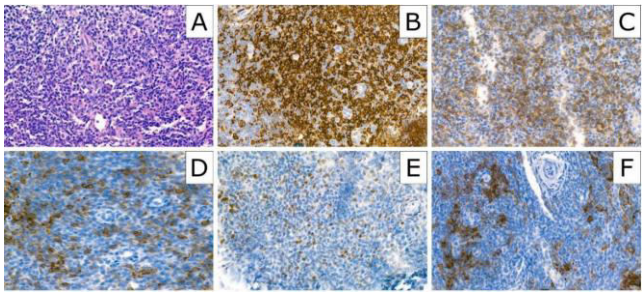
A PET-CT scan was conducted to rule out the presence of an occult tumor. The scan revealed bilateral lymphadenopathies in the cervical, axillary, retropectoral, and right paratracheal regions, as well as in the bone marrow of the vertebral column and pelvis. These areas exhibited hypermetabolic activity, which prompts consideration for a differential diagnosis involving a lymphoproliferative syndrome. **(Panel B)**.



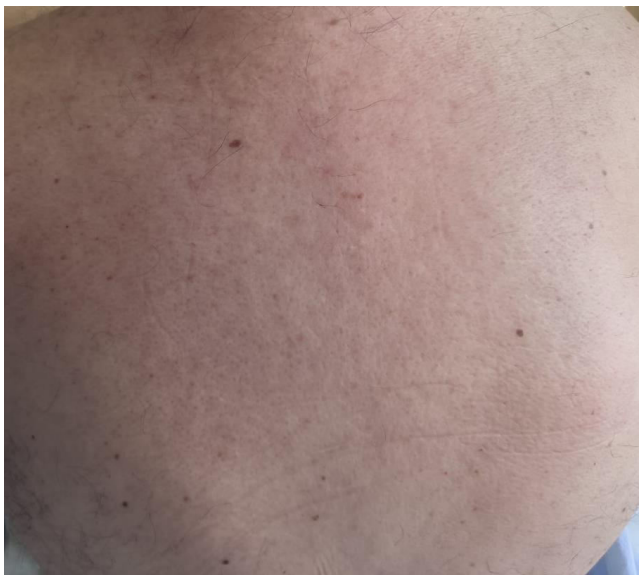
Panel B. PET Study with FDG-F18: Small lymph nodes and some hypermetabolic adenopathy bilaterally in the cervical, bilateral axillary, left retropectoral, right paratracheal, and bone marrow of the vertebral column.

By this time, the patient accomplished five out of the six clinical criteria for the DRESS syndrome, except for the possibility of another alternative diagnosis that could explain this clinical presentation. Because of this, we performed a lymphadenopathy biopsy to rule out the possibility of lymphoproliferative disorder. The anatomopathological findings were compatible with an angioimmunoblastic T-cell lymphoma, showing a proliferation of medium-sized round cells with clear cytoplasm and minimal cytological atypia **(Panel C)**. Once the correct diagnosis was established, intermediate corticosteroids doses were initiated, with an excellent response

and practical disappearance of the skin lesions (**Panel D**) and clinical improvement.



Panel C. HEMATOXYLIN-EOSIN (x40). A): Proliferation of medium-sized round cells, clear cytoplasm, and minimal cytological atypia. Neoplastic cells aggregate in nests around high-endothelial venules, against a polymorphous inflammatory background. IMMUNOHISTOCHEMISTRY (x40): B) CD3 +. C) CD4 +. D) PD1 +. E) BCL6 +. F) CD23 +. Note the striking expansion of follicular dendritic cells forming a mesh with an extrafollicular pattern and surrounding high-endothelial venules.



Panel D. Back. Complete resolution of the erytroderma lesions during hospital admittion.

DISCUSSION

This patient suffered from a rare and extensive systemic affection that can mimic multiples diseases, such as the DRESS syndrome. It is known that exposure to a drug, mostly antibiotics, has been chronologically associated with the onset of the disease in at least one third of the cases, being the DRESS syndrome crucial to be dismissed. This toxicoderma is a complex multisystemic hypersensitivity whose diagnosis and management are troublesome, being crucial the correct identification of the culprit drug to ensure a correct clinical approach. It is known that the most frequently drugs are antibiotics, Alopurinol and antiepileptics, but it is also associated to NSAID, as it was suspected in our patient.

To establish a correct diagnosis for a DRESS syndrome, not only the clinical history has to be probably but also it is obliged to dismiss a lymphoproliferative disorder or an autoimmune disease, like it has been demonstrated in the limited bibliography of this disease, with a completed radiological examination (E.g., lymphadenopathies) and a histological study. It has been registered in the literature 4 types of histological patterns consisting of reactive lymphoid hyperplasia, necrotizing lymphadenopathy, necrotizing lymphadenitis, Hodgkin lymphoma-like and AITL-like. In our case the patient presented a pattern compatible with an AITL-like except for the immunohistochemical confirmation for AITL, consisting in the confirmation of positivity to CD3 and BCL6, typical findings in this kind of tumors (**Panel C**).

CONCLUSIONS

Angioimmunoblastic T-cell lymphoma is a rare but aggressive entity primarily affecting T-cells within the lymphatic system. Its diagnosis and treatment pose a challenge, requiring careful evaluation and multidisciplinary management. Ongoing research is essential to improve our understanding of this disease and develop more effective therapeutic approaches.

CONFLICTS OF INTEREST

The authors declare any conflict of interest.

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