Case Report

Annals of Clinical Reviews and Case Reports

Cutaneous Presentation of Diffuse Large B-cell Lymphoma

Ana F. Batista*, Sara Fontaínhas, Sara Faria, Sónia C. Pereira, Amélia Pereira

Serviço de Medicina Interna, Hospital Distrital da Figueira da Foz, Figueira da Foz, Portugal

***Corresponding author:** Ana F. Batista, Institution: Serviço de Medicina Interna, Hospital Distrital da Figueira da Foz, Figueira da Foz, Portugal. Telephone: +351914300453; E-mail: filipa_batista_@hotmail.com

Citation: Batista AF, Fontaínhas S, Faria S, Pereira SC, Pereira A (2022) Cutaneous Presentation of Diffuse Large B-cell Lymphoma. Annal Clin Revie Cas Repor: ACRCR-107.

Received Date: December 17th, 2022; **Accepted Date:** December 20th, 2022; **Published Date:** December 26th, 2022

Abstract

Dermatologic paraneoplastic syndromes may appear before, after or simultaneously with the diagnosis of malignant neoplasms. They are particularly important because, from a practical point of view, changes involving the skin are more quickly identified at an early stage. Ultimately, this may determine a greater probability of both diagnosis and treatment initiation, with consequent improvement in survival, although the most common scenario associates these lesions with advanced stages of the disease.

The authors present the case of an 85-year-old man with sudden onset of purpura-like skin lesions on the lower limbs, gastrointestinal symptoms and increased immunoglobulin A, compatible with Henoch-Schönlein's purpura, which was interpreted as a paraneoplastic manifestation of Diffuse Large B-Cell Lymphoma. The clinical presentation was exuberant, with generalized adenopathic conglomerade, significant clinical status deterioration and death after 2 months.

Keywords:

Diffuse large B-cell lymphoma; *Henoch-Schönlein* purpura; Paraneoplastic syndrome; Vasculitis

Case Report

The authors present the case of an 85-year-old man, with a history of type 2 diabetes mellitus and benign prostatic hyperplasia, chronically medicated with metformin and finasteride, who went to the Emergency Department due to the acute onset of non-pruritic nor painful skin lesions on the lower limbs, showing cephalic progression, first noticed 3 days before. There were also recent complaints of weight loss (4Kg in the previous week), anorexia, postprandial infarction and nausea, medicated with metoclopramide, already discontinued, despite all symptoms persisted. There were no other B symptoms, myalgia, arthralgia, respiratory or urinary complaints.

On physical examination, he was in good general condition and apyretic. Palpable purpuric and petechial lesions were present on the lower limbs (Figure. 1), already spreading to the upper limbs, with different colorations.

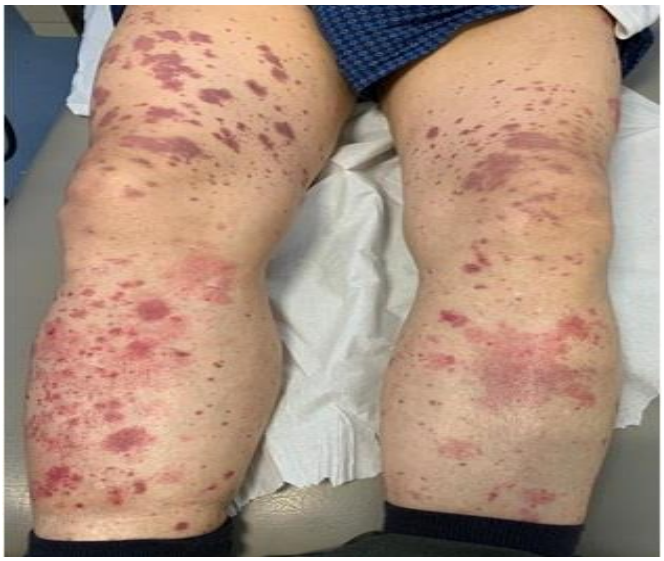

Figure 1: Petechial and purpuric lesions at first medical evaluation.

Analytically, the blood count showed lymphopenia [Lymphocytes 0.53x10^3/µL (N 1.5-3.5)], without anemia [Hemoglobin 14.2 g/dL (N 13.5-18.0)] or platelet count changes. Velocity sedimentation rate and C-reactive protein were within normal reference ranges, 9 mm/1h (N<20) and 4.11mg/L (N<5), respectively. Serological study (HBV, HCV, HIV, CMV and EBV) and immunological study were both negative. Urine sediment examination was not active. Immunoglobulin A (IgA) [733 mg/dL (N 70-400)] was increased, with a normal protein electrophoresis and serum immunofixation. Free light chains in urine showed abnormal results [kappa chain 297 mg/L (N <32.9); lambda chain

103.6 mg/dL (N <3.8)], with a monoclonal lambda band in urinary immunofixation. Chest X-ray had no alterations.

He was discharged on prednisolone, at a dose of 0.5mg/Kg, with a favorable response only after a few days (Figure. 2).

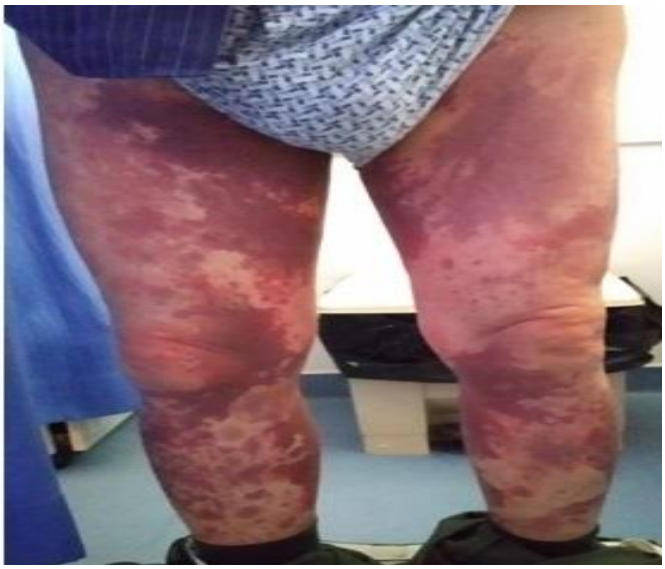

Figure 2: Image of skin lesions in the most active phase.

A thoraco abdominopelvic computerized axial tomography was performed, which was negative for the presence of adenopathies, expansive lesions or organomegalies. The skin biopsy showed a local, perivascular, low-intensity inflammatory infiltrate, consisting of lymphomonocytic cells and perivascular extravasation of red blood cells, and was done with the patient under corticosteroid therapy for the previous 2 weeks.

One month after the first medical contact, despite the partial regression of the skin lesions (Figure. 3), the patient developed multiple and painless cervical, axillary and inguinal bilateral adenopathic conglomerates, which temporally correlated with steroid weaning.

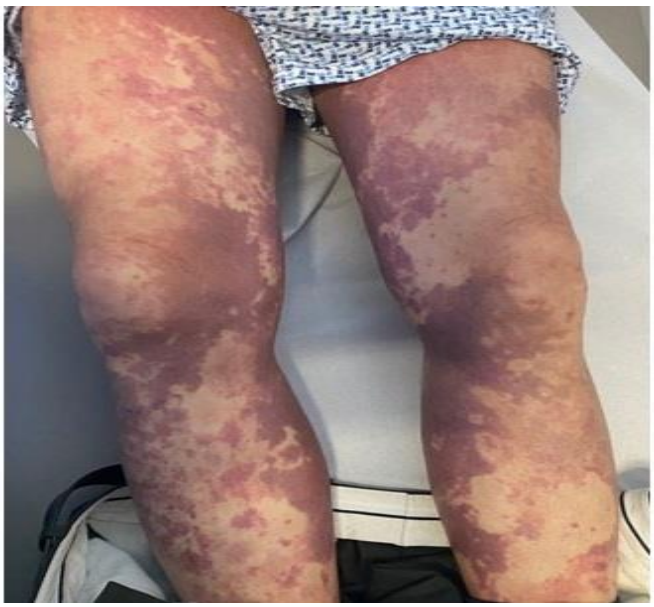

Figure 3: Image of skin lesions showing partial regression.

In view of the generalized adenopathic scenario, an excisional biopsy of one of the inguinal adenopathies was performed, resulting in the diagnosis of Diffuse Large B-Cell Lymphoma (LDGCB), rich in T-lymphocytes.

The subsequent evolution was characterized by an abrupt clinical decline, development of a tumor lysis syndrome and ultimately the patient's death within 2 months, after the initial evaluation, leaving no time for other treatment options, other than the use of corticoids.

Discussion

Although rare in the elderly and with a greater probability of atypical and/or incomplete presentations, the diagnosis of Henoch-Schönlein purpura, which is clinical, was early considered during the diagnostic process [1]. In this specific case, the purpura-like skin lesions, of which Figures 1 to 3 are examples, the concomitant gastrointestinal symptoms and the increase in serum IgA were the key elements – the typical clinical manifestation consists of the tetrad: palpable purpura, abdominal pain, gastrointestinal bleeding, arthritis/myalgias and kidney failure [2]. The observed purpuric and petechial lesions showed a bilateral and symmetrical distribution, predominantly on the lower limbs, as would be expected [3].

The role of skin biopsy, which is decisive when the clinical findings or the atypical/incomplete presentation does not allow the diagnosis to be made, was in this case negatively influenced by the already long evolution of the lesions and the corticosteroid therapy initiated, although in small vessel vasculitis specific anatomopathological alterations may not always be present (the most typical finding is leukocytoclastic vasculitis [1-3]).

Considering the main known triggering factors, it was possible to rule out secondary causes of vasculitis, such as drug-related (chronic and recently introduced medication had no known association with vasculitis) and also infectious causes (lack of focal symptoms, except for gastrointestinal complaints, sustained apyrexia, the absence of elevated inflammatory markers and the negativity of the microbiological/serological study). At this stage, the fact that there was no renal dysfunction conducted the treatment towards symptom control, evidence supporting the use of for abdominal complaints. The focus would then be the exclusion of neoplastic causes, for which the correlation with Henoch-Schönlein purpura is highly probable, only in these age groups [1-3]. The quick development of generalized adenopathies was a strong clue, making the excisional biopsy the next mandatory step to be taken.

With regard to the final diagnosis of LDGCB rich in T lymphocytes, it is important to clarify that it is a rare subtype, with a diagnostic peak in the 4th decade of life, a slight male predominance and is associated with a very aggressive clinical course, of which this case report is an example [4]. The initial presentation was characterized by a paraneoplastic vasculitis, progressed to the development of generalized adenopathic conflomerades and terminated with the patient's death over the course of 2 months. The

rapid onset and aggressiveness of the lymphoma did not allow another type of therapy, in addition to corticosteroid therapy used with the purpose of reducing the size of the adenopathies.

Conclusion

The recognition of typical cutaneous manifestations of vasculitis with a possible association with malignancy can be decisive in the diagnosis and effective treatment of neoplasms, although most of the time these signs only appear when the disease is already in an advanced or incurable stage [5,6]. In the elderly in particular, its study should aim at the exclusion of this secondary cause and should be one of the main differential diagnoses to be considered. One should remember that the investigation should be persued despite normal skin biopsy results because, in most cases, it is unusual for the pathologist to be able to make a diagnosis based only on skin biopsy findings, which are generally nonspecific, showing identical characteristics, regardless the connection between vasculitis and neoplasia is established or not [6].

The presence of both neoplasm and Henoch-Schönlein purpura, which is rare in the adult population, prevails in males, over the age of 60 years and is associated with poor prognosis [2,7,8]. The cases described in the literature are mostly related to solid tumors (mainly lung cancer), but the representation is quite diverse, including hematologic cancers. Therefore, the evaluation of occult neoplasia is suggested in cases of unexplained Henoch-Schönlein purpura and the search for metastatic disease should be done in patients with this type of purpura and known neoplastic history [2,7,8].

References

1. Natalia S., Jonathan Z., Janak Z., Lymphoma presenting as Henoch–Schönlein purpura. Clinical Kidney Journal. 2012; 5: 600–2. doi.org/10.1093/ckj/sfs128
2. Therese M., Dayna F., Jefferson R., Adult-Onset Henoch-Schonlein Purpura associated with a Metastatic Malignancy of Unknown Primary Origin. Hawaii J Med Public Health. 2018; 77: 243–5.
3. Jagpal K., Aneet K., Latha A., A rare cause of rash in elderly: diagnostic dilemma, BMJ Case Reports. 2014. doi.org/10.1136/bcr-2014-205601
4. Olutayo S., Rachael S., Eugene L., T cell/histiocyte rich B-cell lymphoma: A difficult diagnosis to make, Current Problems in Cancer: Case Reports. 2020; 2. doi.org/10.1016/j.cpccr.2020.100041.
5. Zurada JM, Ward KM, Grossman ME. Henoch-Schönlein purpura associated with malignancy in adults. J Am Acad Dermatol. 2006; 55: 65-70. doi.org/10.1016/j.jaad.2005.10.011.
6. Joshua P., David W., Mark P., David W., Henoch-Schönlein Purpura Associated with Solid-organ Malignancies: Three Case Reports and a Literature Review, Acta Derm Venereol 2012; 92: 388–9. doi.org/10.2340/00015555-1288.
7. Josenilson S., Kleyton M., Ana I., Isabeççа L., Aline F., Sandra O., et al, Paraneoplastic cutaneous manifestations: concept and updates, A Bras Dermatol. 2013; 88: 9–22. doi.org/10.1590/S0365-05962013000100001
8. Goldman-Cecil 26th Edition, Chapter 169: Approach to the patient with cancer – Dermatologic Paraneoplastic Syndromes, 1166-95.