CASE REPORT

Primary mediastinal (thymic) B-cell lymphoma presenting as cutaneous vasculitis

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SUMMARY

Cutaneous small vessel vasculitis is a severe neutrophilic vascular inflammation mediated by immune complexes that involves the dermal postcapillary venules.

Neoplasms represent 2–5% of all causes of secondary cutaneous vasculitis. We present a case of a 52-year-old man who was admitted due to a 10-day history of respiratory symptoms and myalgia. From the third day of symptoms onwards, the patient noticed the appearance of cutaneous lesions in the lower limbs with palpable purpura and erythematous papules. Additionally, he reported of asthenia, anorexia and weight loss during the prior month. Chest radiography showed an enlarged mediastinum and thoracoabdominal-pelvic CT scan revealed a bulky left hilar mass. Biopsy of the left superior lobar bronchus' mucosa allowed for the histological diagnosis of diffuse large B-cell lymphoma (DLBL) of the thymus. The patient received symptomatic treatment with improvement of the purpuric lesions and a multiagent chemotherapy regimen was initiated.

BACKGROUND

Although this is a rare pathology, it is of great importance to recognise that cutaneous small vessel vasculitis can be the first sign of neoplasms, particularly those of haematological origin, where we include lymphoproliferative disorders.

CASE PRESENTATION

A 52-year-old man was admitted due to a 10-day history of respiratory symptoms (cough, fever, dyspnoea) and myalgia unresponsive to non-steroidal anti-inflammatory drugs. From the third day of symptoms onwards, the patient noticed the appearance of cutaneous lesions in the lower limbs, sparing the plantar regions. These lesions resembled non-pruriginous palpable purpura and erythematous papules that progressively spread to the forearms and lower abdomen. Additionally, he reported of asthenia, anorexia and weight loss during the prior month. There were no new drugs, including antibiotics, administered during that period. Apart from being a smoker, the patient had no relevant medical history.

INVESTIGATIONS

On clinical examination, we found several cutaneous lesions on the lower limbs, with palpable purpura and multiple vesicles with serous content ([figure 1](#)); the remaining physical examination was normal. Laboratory values were normal except for elevated C reactive protein (14 mg/dL, normal value <0.5 mg/dL). Chest radiography showed an enlarged

Figure 1 Exuberant vasculitis lesions of the right ankle with palpable purpura and vesicles.

mediastinum ([figure 2](#)) and thoracoabdominal-pelvic CT scan revealed a heterogeneous, bulky, left hilar mass with secondary narrowing of the ipsilateral main bronchus and its smaller branches ([figure 3](#)). The tumour invaded the main mediastinal vessels and multiple thoracic, mediastinal and abdominal lymphadenopathies were also present. The mediastinal mass conditioned tracheal and oesophageal deviation to the contralateral side. Bronchofibroscopy confirmed bronchial mucosa infiltration and marked narrowing of the left main bronchus; cytological and microbiological tests were negative, including for neoplastic cells. Punch skin biopsy showed neutrophilic infiltrate with fibrinoid necrosis of blood vessel walls and extravasation of red blood cells consistent with leucocytoclastic vasculitis ([figure 4A, B](#)). Bone marrow aspiration study showed no evidence of abnormal lymphoid infiltration. HIV 1 and 2 serologies were negative. Biopsy of the left superior lobar bronchus mucosa allowed for the histological diagnosis of diffuse large B-cell lymphoma of the thymus (CD79a positive).

Figure 2 Chest radiography: enlarged mediastinum.

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Figure 3 Thoracoabdominal-pelvic CT scan: heterogeneous, bulky, left hilar mass with secondary narrowing of the ipsilateral main bronchus and its smaller branches.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis focuses on the presence of mediastinal mass in a patient with constitutional symptoms and cutaneous vasculitis. Thus we included: thymoma, lymphoma, germ cell tumours or even lung carcinoma as possible diagnostics.

Figure 4 (A and B) Punch skin biopsy showing neutrophilic infiltrate with fibrinoid necrosis of blood vessel walls and extravasation of red blood cells.

Figure 5 (A and B) Several cutaneous lesions on the lower limbs, with palpable purpura and multiple vesicles, with serous content, before (A) and after treatment (B).

TREATMENT

The patient received symptomatic treatment with topic β -methasone and loratadine with improvement of the purpuric lesions (figure 5A, B). As soon as the diagnosis of DLBL was made, a multiagent chemotherapy regimen, R-CHOP (rituximab, cyclophosphamide, hydroxydaunorubicin, oncovin and prednisolone), was initiated.

OUTCOME AND FOLLOW-UP

At present, our patient is still followed by a haematologist and continues to fulfil the chemotherapy regimen initially established. We found a regression of vasculitic lesions of the lower limbs and a significant improvement of respiratory symptoms on admission throughout the treatment period.

DISCUSSION

Cutaneous small vessel vasculitis (CSVV) is a severe neutrophilic vascular inflammation that involves the dermal postcapillary venules. According to the aetiology, CSVV is most frequently classified as primary or idiopathic, but can also be secondary to other processes such as infection or inflammatory diseases, or may be drug-related. Only 2–5% of cases are associated with neoplasms, most commonly plasma cell dyscrasias, myeloproliferative or lymphoproliferative disorders and, less commonly, solid organ carcinomas. The diagnosis of CSVV is obtained by skin biopsy with involvement of small vessels and a histopathological result of leucocytoclastic vasculitis: perivascular

infiltration by polymorphonuclear leucocytes, extravasation of erythrocytes and fibrinoid necrosis of the vascular wall.^{1–6}

The pathogenic association between vasculitis and malignant neoplasms, especially of haematological origin, has not been clarified. CSVV seems to be an immune complex-mediated process caused by an antigen excess that deposits on postcapillary venules and activates complement, as well as the inflammation cascade leading to vascular wall damage.^{2–3–6} The trigger is the excess of circulating antigens that seems to be tumour-associated;^{2–7} despite this, other causes such as infections and drug-exposure should be considered in patients with cancer.^{2–4–8–9} On the other hand, some neoplasms are associated with increased blood viscosity leading to endothelial damage and extensive deposition of immune complexes.²

The cutaneous manifestations can appear previously, simultaneously, or after the diagnosis of malignancy, most frequently as palpable purpura of the lower limbs.^{3–4} If there is a history of solid organ malignancy, a new episode of cutaneous vasculitis may be linked to recurrence of the disease.² There is no evidence that the timing of cutaneous lesion appearance has a prognostic value.⁵ Prognosis seems to be more dependent on the underlying neoplasm than the presence of CSVV.^{3–9}

Learning points

- ▶ Cutaneous small vessel vasculitis (CSVV) typically presents with palpable purpura of the lower limbs.
- ▶ CSVV is most frequently classified as primary or idiopathic but can also be secondary to other processes such as infection or inflammatory diseases, or may be drug-related.
- ▶ Two to five per cent of all CSVV cases are associated with neoplasms, most commonly of haematological origin.

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Contributors ADSL wrote the first draft of the manuscript and rewrote new drafts based on input from coauthors, and was an integral part of the medical team responsible for the patient during his hospitalisation, doing so under the tutelage of HE, who gave input on manuscript drafts. LS was involved in all stages of drafting and reviewed the manuscript. AC was responsible for final review and approval of the submitted manuscript. All authors read and approved the final manuscript.

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REFERENCES

- 1 Fox LP, Shinkai K. Cutaneous vasculitis. In: Bologna JL, ed. *Dermatology*. Vol 2. 3rd edn. Elsevier, 2012:385–91; chapter 24.
- 2 Podjasek JO, Wetter DA, Pittelkow MR, et al. Cutaneous small-vessel vasculitis associated with solid organ malignancies: the Mayo Clinic experience, 1996 to 2009. *J Am Acad Dermatol* 2012;66:e55–65.
- 3 Das M, Chhabra R, Hinton SW. Cutaneous leukocytoclastic vasculitis and myelodysplastic syndrome with little or no evidence of associated autoimmune disorders—a case report and a brief review of the literature. *Am J Med Sci* 2008;336:368–71.
- 4 Carlson JA, Chen KR. Cutaneous vasculitis update: small vessel neutrophilic vasculitis syndromes. *Am J Dermatopathol* 2006;28:486–506.
- 5 Beylot J, Malou M, Doutre MS, et al. Leukocytoclastic vasculitis and malignant hematologic diseases. (12 cases). *Rev Med Interne* 1989;10:509–14.
- 6 Pelosof LC, Gerber DE. Paraneoplastic syndromes: an approach to diagnosis and treatment. *Mayo Clin Proc* 2010;85:838–54.
- 7 Bachmeyer C, Wetterwald E, Aractingi S, et al. Cutaneous vasculitis in the course of hematologic malignancies. *Dermatology* 2005;210:8–14.
- 8 Kurzrock R, Cohen PR, Markowitz A, et al. Clinical manifestations of vasculitis in patients with solid tumors: a case report and review of the literature. *Arch Intern Med* 1994;154:334–40.
- 9 Loricera J, Calvo-Río V, Ortiz-Sanjuán F, et al. The spectrum of paraneoplastic cutaneous vasculitis in a defined population: incidence and clinical features. *Medicine (Baltimore)* 2013;92:331–43.

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