Male with myelofibrosis and ulceronecrotic lesions

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SUMMARY

Granulocytic sarcoma also called myeloid sarcoma is an extramedullary tumour of immature granulocytic cells. It is a rare entity, and mostly accompanied by acute myeloid leukaemia. It is observed during the course of myeloproliferative disorders especially in chronic myeloid leukaemia and myelodysplastic syndromes. Here, we report a case of a 60-year-old male with past history of myelofibrosis admitted to the emergency room due ulceronecrotic lesions, fever and dysphagia. We emphasize the importance of recognizing this entity and its severity.

KEYWORDS: Sarcoma, Myeloid. Leukaemia. Primary myelofibrosis.

CASE

A 60-year-old man was admitted to the emergency room due ulceronecrotic lesions, fever and dysphagia in the last 15 days. He was in follow-up of hypertension and myelofibrosis for three years without complications. On physical examination, periorbital oedema, ulceronecrotic lesions in ocular, oral and genital mucosae (Figure 1), erythematous and purplish lesions on lower limbs, toes (Figures 2 and 3) and face, cervical, axillary, and inguinal lymph node enlargement, and hepatosplenomegaly were observed. Histopathology of a skin lesion on right thigh showed perivascular inflam-
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RESUMO

O sarcoma granulocítico, também chamado de sarcoma mieloide, é um tumor extramedular de células granulocíticas imaturas. É uma entidade rara, e principalmente acompanhada de leucemia mieloide aguda. É observado durante o transtorno mieloproliferativo, especialmente na leucemia mieloide crônica e síndromes mielodisplásicas. Aqui, relatamos um caso de um homem de 60 anos com antecedente de mielofibrose admitida na sala de emergência devido a lesões ulceronecróticas, febre e disfagia. Enfatizamos a importância de reconhecer essa entidade e sua gravidade.


REFERENCES

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DISCUSSION

Granulocytic sarcoma, chloroma or extramedullary myeloid tumour, is a tumour mass of myeloblasts or immature myeloid cells that appears in extramedullary sites, especially bone tissue. It is a rare variant of myeloid neoplasia, formerly known as chloroma, because of its greenish colour, secondary to the expression of myeloperoxidase.

Cutaneous lesions in patients with myelodysplastic syndrome can be separated into non-specific (vasculitis, infections, neutrophilic dermatosis, ecchymosis, panniculitis and erythema multiform) and specific lesions defined by the presence of malignant hematopoietic cells in the skin. The occurrence of cutaneous granulocytic sarcoma in the context of myelodysplastic syndromes is rare and often a sign of poor prognosis. Clinicians must hence be aware to diagnose these lesions early because they can precede peripheral blood and bone marrow transformation to acute myelogenous leukaemia.