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PHOTO QUIZ

A 24-year-old woman with skin ulceration and strawberry gums

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CASE REPORT

A 24-year-old woman was referred by her general practitioner to the dermatologist and the internist because of a three-month history of skin ulcers on her face and right arm. She had also noted redness and swelling of her gingiva in the last four weeks (*figure 1*). She had already been treated with several oral and topical antibiotics without effect. Two weeks before she developed general malaise with low-grade fever, nose obstruction and epistaxis. She had no arthralgias or gross haematuria. On examination, she had three deep ulcerations with a purple margin on her left cheek, on her chin and on the inner side of her right upper arm. She also had erythematous swollen gingiva with petechiae looking like the surface of a strawberry. Besides the skin ulceration and mucosal lesions, examination was not remarkable. On laboratory investigation, C-reactive protein was 21 mg/l and there was a mild leucocytosis (11.9 x10^9/l). Renal and hepatic function tests and urinary sediment were normal. A chest X-ray revealed no abnormalities.

WHAT IS YOUR DIAGNOSIS?

See page 469 for the answer to this photo quiz.

Figure 1A. *Deep skin ulceration with a purple margin on the left cheek*

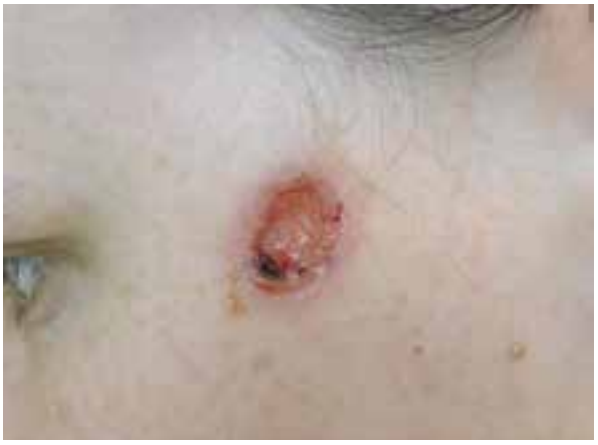


Figure 1B. *Erythematous swollen gingiva with petechiae looking like the surface of a strawberry*

ANSWER TO PHOTO QUIZ (PAGE 464)

A 24-YEAR-OLD WOMAN WITH SKIN ULCERATION AND STRAWBERRY GUMS

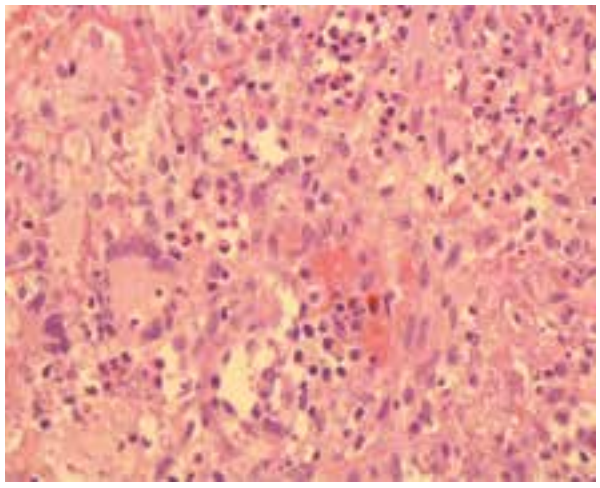
DIAGNOSIS

Granulomatosis with polyangiitis (GPA), previously known as Wegener's granulomatosis, was considered the most appropriate diagnosis based on the clinical picture. Skin biopsy of the lesion on her right arm showed active folliculitis with giant cell reaction and therefore did not contribute to the diagnosis. However, biopsy of the nasal mucosa showed extensive chronic, granulomatous, ulcerating inflammation with vasculitis (*figure 2*), confirming the diagnosis of granulomatosis with polyangiitis. In addition serum anti-PR3 (cANCA) antibodies were positive (4.9 kU/l, reference <2 kU/l) .

GPA is a rare granulomatous necrotising vasculitis of small vessels, affecting vascular structures especially of the upper airways, lungs and kidneys. Patients usually present with constitutional symptoms including fever, migratory arthralgias, malaise, anorexia, nose obstruction, dyspnoea and weight loss.[1] GPA is also known to cause mucosal lesions. Strawberry gums, as found in this patient, are very typical for GPA.[2] Skin lesions, in this case with the aspect of pyoderma gangrenosum, are usually non-specific. Other cutaneous manifestations of GPA are palpable purpura, nodules, petechiae and delayed healing of excision wounds.[3] The American College of Rheumatology criteria for the classification of Wegener's granulomatosis are abnormal urinary sediment (red cell casts or greater than five red blood cells per high power field), abnormal findings on chest radiograph (nodules, cavities, or fixed infiltrates), oral ulcers or nasal discharge, and granulomatous inflammation on biopsy. For purposes of classification, a patient is said to have Wegener's granulomatosis if at least two of these four criteria are present. The presence of any two or more criteria yields a sensitivity of 88.2% and a specificity of 92.0%.[4]

This case illustrates that recognition of the very typical strawberry gums as a manifestation of GPA may lead to an early diagnosis and treatment.

Figure 2. *HE-stained slide of nasal mucosal biopsy shows granulomatous vasculitis*



ACKNOWLEDGEMENTS

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REFERENCES

1. Harrison's Online > Part 15. Disorders of the Joints and Adjacent Tissues > Section 2. Disorders of Immune-Mediated Injury > Chapter 326. The Vasculitis Syndromes: Granulomatosis with Polyangiitis (Wegener's).
2. van Haselen CW, Hermens, FHW, Koot RAC, Smeets JHJM. M. Wegener: een ongebruikelijke klinische presentatie bij een jonge vrouw. NTvDV 2008: 18: 249-54.
3. Ruokonen H, Helve T, Arola J, Hietanen J, Lindqvist C, Hagstrom J. "Strawberry like" gingivitis being the first sign of Wegener's granulomatosis. Eur J Intern Med. 2009;20:651-3.
4. Leavitt RY, Fauci AS, Bloch DA, et al. The American College of Rheumatology 1990 criteria for the classification of Wegener's granulomatosis. Arthritis Rheum. 1990;33:1101-7.