



Facial necrotic ulcerative lesions in an adolescent female with granulomatosis with polyangiitis (GPA)

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Case presentation

A 15-year-old female was referred with forehead lesions starting 3 months before, initially treated as acne. She was also suffering from recurrent otitis media, treated with 3 courses of antibiotics over the last 45 days. Four weeks ago, she was admitted to another institution with symptoms of arthralgias, myalgias, and low-grade fever up to 37.8 °C. Her physical exam revealed ulcerative lesions of the forehead (Fig. 1) and pre-auricular area bilaterally, muscle tenderness, hemorrhagic lesions of the nasal mucosa, aphthous ulcers of the buccal mucosa, gum hemorrhagic lesions, right side perforated tympanic membrane, and otorrhea. No sinusitis, rhinitis, or chronic cough was ever reported. Blood pressure was 115/78 mmHg. Chest X-ray revealed nodular lesions and CT chest confirmed the presence of 2 large nodular lesions up to 1.5 cm of diameter as well as multiple smaller nodules (Fig. 2). The paranasal sinuses imaging revealed mastoid cells, sphenoid sinus, and the ethmoid cells filled with secretions, with demineralization of the septations of the latter. Forehead skin lesion biopsy revealed ulcerative lesions with fibroinflammatory exudate and granulomatous reaction of the dermis, but no lesions suggestive of vasculitis. Blood testing revealed WBC 11360/μL (Ne 65.5%, Ly 23%, Mo 8.1%, Eo 3.3%), Hb 11.3 g/dl, PLT 374 K/μL, CRP 91.8 mg/L, ESR 140 mm/h, Ur 24 mg/dL, Cr 0.5 mg/dL, AST 10 U/L, ALT 7 U/L, and CPK 23 U/L. Urine protein/creatinine ratio was <0.2, and

urinalysis was negative for hematuria. ANCA indirect immunofluorescence was positive (c-ANCA) with PR-3 specificity (300 U/mL) by ELISA. The diagnosis of granulomatosis with polyangiitis (GPA) was established and she was treated with IV methylprednisolone pulses (1 g × 3 days) followed by 60 mg/day with a tapering dose and rituximab 500 mg × 4 doses.

Discussion

In GPA, the prevalence of skin disease has been reported to range between 35 and 50% and can be the first manifestation in up to 8% of cases [1]. Skin involvement is reported in 48% of all pediatric patients with ANCA-associated small vessel vasculitis. In GPA most frequently includes palpable purpura and/or petechial rash (27%), subcutaneous nodules (8%), infarctions (3%), livedo (1%), Raynaud's phenomenon (3%), and subcutaneous swelling (3%) and is often polymorphic [2], very similar to the prevalence in adult studies [3, 4]. Necrotic–ulcerative lesions most commonly occur on the extremities but may also occur on the face and scalp [5]. Acneiform and folliculitis-like papules have been described in patients younger than 20 years of age, often

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Fig. 1 Ulcerative lesions of the forehead

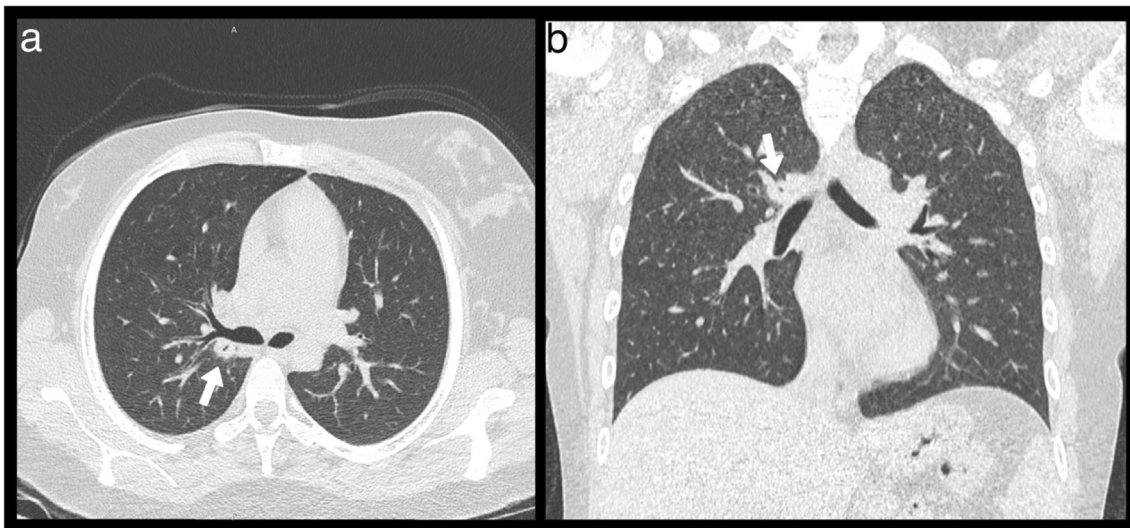


Fig. 2 a Axial and b coronal plane reconstructed CT scan images showing a cavitating pulmonary nodule underneath the right main bronchus (white arrow)

occurring as the initial presenting symptom [1]. Other than acne and folliculitis, pyoderma gangrenosum and other neutrophilic dermatosis, infections, malignancies, and systemic connective tissue disease are included in the differential diagnosis of similar type lesions [1, 5].

Compliance with ethical standards

Disclosures None.

Consent for publication We obtained written informed consent from the patient and her parents for publication of this case report together with any accompanying images.

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