A 24-year-old woman had been followed since 1984 at another institution for recurring skin lesions. The lesions were nonpruritic and would begin as papules before rupturing and becoming necrotic. An initial skin biopsy had been interpreted as being consistent with infected sebaceous cysts. The lesions persisted despite treatment. One episode of otitis media occurred. Symptoms compatible with purulent and hemorrhagic rhinitis were also elicited.

In December of 1986, the patient developed fever, cough, and hemoptysis. An infiltrate was noted in the superior segment of the left lower lobe on chest x-ray examination. Antibiotic therapy was initiated and clinical improvement was noted. A followup chest x-ray film (Fig 1) was obtained. A cavitary lesion was found in the superior segment of the left lower lobe. A repeat skin biopsy was done. The patient was referred to this institution for further evaluation.

On physical examination, the patient was afebrile with a respiratory rate of 18 and blood pressure of 108/72 mm Hg. She did not appear acutely or chronically ill. Examination of the eyes, ears, nose, mouth, pharynx showed no abnormalities. There was no organomegaly. Examination of the skin was remarkable for a 2 cm ulcerative necrotic lesion at the right temple (Fig 2). Smaller lesions were noted to be in various stages of development at the left temple, back, and chest.

Laboratory data showed a hematocrit of 38.1 percent, white blood cell count of $12.4 \times 10^9$, erythrocyte sedimentation rate of 42 mm/h. Chemistry panel result was normal. Urinalysis was normal without proteinuria or hematuria. Antinuclear antibody and rheumatoid factor were negative as was fungal serology. The patient was anergic to skin testing with tuberculin and controls.

The patient underwent bronchoscopic examination and transbronchial biopsies. No endobronchial lesion was noted, but erythema and edema were present at the orifice of the superior segment of the left lower lobe. Endobronchial and transbronchial biopsy results were negative. Bronchial washings and brushings were negative. Results of routine, fungal, and mycobacterial cultures of the washings and biopsy specimens were negative. Due to the presence of a soft tissue nodule in the left maxillary sinus, the patient underwent a left Caldwell-Luc nasal antral mucosal biopsy. Normal nasal mucosa was found. Thoracotomy was performed with a wedge resection of the superior segment of the left lower lobe.

![Figure 1. Cavitary pulmonary infiltrate in the superior segment of the left lower lobe.](image1)

![Figure 2. Ulcerative, necrotic facial skin lesion.](image2)
What is the likely diagnosis?

a) Sporotrichosis
b) Histoplasmosis
c) Systemic lupus erythematosus (SLE)
d) Sarcoidosis
e) Limited Wegener's granulomatosis
f) Blastomycosis

The answer is: e) Limited Wegener's granulomatosis

On pathologic examination of the resected lung specimen, a prominent necrotizing granulomatous process was present (Fig 3). In areas of lung away from this process were arteries surrounded by lymphoid aggregates, chronic inflammation, and fragmented elastica (Fig 4). No infective organism was identified in the special stains of the resected lung or skin biopsy. Routine, fungal, and mycobacterial cultures of the lung lesion were negative. The skin biopsy was consistent with a necrotizing vasculitis.

Wegener's granulomatosis is a disorder of unknown etiology characterized by necrotizing granulomatous inflammation and vasculitis of the upper and lower respiratory tract with glomerulonephritis. The limited form of the disorder is associated with a more benign prognosis and is notable for the absence of renal involvement. A precise etiologic agent or inciting cause has not been discovered, but the clinicopathologic features are consistent with hypersensitivity.

The majority of patients present with symptoms secondary to involvement of the upper airways. Skin rash was the initial presenting symptom in 11 of 85 patients in a series reported by Fauci et al. Skin or mucosal lesions may occur prior to, concomitantly, or after the appearance of involvement in other organs. Papules, vesicles, palpable purpura, subcutaneous nodules, and frank ulcerations may be seen. On pathologic section, papulonecrotic lesions usually show a necrotizing vasculitis with thrombosis of the vessel lumen resulting in ulceration. Necrotizing granulomatous and vasculitic involvement of the skin may be seen separately or in combination though independently as is typical of Wegener's disease. Nonspecific inflammation may be the only finding in the skin or mucosal surfaces in some cases.

Pulmonary infiltrates are common. Multiple, bilateral, nodular infiltrates with a tendency to cavitate are the most common manifestations. Parenchymal lesions may be transient, fleeting, and asymptomatic. Symptomatic lesions result in chronic cough, dyspnea, and hemoptysis. Pleural disease is infrequent. Endobronchial involvement has been described and may be associated with atelectasis on routine radiograph.

The disorder is infrequent. It can be suggested on clinical grounds when the characteristic triad of involvement is present. Fauci et al suggest that the diagnosis be made when there is evidence of involvement in two of three areas: lung, upper airways, and kidney. The diagnosis can be made in the absence of renal involvement, as is illustrated in this case. This case is similar to those reported by Fieberg and illustrative of the superficial protracted phenomenon described by him in which the mucosal and skin manifestations are of long duration and precede the development of more widespread disease. The importance in this instance and in the case presented is in the ability to make an early diagnosis and institute therapy to prevent disfiguration and prevent the occurrence of the more ominous renal disease.

Successful cytotoxic therapy with cyclophosphamide has altered the previously grim prognosis. The disorder had been almost uniformly fatal usually due to renal or progressive pulmonary disease. Long-term and complete remission have now been achieved utilizing cyclophosphamide often in conjunction with corticosteroid therapy. The patient was treated with cyclophosphamide and prednisone. After two months of therapy, there was marked improvement in the skin lesions and complete resolution of the pulmonary lesion.

Collagen vascular and fungal diseases were consid-
ered in the differential diagnosis. Fungal cultures and smears were negative. Pulmonary sporotrichosis is rare, usually affects older men, and may resemble cavitary tuberculosis. The lymphocutaneous form gains entry usually through the skin of an upper extremity. Blastomycosis, histoplasmosis, and sarcoidosis are commonly associated with mediastinal nodal enlargement. Skin lesions in blastomycosis begin as papules before becoming elevated, verrucous, ulcerated granulomas. The tongue, larynx, and oropharynx are frequently involved with ulcerative and granulomatous lesions in histoplasmosis. Erythema nodosum is common in sarcoidosis. Bluish-purple thickened elevations known as lupus pernio may be seen involving the nose, cheeks, ear lobes, knees, lips, and digits. Vasculitis is common in SLE, but not nodular or cavitating pulmonary infiltrates as part of the primary process.

References
1 Carrington CB, Liebow A. Limited forms of angitis and granulomatosis of Wegener's type. Am J Med 1966; 41:457-527
8 Amin R. Endobronchial involvement in Wegener's granulomatosis. Postgrad Med J 1983; 59:452-54