X-RAY FILM OF THE MONTH

Clinical Information

White man, age 59, entered the hospital with a huge chronic progressive ulceration of his left thigh which began about July, 1953. This was accompanied by low grade fever and weight loss. All laboratory studies, including skin tests and cultures, were non-revealing except for eosinophilia of 10 to 15 per cent. Biopsy of the thigh showed granulomatous fat necrosis. All therapeutic measures failed to control the lesion. Dyspnea appeared in December, 1953, and became progressively worse. Bronchoscopy revealed edema and ulceration in several of the large bronchi. The film below was obtained in January, 1954. The patient expired two weeks later.

ANSWER

Wegener's Syndrome

Falling into the general category of non-infectious necrotizing granulomatosis, Wegener's syndrome is basically characterized by three features: necrotizing granulomatous lesions of the upper or lower respiratory tract or both, glomerulitis, and disseminated arteritis similar to that of periarteritis nodosa. Not all features need be present to establish the diagnosis. Related, yet exhibiting certain differences, are Churg and Strauss' granulomatosis and midline lethal granuloma.

Peak age incidence is in the fourth and fifth decades. An allergic history is frequently elicited. The respiratory tract is often the site of the initial symptomatology. There may be persistent rhinitis or sinusitis which may progress to severe destruction of the midline structures of the face. Extensive granulomas of the skin occasionally occur, as in the present case. Pulmonary involvement is heralded by cough, hemoptysis, dyspnea, and chest pain. Signs and symptoms of renal disease develop later in most of the cases. Uremia is the most common mode of death. Hypertension is
not common. Constitutional manifestations such as persistent fever, weight loss, and weakness will, on occasion, be out of proportion to the local symptoms. Anemia, leukocytosis, eosinophilia, and increased sedimentation rate are often present. Almost every organ in the body can be involved by either the granulomatous process or the arteritis. Bacteriological study has been noncontributory.

Positive roentgen findings associated with involvement of the facial area include sinusitis and destruction of bone. Tomography may demonstrate laryngeal or tracheal lesions. The findings in the chest, although nonspecific, are extremely helpful in the diagnosis. Solitary or multiple nodules or infiltrates of chronic nature with central cavitation may occur, as in the present case. These roentgen findings, along with the previously described clinical picture, should suggest the diagnosis.

Almost all cases thus far reported have terminated fatally within six months to two years. No effective treatment has been found.

SELECTED REFERENCES


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