Wegener's Granulomatosis

The destructive granulomatous syndrome first described in detail by F. Wegener in 1936 and 1939[1 2] consists of three features: necrotizing, granulomatous lesions of the respiratory tract; generalized focal necrotizing vasculitis; and necrotizing glomerulitis.[3] The respiratory lesions usually affect the upper air passages, especially the nose and the paranasal sinuses. There is persistent purulent rhinorrhoea, epistaxis, and nasal obstruction succeeded by extensive destruction of the soft tissue, cartilage, and bone of the nose and palate. Great mutilation of the face and eyes may ensue. Sometimes the ears, mouth, larynx, and trachea are also affected. Disease of the lungs is less often a primary event, but is common later in the course of the disease. There is persistent cough, haemoptysis, and pleural pain, and the lung contains areas of consolidation associated with parenchymal destruction. As the condition proceeds there is a considerable constitutional reaction, and remittent fever is a noteworthy accompaniment. Proteinuria and haematuria herald the progressive renal failure that terminates in uraemia, which is the cause of death.[3]

Histologically the lesions in the respiratory tract consist of severe inflammation of the mucosa and submucosa with extensive necrosis and ulceration. There is also a reparative process, with the formation of exuberant granulation tissue containing many of the cells of chronic inflammation such as lymphocytes, plasma cells, neutrophil and eosinophil leucocytes, and especially giant cells of both the foreign body and the Langhans type. The pulmonary lesions are circumscribed masses of similar necrotizing granulation tissue, and they are of firm, rubbery consistency resembling white infarcts. A prominent feature of all these lesions is a patchy necrotizing angiitis affecting especially the smaller arteries and veins. A similar necrotizing vasculitis is found in other organs such as the spleen and kidneys, where it may form the basis of granulomatous masses of necrotic tissue. Finally, there is a widespread inflammation of the glomeruli, with fibrinoid necrosis and later proliferation of the capsular epithelium and capsular adhesions.

Wegener's granulomatosis is rare, only 138 unequivocal cases having been reported by 1967.[4] The victims are usually young or middle-aged adults of either sex who have been previously healthy, and in the great majority of cases death occurs in from one to five months. A few patients survive several years, especially if adrenal steroids are given in the early stages before severe renal damage has occurred. More recently, limited forms of the disease have been noted. Thus C. B. Carrington and A. A. Liebow described 16 cases with prominent pulmonary lesions but with absent or limited manifestations elsewhere. Of these, eight were still alive, two died of unrelated causes, and six succumbed to vascular accompaniments of the syndrome.[5] H. Pambakian and J. R. Tighe have described two women with Wegener's granulomatosis in which lesions in the breast were prominent.[6] In one woman a granulomatous mass in the breast was the presenting feature and this was followed by a similar mass in the thigh. Soon there was evidence of laryngeal and tracheal disease, and death occurred from uraemia and pneumonia seven months after the onset of the disease. The other woman developed the breast lesion in the course of the disease, which started with aural manifestations and proceeded with nasal and pulmonary involvement. She was still alive after two years, having been treated with prednisone, and there was no evidence of renal disease.

The aetiology of Wegener's granulomatosis is unknown. Its microscopical features suggest a close relationship with polyarteritis nodosa, itself a disease of unknown aetiology though usually attributed to hypersensitivity or autoimmunity. Classical polyarteritis nodosa affects medium-sized arteries and gives rise to widespread visceral infarction after the affected vessels have become occluded by thrombosis or reparative fibrosis. There is a variant called microscopical polyarteritis, which affects the smaller blood vessels, producing areas of vascular necrosis surrounded by a profuse inflammatory reaction, but there are no large necrotizing granulomata.[7] Probably, therefore, Wegener's granulomatosis is closely related to polyarteritis but is a separate disease. It should always be borne in mind in the differential diagnosis of progressive necrotizing granulomatous processes occurring anywhere in the body—and especially in the respiratory tract—where no infective agent can be incriminated.[8 9]

[1] Wegener, F., *Verhandlungen der Deutschen Pathologischen Gesellschaft*, 1936, **29**, 202.
[2] Wegener, F., *Beiträge zur pathologischen Anatomie und zur allgemeinen Pathologie*, 1939, **102**, 36.
[3] Godman, G. C., and Churg, J., *Archives of Pathology*, 1954, **58**, 533.
[4] Nielsen, K., Christiansen, I., and Jensen, E., *Acta Medica Scandinavica*, 1967, **181**, 577.
[5] Carrington, C. B., and Liebow, A. A., *American Journal of Medicine*, 1966, **41**, 497.
[6] Pambakian, H., and Tighe, J. R., *Journal of Clinical Pathology*, 1971, **24**, 343.
[7] Rose, G. A., and Spencer, H., *Quarterly Journal of Medicine*, 1957, **26**, 43.
[8] Walton, E. W., *British Medical Journal*, 1958, **2**, 265.
[9] Elsner, B., and Harper, F. B., *Archives of Pathology*, 1969, **87**, 544.

World Medical Assembly

The World Medical Association, of which the B.M.A. is a founder member, meets in Ottawa this year from 12 to 18 September for its 25th annual Assembly. The hosts are the Canadian Medical Association, and at the opening session Dr. Ole Harlem, of Norway, the present president of the W.M.A., will install a Canadian, Dr. A. F. W. Peart, as its new president. A special attraction will be the two sessions arranged by the scientific programme committee on "The Use of Physician Assistants in Primary Health Care," with contributors from the U.S.A., U.S.S.R., Ghana, and Canada, and "Medical Care and Treatment of Psychotropic Drug Problem Patients." The latter discussion will centre on the socioeconomic aspects and on national and international control programmes. The business sessions, which have been remodelled to do away with the cumbersome "reference committee" procedure, will deal with such subjects as medical ethics; medical education—specially topical with the prospect next year of the Fourth World Conference on Medical Education in Copenhagen on the theme "Educating Tomorrow's Doctors;" professional liability; and sociomedical affairs. A meeting of medical editors, now a regular and useful feature of the W.M.A. Assemblies, will discuss the relevance to medical journals of techniques used by the lay press. The business and scientific sessions are open to all physicians, and the Canadian Medical Association, through its president, Dr. H. D. Roberts, extends a warm welcome to all doctors attending the Assembly.[1]

[1] *Canadian Medical Association Journal*, 1971, **105**, 198.