

Fiftieth anniversary

Wegener and Wegener's granulomatosis

Wegener and his background

This year is the 50th anniversary of the publication in 1937 of Wegener's classic paper "On Generalised Septic Vessel Diseases" in the *Proceedings of the German Pathology Association*. To commemorate this anniversary and the 80th birthday of Dr Friedrich Wegener we are publishing the first English translation of the original German article in *Thorax*.

Friedrich Wegener was born on 7 April 1907 in Varel, Oldenburg.¹ His father, Dr Friedrich Wegener, was a physician and surgeon at St Josefsstift Hospital in Varel and his mother, Thyra Cecilia Thyden, a Swedish gymnastic director. Wegener, who was educated in Varel, Wilhelmshaven, and Jever, showed a flair for languages, studying Greek, Latin, and English, and he now speaks Swedish fluently. He was a natural sportsman and was German hammer throwing champion in 1931. After high school Wegener studied botany and zoology, but when in 1926 his father felt that he should be studying something more productive he became a medical student in Munich. He completed his studies in Kiel in 1932 and qualified as MD in 1934 with a thesis entitled *Testicular Tumour*.

Wegener was attracted to the idea that pathology formed the basis of medicine and on 15 March 1933 he became assistant at the Pathological Institute of the University of Kiel under Professor L Jores, R Hueckel, and Martin Staemmler. With the foundation of the Lübeck Medical Academy, Wegener became professor of dissection and teacher of anatomy and histology, a post he held from 1966 to 1969. When he withdrew from the Academy in 1970 he was honoured by a torchlight procession led by the medical students and physicians of Lübeck. He was made "Doctor Honoris Causae" in 1976 by the medical school of Lübeck and retired completely from medicine in 1979. Wegener saw 12 cases of Wegener's granulomatosis during his working life, though he never used that eponym.

The discovery of Wegener's granulomatosis

In January 1934 Wegener saw a 38 year old lorry driver with ulceration of the mouth and stomach, who then developed a saddle nose deformity and deafness.

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This was followed by a pneumonic illness and renal failure and the patient died in June of that year. Histological examination showed a generalised angitis and multiple necrotic granulomas affecting the nose, trachea, lung, kidney, and spleen and a diagnosis of unexplained septic disease was made. Wegener was sure that this was a new condition, clinically and pathologically distinct from periarteritis nodosa, a condition he had studied closely and on which he had presented a paper in 1935.² In 1935 Professor Martin Staemmler became director of the Pathological Institute at Breslau University and Wegener went with him as his assistant and as tutor in dissection, histology and pathology to medical and dental students. Wegener observed two similar cases of angitis and granulomas but could find no written reference to the



Friedrich Wegener.

condition. He was reticent about presenting the cases formally because he did not know the cause and was uncertain about whether there would be similar cases, but Staemmler persuaded him to present them to the 29th conference of the German Pathological Society in Breslau. While preparing his talk, he found that one of his best friends, Heinz Klinger, a Berlin pathologist, had published a report on a case of destructive sinusitis, lung abscesses, and uraemia, characterised by angitis and granulomas, entitled "Borderline Forms of Periarteritis Nodosa,"³ and so felt more confident that he was dealing with a new disease. On 27 September 1936 he presented his three cases under the title "On Generalised Septic Vessel Diseases,"⁴ after which the great German pathologist Professor Ludwig Aschoff told him that he believed that it was "a new and very special disease." Wegener then published a detailed study in 1939 entitled "On a Peculiar Rhinogenic Granuloma with particular involvement of the arterial system and the kidney."⁵ This work ceased, however, with the outbreak of the second world war, during which Wegener worked as army pathologist and then troop doctor until captured by the British. After his release in 1945 he worked as a farmer for two years before becoming assistant and then head physician first at the Pathological Institute of the town hospital and then at the Medical Academy in Lübeck. Wegener's interest in his prewar work was rekindled and he found reports of similar cases that had been mistakenly related to periarteritis nodosa. Two cases were published in 1941 by Postal and Laans from Hamburg⁶ and a further case by Lindsay from the United States in 1944.⁷ In 1947 Ringertz, a Norwegian pathologist, described a special form of periarteritis nodosa, which he called Wegener's granulomatosis,⁸ but it was Sven Johnson, a Swedish pathologist who in 1948 first recognised Wegener's granulomatosis as a distinct entity, separate from periarteritis nodosa.⁹ In 1954 Godman and Churg reviewed 22 cases of Wegener's granulomatosis from published reports, adding seven new cases, and putting forward the

diagnostic triad of (1) necrotising granuloma in the upper and lower respiratory tract, (2) generalised focal necrotising vasculitis affecting arteries and veins, and (3) glomerulonephritis with necrosis and granulomas.¹⁰ It was after the first American paper on Wegener's granulomatosis⁷ that Wegener started to appreciate the significance of his findings.

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