Case histories

Systemic lupus erythematosus

Lupus comes from a Latin word that means wolf—an apt metaphor for the history of this savage and elusive condition. How did a painfully erosive rash, likened to cancer and then tuberculosis, become a chronic systemic disorder? Teasing out the details of this transformation shows how different ways of ordering medical knowledge and practice have framed a series of radically different disease entities under the same resonant name.

For medieval European physicians, "lupus" named a broad and loose category of ulcers—"cankery postumes", "evil sores", "ill-favoured lesions". Some practitioners saw little distinction between lupus and their understanding of cancer, although they noted that lupus only ever affected the skin. In a system of medical thought based on resemblance and metaphor, the branching patterns of veins around tumours evoked the claws of a crab (*cancer* in Latin), while the lesions of lupus seemed to mimic the bites of wolves.

In the 19th and early 20th centuries, as western medicine built an uneasy relation with the new laboratory sciences, this ancient category of skin disease was expanded, fragmented, and finally reframed as a disorder affecting the whole body. In the early 19th century, the English physician Robert Willan and his student Thomas Bateman defined a new category of common lupus or *lupus vulgaris*—nodular eruptions similar to syphilis, typically on the forehead, cheeks, and nose, which led inexorably to ulceration and scarring. After 1882, when the German microbiologist Robert Koch identified the bacterium responsible for tuberculosis, *lupus vulgaris* was identified as a form of cutaneous tuberculosis.

Most early attempts to treat *lupus vulgaris* were crudely physical, cutting away diseased tissue or burning it with caustic chemicals. These interventions rarely provided a cure, and patients suffered gradual disfigurement over decades. But Koch had shown that ultraviolet light killed many kinds of bacteria, and this chimed with the notion, embodied in the sanatorium movement, that tuberculosis might be ameliorated by fresh air and sunshine. In the mid-1890s the Danish physician Niels Ryberg Finsen developed a carbon-arc lamp producing powerful "actinic rays", focused through a quartz applicator pressed into the skin. Although the Finsen lamp could cure *lupus vulgaris*, and its inventor received the 1903 Nobel Prize in Physiology or Medicine, patients had to endure uncomfortable therapy sessions for months or years.

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In 1851 the French dermatologist Alphée Cazenave described a condition he called *lupus érythémateux*—a red rash on the cheeks, occurring most commonly in middle-aged women, and leaving permanent scars but no erosions. 20 years later, the Viennese pathologist Moriz Kaposi observed that those diagnosed with *lupus érythémateux* also displayed more general symptoms such as fever, weight loss, and arthritis. At the end of the 19th century the Canadian physician William Osler drew on Cazenave and Kaposi's observations, and his own work on heart, lung, and kidney complications, to create a new disease category of systemic lupus erythematosus (SLE).

The molecularisation of medicine during and after World War 2 cut across the boundaries of 19th-century clinical specialties, offering a new way of thinking about the processes underlying Osler's symptom-based frame. Working at the Mayo Clinic in 1948, Malcolm Hargraves observed phagocytes engulfing free nuclear material in bone marrow samples from patients with SLE, concluding that the condition was autoimmune at root, and 2 years later John Haserick, a dermatologist at the Cleveland Clinic, worked out the details of the disease pathway. Through the 1950s and 1960s American clinicians developed new drug regimens, based on corticosteroids in conjunction with synthetic antimalarials—still the principle behind modern treatments for SLE.

What it is like to live under this regimen may be another matter. In a moving account of her mother's struggle with SLE, one of the few patient narratives in print, Charmaine Crawford calls it "a zero-sum game: you can't survive without it, but your body slowly deteriorates as a result of it". Though the clinical story of lupus has been well told, its cultural history, particularly the question of what successive diagnostic and therapeutic frames have meant for patients, remains to be written.

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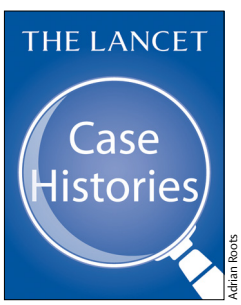

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For more on **Case histories** see **Comment** *Lancet* 2016; **387:** 211 and **Perspectives** *Lancet* 2016; **387:** 217, 737, and 1265

Further reading

Crawford C. When the body attacks: a black woman's fight against lupus. In: Harding SG, ed. Surviving in the hour of darkness: the health and wellness of women of colour and indigenous women. Alberta: University of Calgary Press, 2005: 265–71

Jameson A. More than meets the eye: revealing the therapeutic potential of "light", 1896–1910. *Soc Hist Med* 2013; **26:** 715–37

Potter B. The history of the disease called lupus. *J Hist Med Allied Sci* 1993; **48:** 80–90