

THE BRITISH JOURNAL OF SURGERY

VOL. X.

JULY, 1922.

No. 37.

EPONYMS.

By SIR D'ARCY POWER, K.B.E., LONDON.

V. SIR JAMES PAGET.

THE name of Sir James Paget is associated with a disease of the nipple and with a remarkable change in the skeleton to which he gave the name of 'osteitis deformans'. It is noteworthy that the true pathology of neither of these conditions has yet been worked out.

ON DISEASE OF THE MAMMARY AREOLA PRECEDING CANCER OF THE MAMMARY GLAND.

The paper on "Paget's Disease of the Nipple", as it is now commonly called, appears in the tenth volume of *The St. Bartholomew's Hospital Reports* for the year 1874, pages 87-9. As a classical contribution to surgery it is even shorter than Abraham Colles's description of the fracture of the wrist with which his name is now associated.

Paget writes: "I believe it has not yet been published that certain chronic affections of the skin of the nipple and areola are very often succeeded by the formation of scirrhus cancer in the mammary gland. I have seen about fifteen cases in which this has happened, and the events were in all of them so similar that one description may suffice.

"The patients were all women, various in age from 40 to 60 or more years, having in common nothing remarkable but their disease. In all of them the disease began as an eruption on the nipple and areola. In the majority it had the appearance of a florid, intensely red, raw surface, very finely granular, as if nearly the whole thickness of the epidermis were removed; like the surface of a very acute diffuse eczema, or like that of an acute balanitis. From such a surface, on the whole or greater part of the nipple and areola, there was always copious, clear, yellowish, viscid exudation. The sensations were commonly tingling, itching, and burning, but the malady was never attended by disturbance of the general health. I have not seen this form of eruption extend beyond the areola, and only once have seen it pass into a deeper ulceration of the skin after the manner of a rodent ulcer.

"In some of the cases the eruption has presented the characters of an ordinary chronic eczema, with minute vesications, succeeded by soft, moist, yellowish scabs or scales, and constant viscid exudation. In some it has been like psoriasis, dry, with a few white scales slowly desquamating; and in both these forms, especially in the psoriasis, I have seen the eruption spreading far beyond the areola in widening circles, or, with scattered blotches of redness, covering nearly the whole breast.

"I am not aware that in any of the cases which I have seen the eruption was different from what may be described as long-persistent eczema, or psoriasis, or by some

other name, in treatises on diseases of the skin : and I believe that such cases sometimes occur on the breast, and after many months' duration are cured, or pass by, and are not followed by any other disease. But it has happened that in every case which I have been able to watch, cancer of the mammary gland has followed within at the most two years, and usually within one year. The eruption has resisted all the treatment, both local and general, that has been used, and has continued even after the affected part of the skin has been involved in the cancerous disease.

"The formation of cancer has not in any case taken place first in the diseased part of the skin. It has always been in the substance of the mammary gland, beneath or not far from the diseased skin, and always with a clear interval of apparently healthy tissue.

"In the cancers themselves, I have seen in these cases nothing peculiar. They have been various in form ; some acute, some chronic, the majority following an average course, and all tending to the same end ; recurring if removed, affecting lymph-glands and distant parts, showing nothing which might not be written in the ordinary history of cancer of the breast.

"The single noteworthy fact found in all these cases is that which I have stated in the first sentence, and I think it deserves careful study. For the sequence of cancer after the chronic skin-disease is so frequent that it may be suspected of being a consequence, and must be always feared, and may be sometimes almost certainly foretold. I believe that a nearly similar sequence of events may be observed in other parts. I have seen a persistent 'rawness' of the glans penis, like a long-enduring balanitis, followed after more than a year's duration by cancer of the substance of the glans. A chronic soreness or irritation (of whatever kind) on the surface of the lower lip often long precedes cancer in its substance : and, with a frequency surpassing all other cases of the kind, the superficial syphilitic diseases of the tongue are followed, and not superseded, by cancers which do not always appear to commence in a diseased part of the tongue.

"For an explanation of these cases it may be suggested that a superficial disease induces in the structures beneath it, in the course of many months, such degeneracy as makes them apt to become the seats of cancer ; and that this is chiefly likely to be observed in the cases of those structures which appear to be, naturally, most liable to cancer, as the mammary gland, the tongue, and the lower lip. One may suspect that similar surface-irritation has much to do with the frequency of cancer of the rectum, pylorus, and ileo-cæcal valve, in any of which parts the degeneracy, which might come naturally in old age and make them apt for cancer, may be hastened, and made prematurely sufficient, by an adjacent disturbance of nutrition.

"In practice, the question must be sometimes raised whether a part through whose disease or degeneracy cancer is very likely to be induced should not be removed. In the member of a family in which cancer has frequently occurred, and who is at or beyond middle age, the risk is certainly very great that such an eruption on the areola as I have described will be followed within a year or two by cancer of the breast. Should not, then, the whole diseased portion of the skin be destroyed or removed as soon as it appears incurable by milder means ? I have had this done in two cases, but I think too late. Or, again, when one with a marked family-liability to cancer has syphilitic disease of the mucous membrane of the tongue, with frequent recurrences of inflammation ; should not all the worst pieces of the membrane be removed ? I should certainly advise it, especially if the membrane were ichthyotic, if it were not that the disease is commonly so extensive that good scar-tissue would not be likely to be formed, and that bad scar-tissue, often irritable and ulcerating, is as likely to induce cancer as the syphilitic or ichthyotic patches would have been."

The publication of this paper proved a matter of interest both clinically and pathologically. Those who saw the actual cases and followed up the subsequent course of similar ones were clear that this form of chronic inflammation did not always end in cancer, and that, as Paget stated, local excision was sometimes followed by cure. It was

PAGET'S DISEASE OF THE NIPPLE



Eczema of the nipple in the right breast, occurring three years after removal of the left breast for scirrhus.

*From a drawing by Thomas Codrill, March, 1884,
in the Museum of St. Bartholomew's Hospital*

recognized, however, that cancer occurred very frequently, and the condition was looked upon as 'precancerous'.

The present view held by the majority of surgeons is that cancer of the breast precedes the eczema of the nipple and causes it. Mr. Sampson Handley presents this explanation in the following words (*The British Journal of Surgery*, 1919-20, vii, 189): "A carcinoma starts in the smaller ducts of the breast, perhaps exceptionally from the acini or the larger ducts. Usually, without producing a palpable tumour, it permeates the breast lymphatics widely. The rich plexus of lymphatic vessels around the ducts forms an especially easy and convenient channel for permeation, and the lymphatic block extends along them to the subareolar plexus beneath the nipple. The cutaneous lymphatics about the nipple are now dammed up so that lymph cannot return from them. Later they are themselves permeated, but possibly this is not always the case. At this stage, and before any lump has appeared in the breast, the skin of the nipple and the mucosa of the ducts begin to show changes dependent upon lymphatic obstruction. The epithelium shows disintegration and degeneration of its superficial layers, with proliferation of the deeper layers. These changes are nutritional and non-malignant. The dermis becomes thickened by solid lymphatic oedema. . . . In the rare cases where no carcinoma has made its appearance, though the Paget's disease has lasted many years, it is probable that an atrophic scirrhus, which may have undergone partial or complete cure, preceded the onset of the Paget's disease. But the possibility that the lymphatic obstruction in such cases is of inflammatory origin, and due to a chronic lymphangitis, cannot be altogether excluded."

It will be noticed that Paget's original paper dealt entirely with the clinical aspects of the disease. In 1875—a year after the publication of the memoir—the histological details of two similar cases were described in *The Medico-Chirurgical Transactions*, lix, 107, by Butlin, who added two more in the course of the following year.

Matters rested there for some years, and, as the disease is rare, little notice was taken of it until, on June 4, 1890, Louis Wickham read as his thesis for the Doctorate of Medicine at Paris a "Contribution à l'Étude des Psorospermoses cutanées et de certaines Formes de Cancer Maladie de la Peau dite Maladie de Paget". The thesis opened with the bold statement, "La maladie de Paget est une affection parasitaire du groupe des psorospermoses cutanées, caractérisée par l'inflammation chronique de la peau, des glandes et de leurs conduits, suivie de prolifération épithéliale". ("Paget's disease is parasitic, the result of cutaneous psorosperms, and characterized by a chronic inflammation of the skin, glands, and ducts associated with epithelial proliferation".) The thesis, which was clearly inspired by Darier, who was the head of the Laboratory at the Hôpital Saint Louis, quickly attracted attention throughout Europe and America, and the battle of cancer parasites raged round Paget's disease of the breast for several years. Many observers claimed to have discovered the true parasite of cancer; but no two agreed upon the same, and after a few years the controversy died away. It proved of lasting value, however, because it led skilled histologists to investigate the changes—degenerative and otherwise—which take place in epithelial cells, and many forms of cell-inclusion, vacuolation, and oedematous change became familiar.

The plate of 'Paget's disease of the Nipple' is made by the kind permission of the Treasurer and Governors of St. Bartholomew's Hospital, from a water-colour sketch of a patient sent to the Hospital by Sir James Paget in 1884. The drawing is No. 1057 in the Museum of St. Bartholomew's Hospital.

(To be continued.)