Epithelioma Adenoides Cysticum of Brooke in Mother, Daughter, and two Sons, associated with so-called "Multiple Endothelioma of the Scalp"; its Relationship to Multiple Rodent Ulcer.

By H. G. Adamson, M.D.

Under the name "Epithelioma Adenoides Cysticum," Brooke, of Manchester, in 1892, had described in the British Journal of Dermatology four cases of multiple cutaneous tumours occurring mainly about the face, of epitheliomatous structure, but of benign nature.

For many years this affection had been confused with another type of multiple benign tumour of the skin which was described by Kaposi under the title of "Lymphangioma Tuberosum Multiplex." But it had been conclusively shown by Unna that Brooke's affection was quite distinct from the Kaposi type both in its clinical and in its histological features, and that Kaposi's so-called lymphangioma was really an embryonic sweat gland tumour, and it was now generally known as Syringoma. Its common site was over the front of the upper part of the chest, and not about the face as in Brooke's disease. Brooke's affection was now described as a multiple benign tricho-epithelioma (fig. 1).
Some confusion had also arisen between these two affections of Kaposi and of Brooke and a third type of benign epithelioma which occurred commonly upon the lower eyelid and in which the growths were generally few or single. This third affection was first described by Jarisch in 1894 as "Hæmangio-endothelioma," by Darier as "Hidradenoma" and again by Werther in 1808 as "Tricho-epithelioma." McDonagh had recently studied these growths of the lower eyelid, and stated that they might contain lanugo hair-follicles, sebaceous glands or sweat glands, or undeveloped forms of these organs,

![Image](image_url)

**Fig. 2.**

Case I.—Epithelioma adenoides cysticum. Numerous rounded nodules about the eyelids, at the sides of the nose and scattered in other parts of the face. Rather larger nodules on the temples, forehead and scalp.

that was to say, tricho-epithelioma, sebaceous adenoma, or syringoma. Often two or more of these types were found in one tumour. He regarded them as reversions to the face glands seen on the lower eyelids of deer and other animals.

The lower eyelid tumours were of common occurrence; the multiple tumours of Brooke and the syringoma or lymphangioma tuberosum multiplex of Kaposi were rare.
The cases now shown were typical examples of Brooke's affection. They were of great interest on account of their close resemblance to multiple rodent ulcer in their clinical features, their histology, and in the distribution of their lesions. A further interest was the fact that two of the cases presented those tumours of the scalp which had been called "multiple endotheliomata," but which had been shown by Dubreuilh to be basal-celled epithelioma, and which were demonstrated by the examples before the meeting to belong to the group of cases described by Brooke as "Epithelioma Adenoides Cysticum."

Case I.—F. W., aged 45, housewife. The eruption was first noticed between the ages of 35 to 37. It had increased gradually, and more noticeably after confinements. The eruption consisted of raised rounded nodules, situated in groups: (1) In the naso-labial furrow; (2) at the inner side of the orbit; (3) on the temples. There were scattered nodules on the forehead and cheeks, on the sides of the neck and behind the ears, on the scalp and on the...
shoulders (fig. 2). The nodules were raised, firm, hemispherical, of the colour of the skin, or in the case of the larger nodules, dusky red. Minute vessels could be seen running over the large nodules. They all had much the appearance of the "rolled edge" of a rodent ulcer, but none were ulcerated or crusted, and there were no scars. The nodules on the temples, at the inner orbits and in the naso-labial furrows were small, pinhead to split-pea-sized, and closely packed, with here and there a larger nodule. On the scalp there were many small nodules and three were as large as a small nut or marble. There were several milia on the lower eyelids. A pea-sized nodule had been removed from the forehead for microscopical examination.

Case II.—R. W., aged 21, was the eldest son of F. W. He presented similar nodules on the face, in the same areas, and also several much larger spherical nodules on the scalp and one nodule on the chest below the right clavicle. The nodules on the scalp were as large as unpeeled walnuts and prevented him putting on his hat except at an acute angle. The largest of the nodules on the scalp had appeared three years ago and had rapidly increased to its present size (fig. 3). The nodules on the face had not been noticed until two years ago. The largest nodule and several smaller nodules had been removed from the scalp and examined microscopically.

Case III.—A second son, aged 19, had recently developed nodules on the scalp, but none had been noticed on the face.
Case IV.—I. W., aged 10, daughter of F. W., had a single pinhead-sized nodule on the left naso-labial furrow and one on the forehead. Several smaller nodules could be seen on the cheeks and on the sides of the nose.

The mother of this family stated that she had eight children living, and four of these were affected thus: Girl aged 4, boy aged 7, boy aged 9, unaffected; girl aged 11, a few; boy aged 14, boy aged 16, unaffected; boy aged 19, boy aged 21, both affected. Her mother and one of her maternal aunts had been affected.

In the Lancet of October 17, 1908, the exhibitor had published some cases of multiple rodent ulcer and had pointed out the clinical and histological resemblances of multiple benign cystic epithelioma and multiple rodent ulcer. A comparison of the distribution of the lesions in the case of the mother (fig. 4) with Dr. J. H. Sequeira's well-known record of the distribution of 220 cases of rodent ulcer (fig. 5) showed a striking similarity in areas affected by rodent ulcer and multiple benign tricho-epitheliomata.

But it was not only in their appearance and distribution that these tumours resembled multiple rodent ulcer: their histological characters were also closely similar, so that a section of one of these growths might be pardonably mistaken for one of rodent ulcer. The drawings exhibited (figs. 6, 7, 8, 9) showed that the lesions were made up of oval

\[1\] Lancet, 1908, ii, pp. 1133-38.
Section from nodule on forehead in Case I (fig. 2); masses of oval cells which recall the cells and their arrangement in rodent ulcer.

Rodent ulcer-like growth from one of the nodules of the scalp in Case II (fig. 3). The connexion of the growth with the hair-follicles in some parts, in others with the basal cell layer of the epidermis, is not shown in this drawing.
Section (high power) from large nodule in Case II, showing lobules made up of oval cells with large nuclears and little or no visible protoplasm. The section shows also the hyaline bodies described by Dubreuilh in sections from a similar case of benign epithelioma of the scalp.

Section from a benign lower eyelid growth present since infancy in a man, aged 37, who now had a typical rodent ulcer of two years' duration on the nasolabial furrow. The resemblance of the benign growth (basal-celled epithelioma) to that of rodent ulcer is striking.
cells derived from the basal cell layer of the epidermis, and in the case of one section (fig. 6) there was a suggestion of origin from the pilo-sebaceous follicle. The cells were arranged in lobules with a marginal palisade layer and here and there cystic formations similar to those seen in rodent ulcer. Of special interest was fig. 8, which represented a section from one of the nodules on the scalp of the elder son. It was easy to see how these lesions had been mistaken for endothelioma; but at the same time their resemblance to rodent ulcer was obvious.

As a corollary to these cases the exhibitor showed a drawing (fig. 10) from a section of a congenital lower eyelid tumour in an adult who had also a rodent ulcer in the naso-labial furrow. The lower eyelid tumour was clearly a basal-celled epithelioma and its resemblance to rodent ulcer was striking.

The inference drawn from these cases was that rodent ulcer was a nævoid growth, similar in nature to the benign epitheliomata, but differing in that the latent embryonic foci were aroused from their dormant state at a later period of life, when the cells composing them had retained only their function of proliferation, and had lost their power of differentiation. It was well known that late in life there was a tendency to growth of coarse hairs in the eyebrows and nostrils and elsewhere, and this corresponded with the stimulus to growth of latent embryonic pilo-sebaceous follicles which led to rodent ulcer.

DISCUSSION.

Dr. A. EDDOWES mentioned a case which he showed many years ago, with drawings, at the British Medical Association meeting in Oxford. He had shown the drawings on several occasions since. In his sections he fortunately got staining of the contents of the cystic tubules, which showed a very large number of organisms which were all similar, and had the appearance of minute bacilli in chains, or a small streptococcus. He suggested that every attempt should be made to get cultures from the present cases. Possibly we had here a special organism which had not yet been studied. He believed that the lesions were infective. The patient shown at Oxford had to leave for India, but before he left he removed nearly all the nodules, and there was no recurrence while the patient remained in England. He had since lost sight of the man, whom he advised to see a surgeon on arrival in the East and have the condition watched, because if neglected a fungating process might be set up, or ulceration similar to that of rodent ulcer.

Dr. MACLEOD asked if it was Dr. Adamson's idea that rodent ulcer was invariably a tricho-epithelioma.
Dr. Whitfield said that the diagrams which Dr. Adamson had shown were so striking that one could not neglect the similarity of site between tricho-epithelioma and rodent ulcer. He thought, however, that Dr. Adamson's explanation of the growth of tricho-epithelioma and rodent ulcer respectively was not entirely satisfactory. It was true that rodent ulcer did not often form a good tumour, but this was because it broke down into an ulcer. Actually rodent ulcer made a larger amount of new tissue than did tricho-epithelioma. One could hardly say that tricho-epithelioma was a more robustly growing tumour than rodent ulcer, since the latter seemed the more powerful both as regards producing a greater amount of new growth and invading the tissues of the host.

Dr. Sequeira regarded the communication as very interesting, and especially the suggestion of Dr. Adamson that the rodent ulcer was derived from the rudimentary hair-follicle. In the important debate on rodent ulcer at the Pathological Society in 1894 Dr. Colecott Fox and others placed the origin of rodent ulcer in the hair-follicle. Dubreuilh and Auché also traced the origin in many cases to the pilo-sebaceous follicle. The point raised by Dr. Whitfield he regarded as a good one, because the tumours on the scalp were composed of masses of fairly innocent growth, and were distinct from the growth met with in the case of rodent ulcer when attacking the scalp. Rodent ulcer on the scalp was usually of the superficial cicatrizing type; it did not produce large growths or deep infiltrations. Dr. Adamson's suggestion that multiple rodents may start from congenital anomalies was very important. The speaker had been interested in the origin of many rodent ulcers in early life, and had reported several cases. Recently he saw a case in which the rodent ulcer began as a growth which was described as being a bluish "pimple" at the age of 11; the patient was now only aged 20, and for the last four years he had had a typical rodent ulcer. The old idea that rodent ulcer was always a growth of old age must be abandoned. The early cases were usually diagnosed as lupus.

Dr. Adamson said, in reply to Dr. Whitfield, he had not intended to suggest that the growth of rodent ulcer was less abundant than that of benign tricho-epithelioma. It was probably more active but less stable, for the cells which made up a rodent ulcer had retained their power of active proliferation, while they had lost their tendency to develop into a special type. They thus formed an unstable mass which readily ulcerated and which also did harm to the healthy tissues by its presence and growth amongst them.