MULTIPLE BENIGN EPITHELIOMA OF THE SCALP
(TURBAN TUMORS)

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Tumors of the scalp of a peculiar kind, grouped to resemble bunches of grapes or tomatoes, of various sizes from a pea to a billiard ball, often so numerous as to cover the entire scalp like a turban, have been the subject of much discussion. A few writers consider them as of endothelial origin, but a large majority, including the most recent authorities, look upon them as epithelial. These tumors are slow growing and benign. They are definitely familial and usually occur in the female sex. They are rare, at least in this country.

Few text-books of surgery include these tumors in the long lists of growths which may be found on the scalp. They are not mentioned at all by most writers on sebaceous cysts of the scalp. Text-books on dermatology consider them briefly, some under the old heading "endothelioma capitis," others including them in the chapter on basal-cell epithelioma. In France they are described as "cylindroma of the scalp" or simply as "cylindroma," a term which is also used by German and British writers.

Most of the cases reported are from the German and French literature. Only 3 observations, as far as the writer knows, are recorded in the American literature, one by Cohn (1), as far back as 1892, the second in Sutton's text book of skin diseases (2), and the third by Stillians (3), who reported it at a dermatological meeting in Chicago.²

Ancell's case (4), observed in 1842, is the first of this type of tumor to be recorded. It has been criticized by some authors on account of a concomitant malignant internal growth, considered as metastatic and followed by cachexia and death, but the description is clear and the history typical.

The patient was a woman with small nodules on the face and large tumors on the scalp. The condition was first observed at the age of fourteen, but progressed slowly and caused no trouble

¹ Acknowledgment is due Dr. Philip Batchelder for permission to use the photographs and Dr. B. Earl Clarke for the photomicrographs.

² Since the present paper was submitted for publication one more case, bearing a striking clinical and pathological resemblance to the one here reported, has appeared (Jones, Alden, and Bishop, 54).
until the age of fifty-two. The same condition was present in three female members of the family, who otherwise enjoyed perfect health. Only five months previous to coming under Ancell's care the patient complained of an abdominal growth, and a malignant tumor was found. The scalp tumors showed no sign of malignancy, and the abdominal neoplasm appears clearly as concomitant and not metastatic.

Following Ancell's report, cases were published in extenso, with clear descriptions, illustrations, and histopathology by Poncet (5), Barrett and Webster (6), Barlow (7), Cohn (1), Mulert (8), Kaposi and Spiegler (9), Aitken (10), De Beurmann (11), Friebos (12), van Leewen (13), Coenen (14), Adamson (15), Civatte (16), Pinkus and Watanabe (17), Burnier and Rejsek (18), Delaye et al (19). Nasse (20), Koulieff (21), Rafin (22), Seitz (23), Joseph (24), Drake (25), Sutton (2), and Stillians (3) reported cases briefly at dermatological meetings or in textbooks on skin diseases. Hutchinson's case (26), case 2 of Bérard (27), and Cicconardi's case (28)—fungating tumors involving a large part or the entire scalp, distinctly benign and representing an advanced stage of the disease—are also to be included in this group. The variety of titles chosen by the authors to designate these tumors indicates the difficulty of placing them clinically and pathologically.4

The most popular picture of the condition is the drawing of Poncet's case (5), reproduced by Bérard (27), Spiegler (9), Sabouraud (30), Darier (31), Hazen (32), and Pusey (33). (Fig. 1.)

An extensive résumé of the previously published cases has been made by Dubreuilh and Auché (34) and by Frieboes (12). Zeit (35), Martinotti (36), Wise (37), Savatard (38), Stokes (39), Pusey (33), Gans (40), Crosti (41), McCarthy (42), and Jacoby and Grund (43) published no cases but discussed the subject in their publications.

Thirty-one fully developed cases are recorded in the literature, 19 in females, 12 in males. The age ranges from forty to eighty. In 10 cases nodular formations on the scalp occurred in one or more other members of the family, representing probably an initial stage or an abortive type of the same disease. Of these, 15 occurred in female members of the family, 4 in males. Considering together the fully developed cases and the initial or abortive ones, the total number approximates 50.

Recurrences are reported in few cases. They are to be con-
FIG. 1. SOME STRIKING CASES OF TURBAN TUMORS FROM THE LITERATURE
sidered as exceptional, or as due to incomplete removal or to a near-by tumor mistaken for a recurrence.

Cases of rapidly malignant growths (Oro 45, Haslund 46, Sequeira 47), growths in an initial stage (Dubreuilh and Auché 34, Nicolas et al. 48, Schmidt-Bäumler 49), and solitary tumors (Dalous 50) should not be included among the turban tumors.

REPORT OF A CASE

S. B., male, aged seventy-two, a janitor, born in Sweden and resident in America for many years, came to the Surgical Out-Patient Department of the Rhode Island Hospital on July 7, 1931, for multiple tumors of the scalp. One tumor was excised and another, which spontaneously ruptured, was cleaned and dressed. Radiation of the entire scalp with x-rays caused transitory alopecia but no modification of the tumors. On October 9, 1931, the patient was referred to the Skin Out-Patient Department.

Family History: The patient's mother had died at an advanced age after breaking a hip. She had had several tumors, the size of a pea, on the edge of both ears. The father, 9 brothers, and one son had no tumors. A daughter, aged thirty-eight, had small tumors on the scalp. Her case is reported below (page 881).

History: The patient had suffered the common ailments of childhood and had been in good health since. He gave no history of traumatic lesions on the head. More than twenty years ago he first noticed small growths on the scalp and back, slowly increasing in number and size. His only complaint was that the largest tumors interfered with wearing a hat, and of the unsightly appearance.

Physical Examination: On the face were a few scattered whitish tumors the size of a pin-head and of the clinical appearance of syringoma, and a few of Balzer's adenomata sebacea. On the forehead were three round, pinkish nodules, smaller than a pea. On and above the hair line was a cluster of about 50 tumors, ranging from the size of a pin-head to that of a small cherry (Fig. 2). Several more tu-

Fig. 2. Tumors on forehead, top, and side of the head in case reported

mors were scattered over the rest of the scalp, the size of a cherry or of a small walnut. Some were protruding, lobulated, and freely movable, with a large base. Some were almost flat and more or less embedded in the subcutis. Some had a narrow base and appeared slightly pedunculated.

On the back (Fig. 3) were four flattened and slightly pedunculated tumors, the size of a hazelnut, and a great many tumors the size of peas. There were also a few pigmented moles.

The tumors of the frontal region of the scalp were sharply pink. The largest tumors on the rest of the scalp were purplish blue. The ones on the back were whitish. On the surface they showed enlarged tortuous vessels. The small tumors were of firm consistence; the larger ones were softer and rubber-like. They were smooth and almost hairless.

The Wassermann test was negative and the urine, blood chemistry, and blood count were normal.

Pathologic Examination: Tumors of various sizes both from the patient and his daughter were examined by Dr. Clarke, pathologist of the Rhode Island Hospital, and declared to be basal-cell epithelioma of non-cornifying type.

The excised tumors were fixed in formalin and the sections stained with hematoxylin-eosin, van Gieson, Mallory. Sections from one of the largest tumors show...
an irregularly nodular formation lined by a thin, flat epidermis. A thick capsule of connective tissue surrounds the whole tumor and projects toward the center, dividing cylindrical masses of epithelial cells in palisade formation on the borders. The connective tissue in direct contact with the epithelial masses has undergone

**Fig. 4. Low-power sections of one of the larger nodules, showing the flat, thin epidermis, the capsule of connective tissue with scattered cystic formations, and the cylindrical tumor masses surrounded by connective tissue undergoing hyaline degeneration.**

To the right is shown the upper part of an older tumor mass with blood cavities and hyaline degeneration between the tumor cells.

**Fig. 5. Low-power sections from the nodule shown in Fig. 4, showing the thick capsule of connective tissue and, at the right, a large round tumor mass, with large cavities filled with blood and with hyaline substance.**

hyaline degeneration and forms a well defined ring, deeply stained. The external row of cells is also more deeply stained than the central mass. Among the cylindrical groups of epithelial cells there are small spaces filled with hyaline substance, and scattered about are small cystic formations and large cavities filled with blood or with amorphous material (Figs. 4 and 5).
Numerous sebaceous and sweat glands are visible in the subcutis between the tumor mass and the epidermis. Toward the vertex of the tumor the glands and hairs appear gradually more and more flattened, atrophic, or missing, on account of the pressure of the tumor toward the epidermis.

![Low-power section from a small nodule, showing mostly connective tissue, scanty sebaceous glands, and a solitary tumor mass, representing an abortive formation.](image)

At a few points, round masses of tumor cells seem to have a connection with the sebaceous glands and, possibly, to originate from them. No direct connection of the tumor with the epidermis is noticeable.

Sections of one of the pea-sized tumors show the following similar structure. There is a large solitary cylindrical mass of epithelial cells (Fig. 6) with less marked hyaline degeneration in the center and at the periphery. A large part of the nodule is formed by connective tissue containing normal glands. The epidermis is thin and flat. No cyst formations are present. No point of origin is appreciable. These small nodules appear as abortive tumors rather than as young formations.

Treatment: On account of the age of the patient and the little trouble given by the disease, one or two of the largest tumors were excised at a time. Some were removed with the cutting current; a flap was then made and the base fulgurated and covered with the flap to obtain the best cosmetic result. Some of the small growths were desiccated. One, the size of a small walnut, was treated by implantation, at the base, of 7 needles of 5 mg. radium, left in place for seven hours. While for four months the irradiated tumor seemed unmodified, after seven months it was reduced to one third of its original size.

Spontaneous involution was never observed by the patient, nor recurrence up to the present time. One tumor, partially destroyed, secondarily infected and discharging for about three months, showed no signs of recurrence or of malignancy.

S. B., married daughter of the patient just described, aged thirty-eight, housekeeper, was seen on Jan. 29, 1932. She had no complaints except for a few small tumors on the scalp, which she feared would grow as in her father's case.

She showed four nodules the size of a pea or a bean, of pinkish hue, firm, hairless and indolent, of about ten years' duration. There were three smaller nodules,
the size of a pinhead, which she had first noticed only a few months before. The
largest tumor, on the vertex, had been removed five years earlier and had recurred,
probably because of incomplete removal. One located on the nucha, near the hair
line, was embedded deeply in the subcutis and was treated by electrodesication.
The three smallest tumors on the scalp were treated similarly. The others were
excised in toto with the cutting current, and the site was fulgurated. A recovery
with perfect scar formation followed, and at the present time no sign of recurrence
is noticeable.

The largest nodule excised from the vertex shows essentially the same structure
as was found in the tumors removed from the patient's father. It appears as a
flat nodule with the same thin, flat epidermis (Fig. 7), with cylindrical masses of
epithelial cells separated from each other by a well defined band of connective tissue
undergoing hyaline degeneration. Small spaces filled with hyaline substance are
present between the tumor cells. The hyaline degeneration is more evident than in
the father's tumors and there are fewer cystic formations.

Diagnosis

A clinical diagnosis is fairly easy. The history is invariably
the same: onset at fourteen to thirty years of age; full develop-
ment between fifty and seventy; occasionally trauma with rupture
and quick recovery; slow growth and slow multiplication up to the
number of 500; other cases in the family, chiefly in the female
members; no complaints except for the unsightly appearance and
the occasional rupture of some of the tumors, with secretion and
the formation of a fungating mass, which may be considered, at
first impression, as malignant (Hutchinson, 26, Bérard, 27; Cic-
conardi, 28).

Fig. 1 shows some of the most striking cases in the literature.
Typical, also, are the pictures of the cases of Joseph (24) and
Delaye et al (19). Some pictures of published cases are less clear
and some are poor, as the colored plates of De Beurmann (11) and
Sequeira (47). The former shows a pathologic process of the nose
of indeterminate appearance; the latter a rapidly malignant growth
which is inconsistent with turban tumors.

In regard to the differential diagnosis, the most common mis-
take is a diagnosis of wen. The number, ordinarily one or two;
the color, pale or as normal skin; the shape, spherical, with no
lobulations; the pultaceous consistence; the absence of concomitant
nodular formations on the face, point to a diagnosis of sebaceous
cyst. The history of a similar affection in the family, the long
duration and slow growth of the tumors, their large number, their
varied size, their grouping, their lobulated or pedunculated shape,
firm, rubber-like consistence, and pink or bluish hue, lead fairly
easily to a diagnosis of turban tumors.

A correct diagnosis is important in early cases, when the tu-
mors can be easily removed by electrodesication, with very slight
likelihood of recurrence. Cases involving the entire scalp can be
treated successfully by removal of a few tumors at a time. A
Fig. 7. Low-power and high-power views of the same section from a nodule removed from the patient's daughter, showing the cylindrical masses of epithelial cells surrounded by a thick band of hyaline substance.

Fig. 8. High-power view (right) of point A of the left photomicrograph, suggestive of a sebaceous gland undergoing transformation into a tumor mass.
correct diagnosis is also of great practical importance in advanced and complicated cases, as those reported by Hutchinson (26), Bérard (27), and Cicconardi (28). In such cases, with fungating masses covering the scalp in part or entirely, and representing a late stage of the disease, the surgeon may be uncertain whether or not to attempt operation. The clinical appearance (lobulation, etc.) and the history should lead to the right diagnosis. Such cases have been treated successfully by surgical excision of the entire tumor mass.

**Pathogenesis**

The cylindrical tumor masses appear to be made up of epithelial cells surrounded by connective tissue undergoing hyaline degeneration, as is part of the tissue between the tumor cells.

These tumors have been regarded as basal-cell epithelioma of nevus origin by Frieboes (12), Watanabe (17), Hoffmann (52), Gans (40), McCarthy (42), and others. Ricker and Schwalb (53) believed that they were derived from the sweat glands. A more satisfactory explanation regards them as arising from the pilosebaceous system, particularly from the epithelium of the sebaceous glands. In some sections of the case here reported the tumor mass seemed to be very near, and to constitute part of a sebaceous gland undergoing transformation into an epithelial tumor mass. This would confirm the impression of such writers as Poncet (5), Bérard (27), Dubreuilh and Auché (34) and others.

An origin from the sebaceous glands would make clearer the relationship between the so-called adenoma sebaceum of Pringle and Balzer and Ménetrier, the epithelioma adenoid cysticum of Brooke, and the turban tumors.

**Nomenclature**

The term "multiple, benign, familial, nodular, turban-like, basal-cell epithelioma of the scalp, with cylindromatous degeneration" would seem to be as complete and clear a designation of these tumors as is possible, but it is obviously too long. A clinical heading should give a quicker and clearer idea of what is referred to, and the term "turban tumors," recalling the appearance of the Turkish head-dress, is still the shortest and the best. Uniformity in nomenclature would save much space and time.

**Treatment**

X-rays have been of no value in the present case. Radium was used in a small dose and only on one tumor. It is possible that by the use of a large amount of radium a better result might have been obtained and the annoyance of profuse bleeding during sur-
gical removal might have been avoided. Owing to the relative benignity of this type of tumor, I believe the best results can be obtained, as in the present case, by excising a few tumors at a time with the scalpel or with the cutting current, thus avoiding the possible dangers of heavy irradiation. The profuse bleeding is better checked by ligation of the vessels than by excessive use of the coagulating and fulgurating current, which may lead to sloughing and to delayed recovery.

**Summary**

The literature on "multiple, benign, familial, nodular, turban-like, basal-cell epithelioma of the scalp, with cylindromatous degeneration," called also "endothelioma capitis," Spiegler's tumors, cylindroma, etc., has been reviewed. The nature of these rare tumors is established as epithelial by the majority of authors. An origin from the sebaceous glands is the most reasonable, although this is difficult to prove.

A strict relation between adenoma sebaceum, epithelioma ade-noide cysticum and the turban tumors is clear, and they must be considered as belonging to the same group, in different stages, of the same nevic disease.

The number of names under which this syndrome is listed demonstrates the difficulties of classification. At any rate the ancient denomination of "endothelioma capitis" should be dropped, as well as "cylindroma," the hyaline degeneration being a characteristic not limited to these tumors. "Multiple benign epithelioma of the scalp," or, from the striking clinical appearance, "turban tumors," are suggested as the best names.

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