

Section of Surgery

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Turban Tumours

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TURBAN tumour is the popular name given to a type of basal cell carcinoma which arises deep in the dermis from the sweat glands or the pilo-sebaceous system. This definition is a simplification of much discussion in the literature as to the true nature and classification of the neoplasm. They were first described by Ancell in 1842 and were originally thought to be of endothelial origin, but, in 1933, Ronchese concluded that they were subepidermal in origin, beginning in the epithelium of the sebaceous glands, although others at that time believed that the sweat glands were the primary site. In 1943, Warvi and Gates differentiated between tumours arising from the sweat glands and sebaceous glands, naming the first syringomata and the second epitheliomata adenoides cysticum. However, they did recognize the possibility of a common cause and suggested that a congenital abnormality of the dermis might give rise to a variety of unusual epithelial lesions. Willis, in 1953, grouped under the term subepidermal basal cell carcinoma lesions arising from either sweat glands or pilo-sebaceous epithelium.

The lesions may involve any part of the skin, but most frequently the scalp, face, neck, chest, back, and external genitalia. When the scalp is prominently involved, turban tumour aptly describes its appearance. The familial nature of the disease is well recognized; Evans (1954) reported a 33½% incidence of multiple involvement. This hereditary tendency of the disease is well illustrated in the following cases. They are discussed in more detail by Chalstrey (1955).

CASE HISTORIES

R. W., a 62-year-old farmer, was admitted to hospital in March 1955, complaining of multiple nodules on the head and body. At the age of 19 he first noted a swelling on his head. During the following years a few nodules appeared on his body and by the age of 22 numerous tumours had appeared on his scalp. Many local excisions were carried out but as more were excised, more appeared. Five years ago the majority of the lesions involving the back and chest were removed. In 1954, multiple large tumours were present on the scalp but despite local removal of some two dozen in August of that year, they continued to multiply and become larger. At the time of admission there was massive involvement of the scalp. Some of the tumours had broken down and discharged a foul-smelling material. They ranged in size from 0.3 to 4.0 cm. in diameter and were of a variety of shapes, some being almost flat and embedded in the surrounding skin, others lobulated and freely movable on a short, wide base, and a few pedunculated (Fig. 1). Most had a pinkish, fleshy appearance, while others were bluish in colour. The smaller nodules tended to be firm in consistency but the larger ones were soft and rubbery and gave the impression of being cystic. Similar tumours were also present to a lesser degree on the face, neck, back, upper chest scrotum and calves.

On March 18, 1955, radical mass excision of the most severely affected area of the scalp was carried out with the application of split-thickness grafts to the defects. Fig. 2 shows the scalp seven months after operation. Histological section showed subepidermal basal cell carcinoma consisting of nodular formations covered by a thin, flat, intact epidermis (Fig. 3). Connective tissue capsules lined each tumour and projected inwards, dividing the tumour into clumps of basal cells lined by hyaline membranes. Small round deposits of hyaline material were also present between the cells and in some places there were cystic spaces and pseudoglandular formations.

Since operation the patient's head has had a total of 3,150 r at 250 kV which has caused a noticeable shrinkage of the remaining lesions.

Fig. 4 shows the patient's family tree; 12 persons are affected, 7 being female. Evans found a similar sex distribution in his collected cases. The patient's son developed turban tumours at the age of 13. A number have been excised and all have the typical histological picture of subepidermal basal cell carcinoma. One daughter first noted the tumours when she was 19. Now at age 35, multiple tumours are present over the scalp, forehead, cheeks and chin. A number have been excised and microscopic study of these reveals a similar appearance to the lesions of her father. The patient's 60-year-old brother is also affected; his tumours first appeared at age 20 and recently have been enlarging. In addition he has involvement of his neck, chest and back. The patient's youngest sister



FIG. 1.—Side of patient R. W., showing the involvement of the scalp and face by turban tumour.

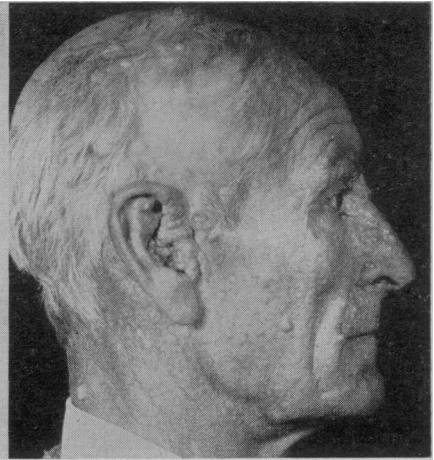


FIG. 2.—Seven months after mass excision of turban tumours and grafting. Side view; compare with Fig. 1.

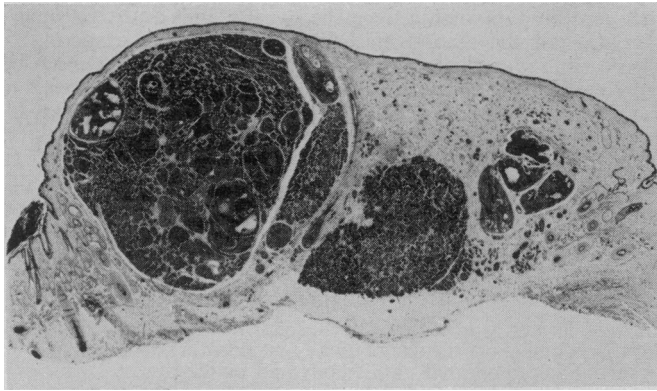


FIG. 3.—Subepidermal basal cell carcinoma $\times 5$. Note the intact epidermis, encapsulation and cystic spaces.

first became involved at age 29. Two other brothers and one sister in addition to the patient's mother, two aunts and grandmother have similar disease.

DISCUSSION

Turban tumours or subepidermal basal cell carcinoma are of rare occurrence compared to the more usual basal cell carcinoma which arises in the epidermis and is often called rodent ulcer. However, the marked disfigurement they may cause and their hereditary nature make them an interesting study. Evans (1954) surveyed the literature between 1842 and 1954 and collected 47 cases. Lyon (1955) reported another case to this Society. There was no familial history in this case. Undoubtedly there are many more cases which have not been reported or, due to disagreement as to the pathological classification, have been listed as another entity. It would seem best to discard the multitude of terms used in the past to describe these tumours and to classify them as subepidermal basal cell carcinomata of sweat or pilo-sebaceous gland origin.

SUMMARY AND CONCLUSIONS

A case of subepidermal basal cell carcinoma is described in which 11 other members of the same family have the disease. Onset is usually in adolescence or early adult life. At first the tumours enlarge and multiply slowly but by the sixth decade marked involvement of the scalp and other areas of the skin is present. The lesions do not metastasize but may occasionally erode underlying bone by pressure as in Lyon's case. They are painless and do not impair general health, but become unsightly, sometimes ulcerate, and often lead

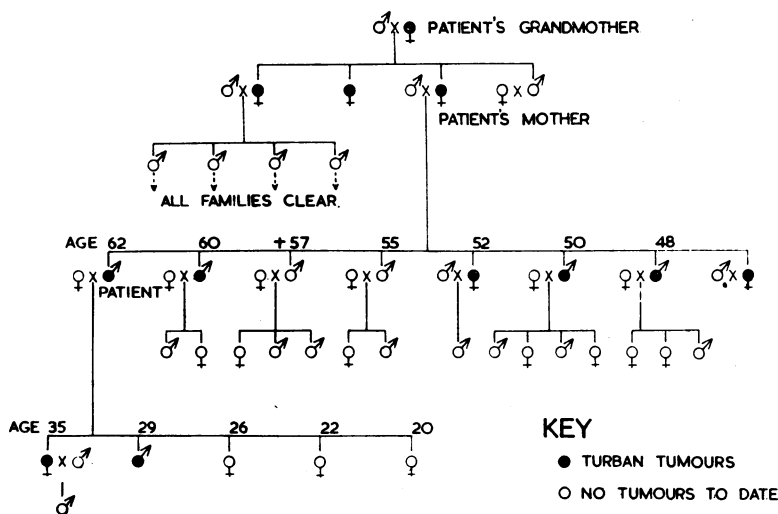


FIG. 4.—Individuals with turban tumours in one family.

to marked psychological depression of the patient. Pathological study shows them to be basal cell carcinomata beginning in the epithelium of sweat or sebaceous glands and proceeding to nodular tumours of varying size with an intact epidermis over them. Hyaline deposition with cystic and pseudoglandular formation is common and occasionally calcification occurs within them. When the disease appears, there is one chance in three that it will involve other members of the patient's family. It is an example of a Mendelian dominant trait, apparently stronger in the female sex.

The treatment of choice is surgical excision with skin grafting as necessary. X-ray therapy is of some value in that it will cause regression, but it does not cure. Evans found that those lesions treated by roentgen rays alone usually recur after a few months.

A complete descriptive term for the disease would be multiple, benign, familial, nodular, subepidermal, basal cell epithelioma, but this is far too cumbersome. When, as is usual, the scalp is the main site of involvement, the resemblance to an Eastern head-dress makes the term turban tumour a fitting title.

Acknowledgments.—Figs. 1–4 are reproduced from *St. Bartholomew's Hospital Journal* by kind permission.

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Orlon Prostheses in Arterial Surgery

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BECAUSE of difficulties encountered in obtaining suitable autopsy material for stored arterial homografts and the risk of infection associated with this method, an investigation into the use of plastic arterial prostheses was undertaken.

Orlon fabric was chosen as the most suitable material. Tubes of this fabric are easily made, the seam being sewn with an ordinary sewing machine. A cuff is turned back at each end of the tube so that a double thickness of orlon is available for the anastomotic suture. The completed tubes are sterilized by boiling or autoclaving. Orlon prostheses were used, experimentally, to replace the terminal aorta in dogs, and were found to function well and remain patent over long periods. Excised specimens showed that the tube becomes surrounded by a layer of hyaline collagenous tissue which penetrates the mesh of the