

TRICHOEPITHELIOMA

(Two Clinico Histological Cases Reported)

By

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Trichoepithelioma, also known as Epithelioma Adenoides Cysticum of Brooke, and Multiple Benign Cystic Epithelioma, is a tumour of adnexal origin. This name is preferable to the other names, because it indicates that differentiation in this tumour is directed towards hair structures.

Trichoepithelioma usually occurs as multiple lesions, but solitary lesions have also been reported. In multiple trichoepithelioma the condition is dominantly inherited (Gaul)². Generally, the lesions appear at puberty and gradually increase in number and size. The lesions are numerous rounded yellowish or pink nodules, from pinhead to pea in size, usually on the face and occasionally on the trunk. Telangiectasia is common. Occasionally one or several lesions become ulcerated because of change into a basal cell epithelioma. Solitary trichoepithelioma, which is not hereditary, is characterised by a slow-growing nodule usually in adult life, the lesion being present on the face or elsewhere on the skin (Traenkle³, Nikolowski⁴, Zeligman⁵).

Adamson⁶, Little⁷ and Savatard⁸ reported cases in which one or several of the lesions of trichoepithelioma after having persisted as such for many years, developed into ulcers with the histologic appearance of basal cell epithelioma. The close relationship of trichoepithelioma and basal cell epithelioma has been explained by assuming that they have a common genesis from pluripotential cells that may develop towards hair structures; i. e. from primary epithelial germ cells (Lever)¹. Thus, the two types of tumors differ merely in the degree of maturity of their cells (Adamson). Since cells at various degrees of maturity may occur in the same lesion, one may find in trichoepithelioma, areas consistent with the histologic picture of basal cell epithelioma and vice versa; also if active growth occurs in a lesion of trichoepithelioma, the newly formed cells may be less differentiated than the older cells, and the lesion may grow as a basal cell epithelioma.

A close relation exists not only between trichoepithelioma and basal cell epithelioma, but also between trichoepithelioma and other types of benign epitheliomas, such as syringoma and cylindroma. Hartzell⁹, McDonagh¹⁰, Weidman and Besaneon¹¹ have reported the simultaneous occurrence of syringoma an apocrine epithelioma and trichoepithelioma in the same patient and even in the same lesion. (Ingels¹², Lever¹³) While Adamson⁶, Watanabe¹⁴, Schlammadinger¹⁵, Schucman & Weber¹⁶, Savatard⁸, Kleine-Natrop¹⁷ have reported the simultaneous presence of cylindroma an apocrine epithelioma and trichoepitheliom in the same patient.

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Histopathology. On Histologic examination, trichoepithelioma appears as a well circumscribed tumor. Horn cysts represent the characteristic lesion. They consist of a fully keratinised center surrounded by a shell of flattened "basal cells" without prickles. The keratinization in the horn cysts is abrupt and complete, not gradual and incomplete as in the horn pearls of squamous cell carcinoma. This process corresponds to the abrupt development of the horn cells of the hair matrix cells (which also are cells without prickles) Lever is of the opinion that the cells surrounding the horn cysts are hair matrix cells and that the horn cysts represent attempts at hair shaft formation. Occasionally one sees squamous cells with distinct intercellular bridges around some of the horn cysts. They represent outer hair sheath cells. Since the outer hair sheath develops from squamous cells at a time when the hair germ already has advanced to a rather high stage of differentiation, the presence of squamous cells around the horn cysts is evidence of rather high differentiation.

In addition to horn cysts, irregularly shaped islands and intertwining strands composed of "basal cell" are present. Such areas are indistinguishable from basal cell epithelioma (Goldman)¹⁸. Abortive hair papillae and hair shafts are seen occasionally. Since hair papillae contain a high concentration of alkaline phosphatase (Hardy)¹⁹, their presence can be demonstrated by the use of the Gomory stain for alkaline phosphatase. Calcification of the horn cysts may occur and evoke a foreign-body giant-cell reaction in the adjacent connective tissue.

In multiple trichoepitheliomas, not infrequently some of the lesions show relatively little differentiation, containing only a few horn cysts but many areas with the appearance of basal cell epitheliomas (Summerill & Hutton²⁰, Traenkle³). Such lesions are indistinguishable histologically from a keratotic basal cell epithelioma, which may also show horn cysts. Thus, since no sharp line of demarcation exists histologically between multiple trichoepithelioma and basal cell epithelioma, it may be necessary in order to arrive at a diagnosis in a given case, to have knowledge of clinical data, such as the number and the distribution of the lesions, presence of hereditary transmission and the age at which the lesions first appeared.

Solitary trichoepitheliomas are histologically designated by lesions showing a high degree of differentiation towards hair structures. Lesions with relative little differentiation towards hair structures are best classified as keratotic basal cell epitheliomas. Thus, for a lesion to qualify for the diagnosis of solitary trichoepithelioma, it should contain numerous horn cysts as abortive hair papillae and hair shaft and only few areas with the appearance of basal cell epithelioma (Traenkle³, Nikolowshi⁴, Zeligman⁵).

CASE HISTORIES

Case I Patient S. B. Sex—Male Age—36 years.

Complaint—Attended the out-patient department of the hospital with history of lesions over the face for the last 2 years. The lesions were slowly progressive in size and not accompanied by any pruritus.

Examination of the patient showed that the lesions were smooth, shiny well defined papular lesions of normal skin colour and firm to feel located on the forehead nose, naso-labial folds and on the upper lip. Lesions were varying in size from 1 mm to 4 mm in diameter.

Skin Biopsy; The characteristic features of trichoepithelioma namely the presence of keratinizing or horn cysts was evident. In addition the presence of immature hair structures and abortive hair papillae were occasionally seen. Thus confirming a diagnosis of Trichoepithelioma.

Case II. Patient H. D. Sex—Female Age—75 years.

Complaint—History of lesion over the nose for the last 4/5 years. The lesion was very gradually increasing in size, there was no pruritus, but history of smarting sensation on exposure to sun light.

Examination revealed a solitary lesion over the nose, of normal skin colour and firm to feel. The size was 4–6 mm in diameter.

Skin Biopsy: Section showed in addition to the horn cysts, irregularly shaped islands and intertwining strands composed of basal cells, indistinguishable from that of basal cell epithelioma. There were larger areas showing the appearance of basal cell epithelioma as compared to the number of horn cysts, abortive hair papillae and hair shafts.

DISCUSSION

Of the two cases presented, Case I does not present any diagnostic problem. The histological pattern conforms to the diagnosis of trichoepithelioma. The only note worth factor, is the late development of the lesions, occurring at the age of 34 years, as opposed to the commonly seen occurrence of trichoepithelioma at or round about puberty.

In Case II the histological pattern presents the characteristic of both trichoepithelioma and basal cell epithelioma. Lever states that since there is no sharp line of demarcation existing in differentiating between trichoepithelioma and basal cell epithelioma, histologically, it may be necessary, in order to arrive at a diagnosis in a given case to have knowledge of clinical data, such as the number and distribution of the lesions, and the age at which the lesions first appeared. Besides, histologically to qualify for the diagnosis of trichoepithelioma, the lesion should contain numerous horn cysts, abortive hair papillae and hair shafts and only few areas with the appearance of basal cell epithelioma. In Case II the age of onset of the development of the lesions being 70 years, the histological evidence relatively of large areas showing the appearance of basal cell epithelioma and fewer horn cysts, abortive hair papillae and hair shafts go more in favour of the lesion being a keratotic basal cell epithelioma.

CONCLUSION

1. The clinico-histological picture of two cases is reported.
2. Case-I presented the typical histological pattern of trichoepithelioma. Note worthy factor, being the appearance of the lesions at the age of 34 years, as opposed to the commonly seen occurrence at or round about puberty.

3. Case-II presented the combined characteristics of both trichoepithelioma and basal cell epithelioma. However, in view of the nature of the solitary lesion, occurring at the age of 70 years and the histological evidence relatively of larger areas showing the appearance of basal cell epithelioma and fewer horn cysts, abortive hair papillae and hair shafts, the lesion was regarded as a keratotic basal cell epithelioma. ✓

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