

Early Diagnosis of an Unusual Entity- Turban Tumour

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Research Article

Abstract: Cylindromas are benign neoplasms that commonly occur as solitary, slow-growing nodules on the head and neck regions, especially the scalp and face. Large variants usually composed of multiple coalescing tumors on the scalp and forehead ("turban tumors"). Despite its histologically benign behaviour, the disease process is distressing for the patients. We present our experience of this condition in which diagnosis was made on histopathology in the early course of disease, helping in early and better management of the patients.

Keywords: Turban tumour.

Introduction:

Dermal cylindromas are usually benign neoplasms of the eccrine sweat glands. The tumor usually occurs in the sixth decade of life [1]. There is no sex predilection. It presents most commonly on the head, neck, or scalp as slowly growing, pink to purple, solitary or multiple, smooth surfaced nodules, which can rarely grow and coalesce to produce the characteristic turban-like mass (turban tumor) [2].

Cylindromas have also been reported at other extracutaneous sites including salivary gland, bronchus, lung and kidney. Brooke-Spiegler syndrome or familial cylindromatosis is a rare familial condition, which is inherited in an autosomal dominant fashion, with variable expression and penetrance. The syndrome is associated with the occurrence, generally from childhood or adolescence, of multiple dermal cylindromas. The lesions are widely distributed

not only on the head and neck, but also the trunk and extremities, and are occasionally associated with other dermal adnexal tumours such as spiradenoma [3].

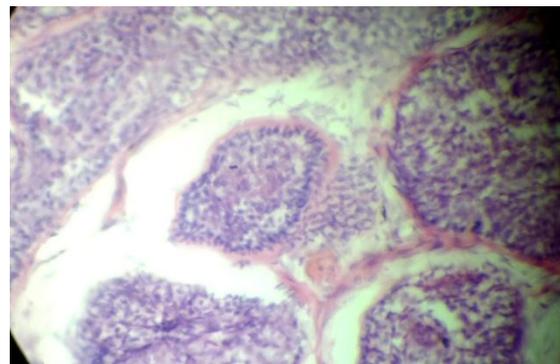


Figure01: (Low power view -10 X) dermal proliferation of basophilic lobules which molded together in a jigsaw puzzle configuration

Case report:

A 40 yr female patient was admitted to our rural hospital, with the complaint of multiple scalp swelling noticed since last 01 years, which were gradually increasing in size. The size of larger swelling at the time of admission was 3x2.5 cm. There was no history of trauma or fever. Examination revealed a firm, lobulated, nonpulsatile tumour with firm consistency. There were two more swelling which were small in size with similar examination findings. No

prior history of trauma or surgery was present. She underwent excision of the larger tumour. The specimen was then sent to pathology department for histopathological study. We received skin covered tissue mass measuring 3x2.5x2.5cm. On microscopic examination multiple sections studied show, dermal proliferation of basophilic lobules which molded together in a jigsaw puzzle configuration. At the periphery, there was a thick eosinophilic PAS+ basement membrane material (type IV collagen). The nests were also punctuated by small round collections of this material with similar staining qualities. [fig 01 &02] No evidence of malignant cytological features in the multiple sections studied. Hence we confirmed our diagnosis of turban tumour on the basis of presentation and histopathology.

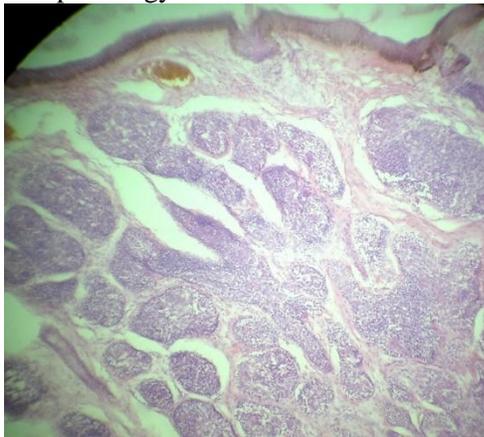


Figure02: (High power view -40 X) Lobules with Thick eosinophilic basement membrane material (type IV collagen) at the periphery.

Discussion:

In 1842 Ansell' first described an entity characterized by a striking clinical picture of multiple disfiguring tumours located on the face and scalp, which rapidly recurred after excision, had a distinctive histology, and showed familial occurrence.[04] Cylindromas are benign neoplasms that commonly occur as solitary, slow-growing nodules on the head and neck regions, especially the scalp and face. Large variants usually composed of multiple coalescing tumors on the scalp and forehead ("turban tumors").The frequency of (multiple) eccrine dermalcylindroma is still uncertain, although most authors consider it rare. The

primary histological features of cylindromata are cystic and duct-like structures with cords of basophilic cells and a prominent acidophilic hyaline basement membrane. Microscopic examination usually shows multiple foci of closely clustered epithelial cells with uniform size and shape and a palisading arrangement within nodules surrounded by a hyaline wall. The oval to round islands of basophilic cells may have the appearance of an assembled jigsaw puzzle.[05]

The tendency to produce multiple cylindromas often is inherited as an autosomal dominant trait.[06] Hereditary cylindromatosis is a rare autosomal dominant disease that is characterized by the development of multiple benign adnexal neoplasms, including cylindromas, spiradenomas, sebaceous epitheliomas, and trichoepitheliomas.[07]

The differential diagnoses of tumors on the scalp may include benign growths, such as trichilemmal cyst, which is composed of squamous epithelium without a granular layer with abrupt homogeneous keratinization. In addition, malignant tumors, such as metastatic carcinomas may spread to the skin via blood or lymphatics. The most common sources are breast, colon, and bronchial carcinomas, as well as metastatic melanoma. In skin metastases, the epidermis is usually uninvolved and biopsy shows a nodular proliferation of malignant cells [08]. Proliferating tricholemmal tumor is a benign tumor originating from the outer root sheath of a hair follicle. It is usually a solitary lesion and most commonly occurs in elderly women. Although considered biologically benign, it may be locally aggressive. In rare instances, malignant transformation has been reported, evidenced by regional or distant metastases. [09]

To conclude we present our experience of this condition in which despite its histologically benign behavior, the disease process is distressing for the patients and diagnosing it on histopathology in the early course of disease, help in early and better management of the patients.

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