

# Eccrine cylindroma of the face and scalp

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## ABSTRACT

Cylindroma is a benign skin adnexal tumor of eccrine differentiation. Clinically, they are disfiguring lesions that can mimic a malignancy. Cylindromas can be single or multiple and commonly involve the scalp (turban tumor) and face. Multiple cylindromas can have a syndromic association as seen in Brooke–Spiegler syndrome and familial cylindromatosis. We present a case of non familial cylindroma of the face that clinically mimicked a basal cell carcinoma. The histopathology was confirmatory. Herein we also highlight the utility of a simple and inexpensive histochemical stain Periodic Acid Schiff in supporting the diagnosis.

**Key words:** Brooke–Spiegler syndrome, cylindroma, periodic acid Schiff, PAS

## INTRODUCTION

Cylindroma is a benign skin adnexal neoplasm that presents in the head and neck region. The tumor may be single or multiple,<sup>[1]</sup> and is commonly located on the scalp. Histopathology is diagnostic with characteristic ‘jigsaw puzzle’ architecture and features of ductular differentiation. We present one such case.

## CASE REPORT

A 58-year-old man presented to the outpatient department with the complaints of a painful swelling over the right side of the face and scalp that gradually increased in size since two years. On examination, the facial swelling appeared bosselated and fungating, with surface ulceration [Figure 1]. Our clinical diagnosis was basal cell carcinoma. A trucut biopsy was done from the face as well as the scalp nodule [Figure 2]. Histopathological examination from both the swellings were identical and revealed lobules of epithelial cells separated by thin acellular eosinophilic septae, which imparted a mosaic or “jigsaw” puzzle architecture on low power [Figure 3]. The lobules comprised of a peripheral layer of dark staining basaloid cells and pale staining cells in the center. Tubular lumina suggestive of ductular differentiation were present at multiple foci. The periodic acid Schiff (PAS) stain highlighted the acellular basement membrane-like material surrounding the lobules as well as

entrapped masses of hyaline material within the lobules [Figure 4], thereby supporting a diagnosis of eccrine cylindroma. There was no significant family history to support a diagnosis of Brooke–Spiegler syndrome or familial cylindromatosis.

## DISCUSSION

Eccrine cylindromas are uncommon benign skin adnexal tumors. The cell of origin is from the pluripotent stem cells in the folliculo-sebaceous-apocrine unit.<sup>[1]</sup> The tumor continues to incite the curiosity of dermatologists and dermatopathologists, due to its syndromic association as well as due to its unique histopathological appearance. Brooke–Spiegler syndrome is an autosomal dominant cutaneous tumor syndrome, which presents with multiple skin adnexal neoplasms including cylindromas, trichoepitheliomas, and rarely spiradenomas.<sup>[2]</sup> It results from mutation in a tumor suppressor gene *CYLD*.<sup>[3,4]</sup> The syndromic tumors have an early age of onset and are multiple. Multiple

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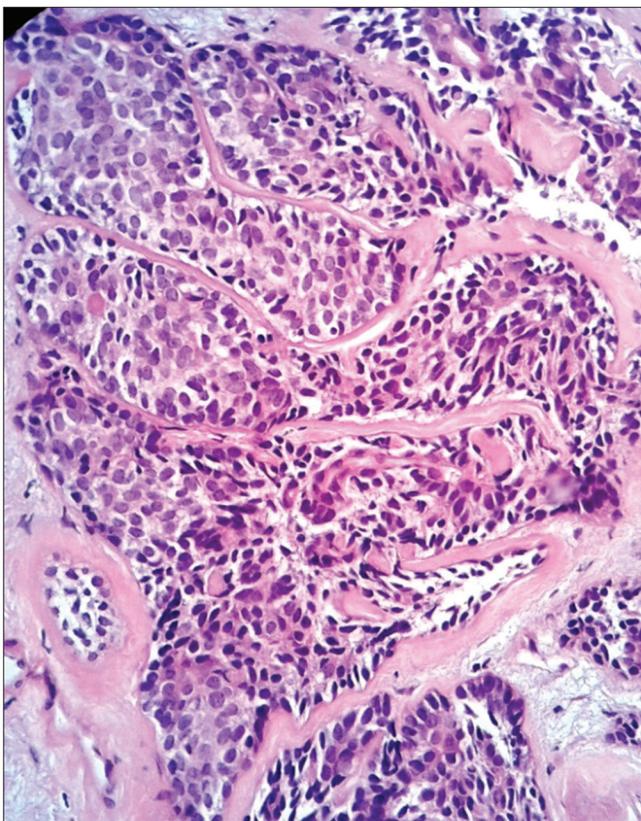
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**Figure 1:** Bosellated and ulcerated swelling on the face



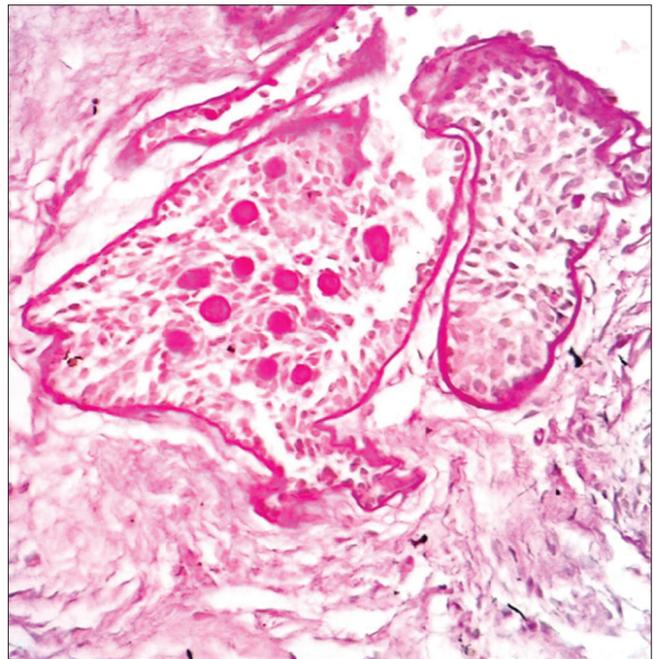
**Figure 2:** Scalp swelling



**Figure 3:** Low-power view (H and E ×10) showing islands of epithelial cells giving a jigsaw puzzle appearance

scalp lesions can cover the entire scalp like a turban, earning it the name of turban tumor. The lesions can be smooth and pedunculated or ulcerated and disfiguring. This necessitates biopsy to rule out malignant neoplasms such as basal cell carcinoma.

Histopathology is diagnostic with islands of cells separated by hyaline basement membrane material that is PAS positive. PAS-positive hyaline material in globules is also seen within the lobules.<sup>[6]</sup>



**Figure 4:** PAS stain highlighting the eosinophilic ribbons surrounding the islands and hyaline globules (H and E ×40)

In conclusion, eccrine cylindromas are rare tumors that can clinically masquerade malignancy. Biopsy clinches the diagnosis as the histopathology and histochemical staining properties are characteristic. Biopsy of more than one lesion is necessary in cases of multiple lesions. This in conjunction with a carefully elicited family history is mandatory to suggest or refute a syndromic association such as Brooke–Spiegler syndrome.

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### Conflicts of interest

There are no conflicts of interest.

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