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Cylindroma Mimicking Neurofibroma

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ABSTRACT

Introduction

Cylindroma is an appendageal tumour of the skin that is usually benign in nature, arising from pluripotent stem cells in the follicular-sebaceous-apocrine unit. Usually, multiple cylindromas are a component of Brooke-Spiegler syndrome (BSS) or occur only as skin lesions in familial cylindromatosis (FC). The site of the occurrence of tumours in this condition is the scalp.

Case Report

Here, we are reporting a case of multiple cylindromas in a 47-year-old female who presented with lesions on the face and scalp that were initially suspected to be neurofibroma as there were no features of Brooke-Spiegler syndrome or significant familial history pertaining to this presentation, but the histopathological diagnosis was suggestive of cylindroma.

Discussion

These are skin appedegeal tumours which commonly occur in the face and scalp. Their presentation resembles that of Neurofibromatosis and hence histopathological examination holds significance here. The main modality of treatment is excision of the lesions with management of the vascularity around the lesions, if present.

Keywords

Cylindroma; Pluripotent Stem Cells; Neurofibroma; Brooke Spiegler Syndrome; Scalp

Cylindromas are uncommon benign skin adnexal tumours.[1] The literature contains very few reports of these benign tumours. The benign tumour of the eccrine sweat glands is known as dermal cylindroma. The head and neck are the most frequently affected areas.[4] Mostly, they present as a single lesion, while multiple presentations are rare. The Brooke-Spiegler syndrome is frequently connected to several occurrences.[5] The autosomal dominant characteristic that causes Brooke-Spiegler syndrome is inherited irregularly.[6] Benign cylindromas occur more frequently in women, present as solitary or multiple lesions, and are autosomal in inheritance. Malignant cylindromas can occur in Brooke–Spiegler syndrome which is a rare autosomal dominant disorder characterized by the presence of various adnexal tumors such as multiple cylindromas, spiradenomas and trichoepitheliomas[7] although the probability of malignant transformation is very rare.[2] The tumours are generally asymptomatic, but a few patients can present with multiple, smooth, firm, pink to red, pedunculated nodules of various sizes that are painful.[8]

Case Report

A 47-year-old female presented with lesions over the face and scalp for the last 10 years. She initially consulted the department of dermatology and was then referred to the department of otolaryngology as the patient complained of a smelly nose and obstructed airflow in the left nasal cavity. On examination, multiple lesions on the face and scalp were present, the largest one being at the tip of the nose, causing difficulty in breathing, and on the forehead, causing a cosmetic blemish. The patient had previously undergone multiple excisions of the lesions under general anaesthesia. There was no history of similar presentations

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in the family. Surgical excision of the two lesions, one in the nasal cavity obstructing the airway and the other one on the forehead under general anaesthesia was done in our case. The patient was followed up for 6 months, and there was no further difficulty for the patient.



Fig.1. Clinical presentation of the patient with multiple Cylindromas on face, scalp

Although the clinical presentation hinted towards neurofibromatosis, on histopathological examination of the biopsy specimens from the nose and the forehead, macroscopic examination showed three grey-white to grey-brown soft tissue masses, each measuring 3.5 x 2.8 x 2.6 cm, 2 x 1.5 x 1.5 cm, and 1 x 1 x 0.6 cm. The cut section showed greyish-white and homogenous areas. Microscopic examination revealed a compact nest of basaloid cells with peripheral palisading arranged in the form of lobules like a zig-zag puzzle, and these nests of tumour cells were surrounded by a thick basement membrane. The tumour cells were of two types: undifferentiated epithelial cells and differentiated ductal cells.

Given the above-mentioned findings, although the patient presented with features resembling neurofibroma but with no lesions on the scalp and no features related to Brooke Spiegler syndrome, the histopathology showed features consistent with cylindroma. Due to this disparity between the clinical and histopathological data, we are reporting the case.

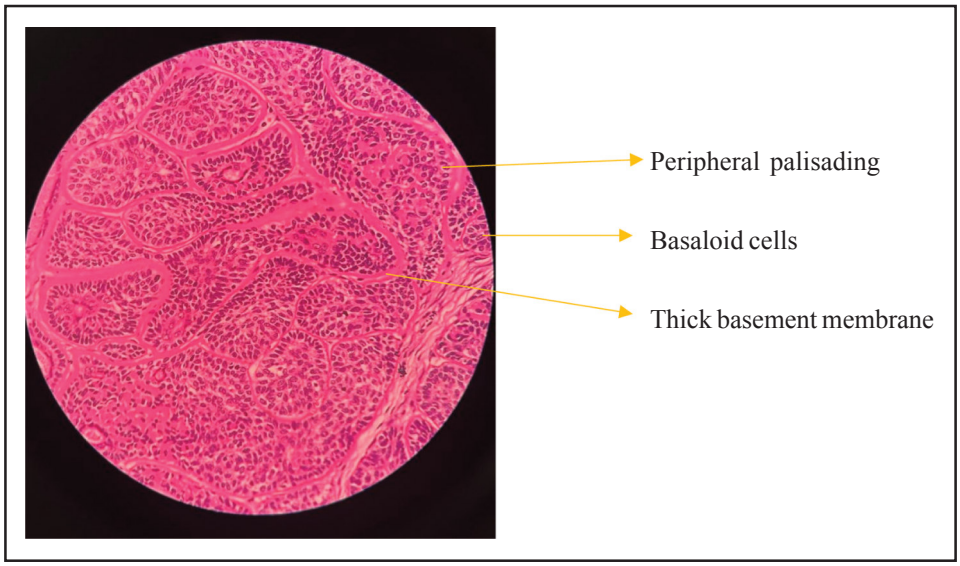


Fig. 2. Histopathological picture: Hematoxylin and Eosin stain- 40x: showing Basaloid cells with peripheral palisading

Discussion

Cylindromas, also known as cutaneous or dermal cylindromas, are unusual and usually skin appendage tumours that are benign, slow-growing lesions, although there have been rare cases of malignant transformation and even metastases reported.[4] They can be seen along with spiradenomas and trichoepitheliomas.[1]

The probability of disease manifestation is nine times higher in females than in males. Multiple cutaneous cylindromas can occur on the scalp and resemble a turban, thereby leading to it being referred to as a "turban" tumour[1].

Cylindromas can have either sporadic or familial modes of occurrence. Sporadically occurring cylindromas usually present as solitary lesions that are slow-growing, painless, round nodular lesions that commonly occur in the older age group.[1] Familial or syndrome-associated lesions are usually multiple, rapidly growing lesions that can ulcerate, become painful at later stages, and commonly occur at an earlier age.[7] Cylindromas that occur sporadically usually exhibit the expression of MYB-NFIB (v-myb avian myelobastosis viral oncogene homolog-nuclear factor I/B) fusion transcripts, where MYB is an oncogene that, when fused with the transcription factor gene NFIB, forms an oncoprotein that stimulates the neoplastic process.[1, 3]

Familial studies in patients who are suspected to have Cylindromatosis Syndrome or genetic testing looking for mutations in the CYLD (**CYLD Lysine 63 Deubiquitinase** which is a Protein Coding gene) cylindromatosis gene can be performed.[3]

The head and neck region has the highest likelihood of occurrence of lesions in Cylindromas, followed by the trunk and extremities, while the palms and soles are usually spared.[3]

Radiologic exams aid in finding the extent of the lesions that might occasionally involve the underlying osseous structures or are vascular in nature before formulating a further plan of care.

Although there is no definitive treatment for this condition owing to its genetic origin, early diagnosis and currently, multiple, recurrent excision of the lesions under local or general anaesthesia is the main modality of treatment in multiple cylindromas. Large lesions should be imaged to determine vascularity and involvement of surrounding tissues, which will determine the further mode of management. Pre-treatment embolization is preferably done in patients with large and multiple cylindromas to minimise intra-operative blood loss as the lesions can be vascular.[3] Laser ablation has also been tried in a few cases with temporary promising results, but it is not a curative solution.

Recently, a therapeutic attempt has been made to treat single cylindromas in Brooke Spiegler Syndrome with topical application varying concentrations of salicylic acid which acts by interfering with the NF-êB signalling pathway.[9]

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