Case Report

Dermal cylindroma of the scalp

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ABSTRACT

Dermal cylindroma is a benign neoplasm of the eccrine sweat glands, which presents in the head and neck area in majority of cases. In its most florid presentation, the entire scalp can be involved, leading to the descriptive label turban tumor. They most commonly occur on the head and neck as solitary or multiple tumors. Solitary cylindromas occur sporadically and typically are not inherited. Multiple tumors are observed in an autosomal dominantly inherited manner. When nodules enlarge and coalesce on the scalp, they form the distinctive turban tumor feature. We present a case of multiple form of dermal cylindroma, occupying the anterior scalp in a 38-year-old male patient.

Key words: Dermal cylindroma, scalp, turban tumor

Introduction

Cylindromas are appendage tumors of uncertain histogenesis. They may occur as solitary or multiple lesions. In case of multiple lesions numerous small papules and/or large dome-shaped nodules are present on the scalp like a turban, hence the name turban tumor. Scattered nodules are present occasionally on the face and in rare instances on the extremities.[1] Multiple cylindromas may be associated with other cutaneous and extracutaneous tumors.[2] We report a 38-year-old male patient who had multiple cylindromas on the anterior region of the scalp.

Case Report

A 38-year-old male patient presented with a long-standing history of multiple non-tender scalp swellings from 2.5 to 3.5 cm in diameter, with progressive growth over the past one year. The skin over swellings was normal [Figures 1 and 2]. Clinical examination revealed multiple nodular lesions coalesced on the anterior region of the scalp, which was solid and movable from the underlying surface. Routine blood investigations were normal. Excisional biopsy was planned under general anesthesia followed by primary closure [Figures 3 and 4]. The excised specimen was sent for histopathological examination, which showed lobules of epithelial cells arranged in a jigsaw or mosaic pattern. Prominent red basement membrane-like structure encircles the tumor lobules. Each lobule shows a peripheral lining by dark basaloid cells and an inner larger and paler zone of cells, which was suggestive of dermal cylindroma [Figure 5]. Follow-up was done regularly for one year and there were no signs of recurrence [Figure 6].

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Discussion

Cylindroma is an uncommon tumor, affecting females about twice as frequently as males. It is often familial and its inheritance is determined by an autosomal dominant gene. It has been reported to follow X-ray epilation *of* the scalp.[3] In cases with solitary lesion, there is no family history association.[1] The tumors are frequently multiple, smooth, firm, pink to red in color and often somewhat pedunculated. Some tumors may be painful. The commonest site is the scalp and adjacent skin. The tumors may be almost hairless when pedunculated, but the smaller lesions form dermal nodules with little loss of hair over them. A proportion of lesions occur on the face and neck, in less than 10% of cases they are situated on the trunk and limbs.[3]

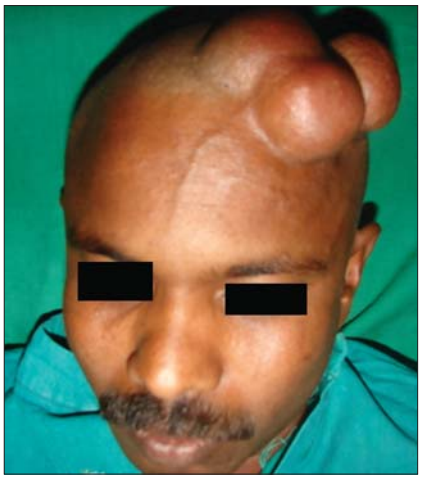

Figure 1: Preoperative frontal view of the patient showing multiple swellings on the anterior scalp

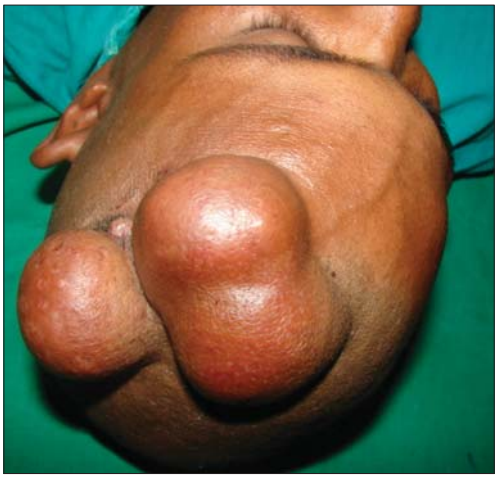

Figure 2: Preoperative superior view of the patient showing multiple swellings on the anterior scalp

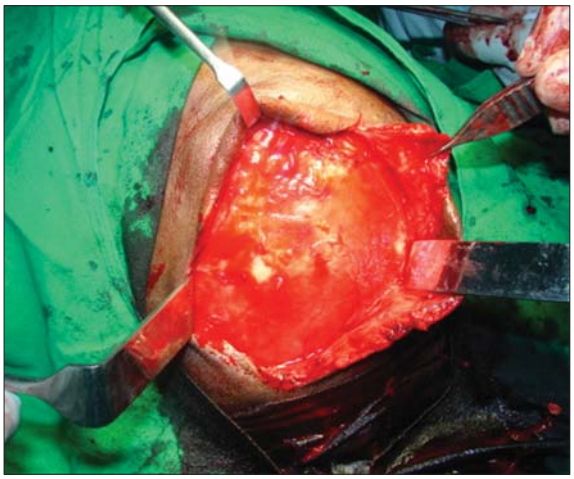

Figure 3: Intraoperative view after surgical excision of multiple swellings

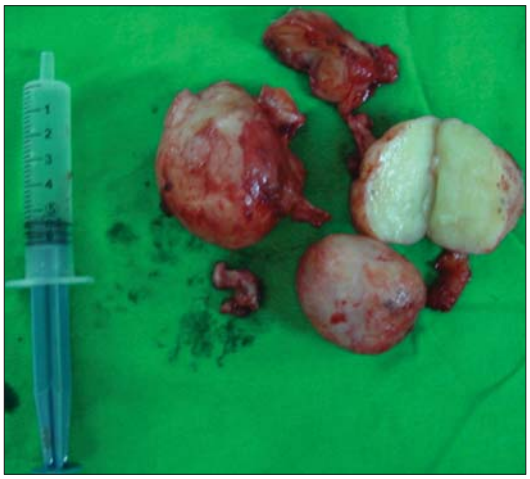

Figure 4: Excised specimen showing multiple swellings with a glistening, smooth, faintly lobulated surface. Note the cut section of the tumor showing solid mass inside

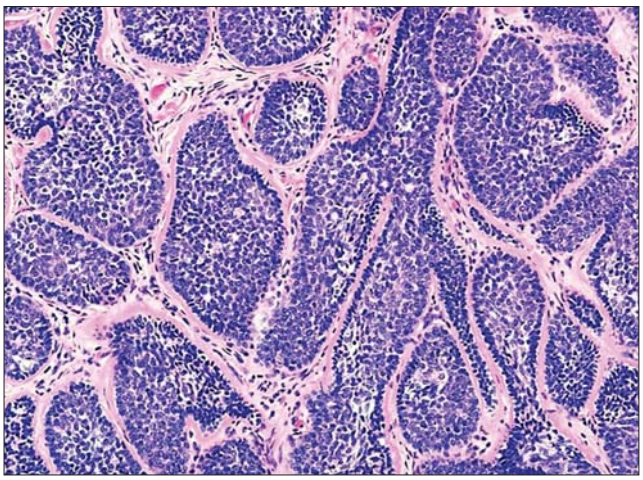

Figure 5: Histopathology showing lobules of epithelial cells arranged in a jigsaw or mosaic pattern. Prominent red basement membrane-like structure encircles the tumor lobules. Each lobule shows a peripheral lining by dark basaloid cells and an inner larger and paler zone of cells which was suggestive of dermal cylindroma

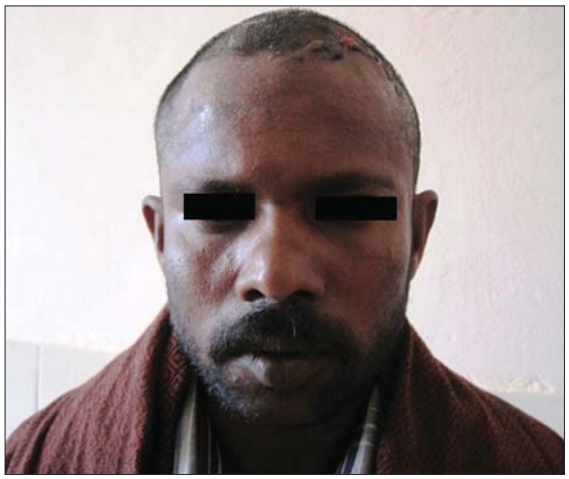

Figure 6: Postoperative frontal view of the patient after one week

Cylindromas are appendage tumors previously thought to be of apocrine differentiation. While phenotypic features differ between cylindromas and spiradenomas, recent studies have shown immunohistological and cytomorphological overlap, with both tumors exhibiting apocrine, eccrine, secretory, and ductal features. Therefore, the cellular origin of cylindromas

remains unknown.[4,5] Cylindromas are most likely a very primitive sweat gland tumor differentiating toward either the eccrine or apocrine line. The cause of sporadic, solitary cylindromas is largely unknown; however, genetic studies of sporadic cylindromas show loss of heterozygosity at and around the CYLD locus in a substantial number of cases, suggesting that this gene also plays a role in the development of sporadic tumors. Familial cylindromatosus is inherited in an autosomal dominant fashion, and the responsible gene, CYLD, is located on band 16 q 12-13.[6,7] Tumors exhibit loss of heterozygosity, implicating the gene as a tumor suppressor gene.

Histopathological findings suggest that cylindroma is a dermal tumor without attachment to the epidermis. The lesion is composed of numerous oval and polygonal nests molded into a jigsaw-like pattern. Masses of epithelial cells are surrounded and penetrated by a hyaline sheath closely resembling a basement membrane.[8] This sheath separates the tumor from the dermal mesenchyme, yet does not interfere with tumor growth and proliferation. A lack of lymphoid tissue is a histological feature that differentiates cylindromas from spiradenomas. Spiradenomas show a unique prominent presence of lymphocytes. Cylindromas, on the other hand, demonstrate a large number of prominent dendritic cells that most likely represent Langerhans cells that permeate the tumor.[8,9] Surgical management for solitary lesions, the treatment of choice is surgical excision. Other treatments include electrodesiccation/curettage and cryotherapy. For small cylindromas, the carbon dioxide laser may be used.[10] Retamar *et al.* used carbon dioxide laser to treat facial trichoepitheliomas in twp patients, with good results.[11] Multiple cylindromas usually require extensive plastic surgery that may be obviated by progressively excising a group of nodules in multiple procedures. Follow-up care of patients with multiple cylindromas is recommended because of the tendency for new lesions to develop and to prevent the risk of malignant degeneration.

References

1. Hashimoto K, Lever FW. Tumours of the skin and appendages. In : Dermatology in General Medicine, (Fitzpatrick TB, Arndt KA, Clark HW, Jn, *et al* eds). New York: McGraw Hill, 1971; 452.
2. Raman M, Singh N. Familial multiple cylindromas. Indian J Dermatol Venereol Leprol 1991;57:104-6.
3. Mackie RM. Tumours of the skin appendages. In: Champion RH, Burton JL, Ebling FJG, editors. Textbook of Dermatology. 5th ed. Oxford: Blackwell Scientific; 1992. p. 1520-1.
4. Stegmeier F, Sowa ME, Nalepa G, Gygi SP, Harper JW, Elledge SJ. The tumor suppressor CYLD regulates entry into mitosis. Proc Natl Acad Sci U S A 2007;104:8869-74.
5. Massoumi R, Paus R. Cylindromatosis and the CYLD gene: New lessons on the molecular principles of epithelial growth control. Bioessays 2007;29:1203-14.
6. Fukuda M, Hiroi M, Suzuki S, Ohmori Y, Sakashita H. Loss of CYLD might be associated with development of salivary gland tumors. Oncol Rep 2008;19:1421-7.
7. Parren LJ, Bauer B, Hamm H, Frank J. Brooke-Spiegler syndrome complicated by unilateral hearing loss. Int J Dermatol 2008;47 Suppl 1:56-9.
8. Carlson RM, Haddad L, Pui JC. Brooke-Spiegler syndrome with associated pegged teeth. Cutis 2008;82:345-9.
9. Ishihara M, Mehregan DR, Hashimoto K, Yotsumoto S, Toi Y, Pietruk T, *et al.* Staining of eccrine and apocrine neoplasms and metastatic adenocarcinoma with IKH-4, a monoclonal antibody specific for the eccrine gland. J Cutan Pathol 1998;25:100-5.
10. Rallan D, Harland CC. Brooke-Spiegler syndrome: Treatment with laser ablation. Clin Exp Dermatol 2005;30:355-7.
11. Retamar RA, Stengel F, Saadi ME, Kien MC, Della Giovana P, Cabrera H, *et al.* Brooke-Spiegler syndrome - report of four families: Treatment with CO2 laser. Int J Dermatol 2007;46:583-6.

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