

Cylindroma of the scalp

Sir,

A 56-year-old woman presented with a long-standing history of slow-growing multiple scalp nodules. Her daughter and aunt also had a history of similar cutaneous lesions. Physical examination revealed multilobulated, large, epiphytic pink nodules, 2.5 cm by 6 cm in the greatest dimension, on the scalp [Figure 1]. The nodules were painless, solid, and movable from the underlying surface. Results of the routine laboratory examinations were normal. The histopathology of the scalp nodule revealed a well-defined dermal lesion composed of islands and cords of dark basaloid cells arranged in a jigsaw puzzle-like pattern and separated by hyaline basement membrane material, which was suggestive of cylindroma [Figure 2]. This patient underwent staged procedures, with debulking surgery followed by contouring with split-thickness skin graft. After 1 year, she was lost to follow-up.

Cylindromas are benign skin appendage tumors. They can be seen in conjunction with spiradenomas and trichoepitheliomas, and this condition is known as Brooke-Spiegler syndrome (BSS).^[1] BSS is a rare autosomal dominant disorder caused by mutations of the cylindromatosis gene (CYLD) located at 16q12-q13.^[2] The penetrance of the gene has been estimated to be between 60% and 100%.^[3] Histologically, the tumors are circumscribed, nonencapsulated dermal nodules composed of islands and cords of basaloid cells surrounded by a thick, hyalinized, periodic acid-Schiff (PAS)-positive basement membrane. The cells are arranged in an interlocking "jigsaw-puzzle"-like architecture.^[4]

Treatment options in BSS are mainly for cosmetic concern. For solitary tumors, surgical excision is the treatment of choice. Other forms of therapy include electrodesiccation, curettage, cryosurgery, and excision with the use of carbon dioxide or neodymium-doped yttrium aluminum garnet; Nd:Y3Al5O12 (Nd:YAG) laser.^[5,6] Topical 5% imiquimod cream has been advocated as a useful treatment.^[7] Recently, it was suggested that targeting nuclear factor (NF)- κ B in cylindroma therapy may be of therapeutic potential in patients with multiple tumors in the course of BSS. In one case of familial trichoepitheliomas, significant improvement was achieved with a combination of adalimumab and aspirin. This combination was chosen in order to block tumor necrosis factor alpha (TNF α)-induced activation of NF- κ B at two different levels of the signaling pathway.^[8] Several therapeutic agents



Figure 1: Physical examination revealed multilobulated large epiphytic pink nodules, 2.5 cm by 6 cm in the greatest dimension, on the scalp

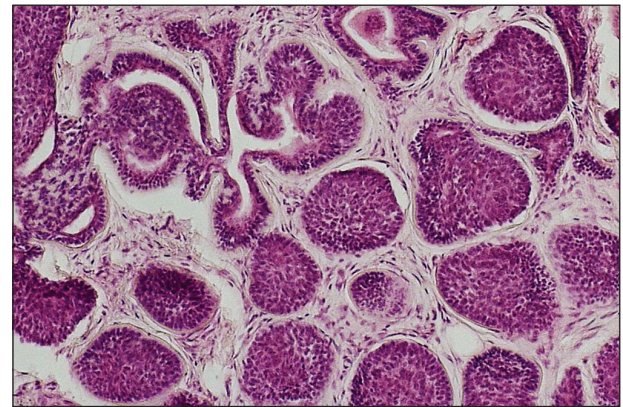


Figure 2: The histopathology of the scalp nodule revealed a well-defined dermal lesion composed of islands and cords of dark basaloid cells arranged in a jigsaw puzzle-like pattern

including sodium salicylate, cyclooxygenase (COX)2 inhibitors (i.e., celecoxib), corticoids and prostaglandins as well as phytomedicinal products inhibit NF- κ B activity and thus, may be of benefit in BSS.^[9] However, their possible clinical efficacy remains to be elucidated.

AUTHOR'S CONTRIBUTION

MRM contributed in the conception of the work, conducting the study, and revising the draft. NM and NSA designed the study and were responsible for the overall study management. All authors approved the final version of the manuscript, and agreed with regard to all aspects of the work.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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DOI:

10.4103/1735-1995.170638

How to cite this article: Mogaddam MR, Maleki N. Cylindroma of the scalp. *J Res Med Sci* 2015;20:923-4.