

flow images before and after treatment as a complement to clinical observation.

The present study revealed considerable agreement between the 2 observers, which in the case of X flow was 100%. Therefore, we believe that X flow mode provides a better evaluation of PWS, since it is more efficacious for detecting small-caliber, low-flow vessels.

Conflicts of Interest

The authors declare that they have no conflicts of interest.

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A Simple and Effective Method for Treating Cyndromas in Brooke-Spiegler Syndrome[☆]



Método simple y efectivo para el tratamiento de los cilindromas en el síndrome de Brooke-Spiegler

Dear Editor:

The genetic skin disease known as Brooke-Spiegler syndrome (BSS) is a rare autosomal dominant condition with more than 50 cases reported to date.¹ BSS predisposes to benign adnexal tumors that generally first appear in adulthood and involve the folliculo-sebaceous-apocrine unit. It is characterized by multiple cyndromas, trichoepitheliomas, and, occasionally, spiradenomas.² The tumors appear simultaneously, as in BSS, or in isolation, as in familial cyndromatosis and multiple familial trichoepithelioma. Consequently, they seem to have a common origin in a pluripotent cell in the folliculo-sebaceous-apocrine unit, with several phenotypic variants.^{3,4}

Various mutations have been identified in the tumor suppressor gene *CYLD* on chromosome 16q12-913, thus indicating an association with BSS. *CYLD* inhibits the tumor necrosis factor α pathway, diminishing expression of nuclear factor κ B (NF- κ B), a transcription factor that in turn regulates various antiapoptotic genes involved in the

proliferation of adnexa. A loss of the suppressive function of *CYLD* leads to greater resistance to apoptosis and, consequently, the appearance of these tumors.^{3–5}

Clinically, cyndromas present as pink nodules on the scalp and behind the ears (Fig. 1). Trichoepitheliomas appear as papules or small translucent or flesh-colored nodules and are located mainly in the center of the face, especially in the nasal region. Spiradenomas, on the other hand, usually present as solitary bluish or flesh-colored nodules that are often painful.²

The emotional impact of recurrence of these lesions on the face and scalp makes treatment essential. Our literature search revealed various options, such as electrocautery, dermabrasion, radiofrequency, cryotherapy, CO₂ laser,⁶ radiotherapy (in special cases),⁷ and, recently, enucleation.⁸ The treatment recommended for solitary cyndromas is surgical removal, whereas partial or total skin grafting has been proposed for multiple lesions.⁹ NF- κ B



Figure 1 Cyndroma on the scalp.

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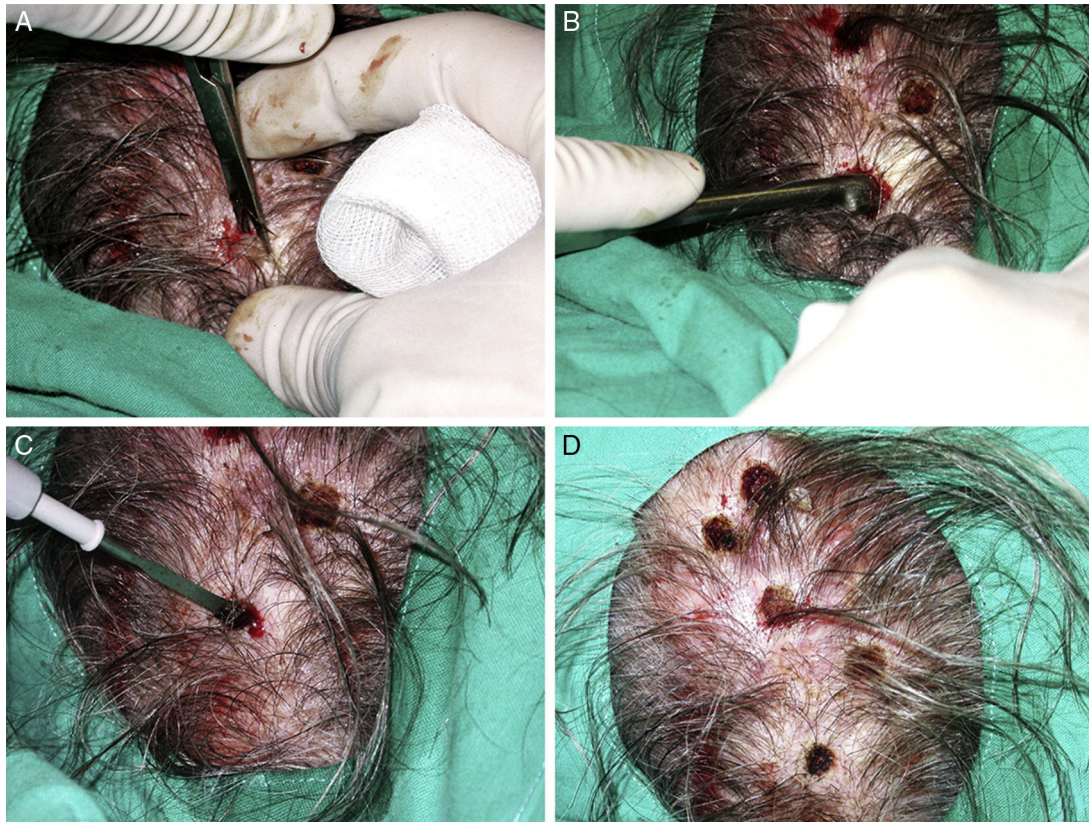


Figure 2 A, Incision and debulking. B, Curettage. C, Electrocautery to stop bleeding. D, Immediate postoperative findings.

inhibitors, such as acetylsalicylic acid and its derivatives, may prove useful both for prevention and for treatment of cylindromas when applied locally.¹⁰ Therefore, treatment should be tailored depending on the patient, size of the lesion, number of lesions, therapeutic options available, and the dermatologist's experience.

We report the case of an 80-year-old woman with BSS who presented with multiple cylindromas on the scalp that had recurred since adolescence, together with occasional spiradenomas. Her family history was remarkable in that one of her daughters, aged 49 years, had had numerous facial trichoepitheliomas since puberty. She has been followed regularly in our department for the last 10 years, and several lesions have been removed using elliptical excision and direct closure. Given the high number of lesions and the excisions performed, we sought an alternative that would make for more efficient surgery and benefit the patient by reducing the number of trips to the hospital. Five years ago, we began to perform a technique that combined cytoreduction and subsequent curettage of the growths. Our technique is minimally invasive and efficacious.

After administration of local anesthetic (mepivacaine 2%), an incision is made with a scalpel or scissor cut to enable resection of most of the tumor mass and create a portal of entry for the next step. After debulking, the remaining lesional tissue is removed by intense curettage, and bleeding is stopped using electrocautery. The resulting wound heals by secondary intention (Fig. 2). Complete

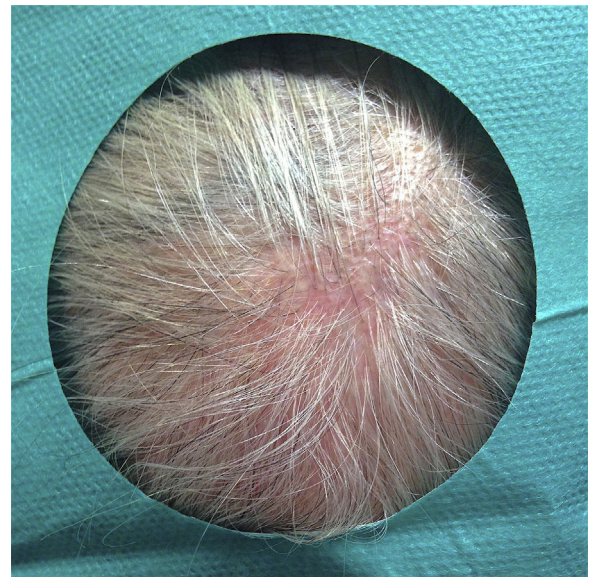


Figure 3 Outcome 12 months after surgery.

scarring is achieved within approximately 14 days based on daily dressing with antiseptic and antibiotic ointment. The wound progresses without complications after surgery, and the cosmetic outcome is favorable at the 12-month visit (Fig. 3). Regular follow-up has revealed a reduction

in the number of cylindromas and a rate of recurrence of the lesions removed lower than 25%. When the lesion did relapse, the growth was always smaller than the original lesion, thus facilitating surgery.

Our technique enables us to reduce the volume of the lesions by eliminating several growths during the same procedure. This is particularly advantageous in patients who must undergo repeated procedures. It also enables us to concentrate on the emergence of new lesions, since cylindromas rarely recur after removal using this technique. This is because elimination of the lesions by combining several techniques in one procedure (eg, debulking with scissors, followed by curettage of the wound bed, and subsequent use of an electric scalpel) enables almost total necrosis of any remaining cells and destruction of the hair follicles, which are a potential source of new tumors.

Conflicts of Interest

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