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A Case of Malignant Transformation of Solitary Recurrent Cylindroma on Scalp

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A 78-year-old male presented with an asymptomatic pinkish multi-nodular mass on his frontal scalp. The lesion had recurred twice after incomplete surgical excision. Initial punch biopsy was diagnosed with cylindroma. He revisited after one year with exophytic enlargement of the mass, and two staged Mohs micrographic surgery identified well-differentiated malignant cylindroma. Histopathology in the lower dermis and periosteal layer showed atypical cells with mitosis and hyperchromatic nucleoli with increased Ki-67 index of 10% to 30%. The postoperative wound was successfully treated with negative wound pressure therapy (NPWT) and secondary intentional healing. We report this case showing malignant transformation of solitary cylindroma, and good result for secondary intention healing using NPWT for postoperative defect.

Keywords: Adenoid cystic carcinoam, Malignant adenoid cystic carcinoma, Mohs surgery, Negative-pressure wound therapy

INTRODUCTION

Cylindroma is a benign skin appendage neoplasm that typically presents as a single smooth nodule on the head and neck region in adults[1,2]. Malignant cylindromas have rarely been reported, most of which are derived from multiple cylindromatosis so that solitary well-differentiated cases are uncommon[1-4]. Herein, we report an interesting case of malignant transformation of a recurrent well-differentiated solitary type cylindroma treated with secondary intentional healing assisted by negative wound pressure therapy (NPWT).

CASE REPORT

A 78-year-old male presented with an asymptomatic pinkish multi-nodular mass on his frontal scalp that reportedly emerged several decades earlier (Fig. 1A). The lesion recurred twice after incomplete surgical excision without pathological confirmation. There was no known family history of similar disease, trauma history, or current medical history. Skin biopsy demonstrated a homogeneous basaloid cell nest arranged in a jigsaw pattern, composed of central cells with eosinophilic cytoplasm and peripheral cells with basophilic nuclei and scant cytoplasm. The patient was diagnosed with a cylindroma but refused further treatment due to economic circumstances.

He returned after one year, by which time the mass that had increased in size to 5.0 cm×2.5 cm (Fig. 1B). At this point, he was willing to undergo surgery. In the first stage of Mohs micrographic surgery, the deeper margin was positive for tumor cells. The second stage of excision was performed deep down to the periosteum, and a clear resection margin was confirmed. The final surgical defect was 7.2 cm×4.5 cm and was successfully treated with secondary intentional healing using a NPWT device (Fig. 2A~C). The device used in this patient was PICO single use NPWT system (Smith and Nephew, London, UK). During the first month, the patient received the PICO NPWT dressing twice a week, and after a month, sufficient wound healing was observed (Fig. 2D).

Histopathologically, the upper dermis of the main tumor mass was similar to that seen in the initial biopsy. However, the lower dermis and periosteal layer had some stromal infiltra-

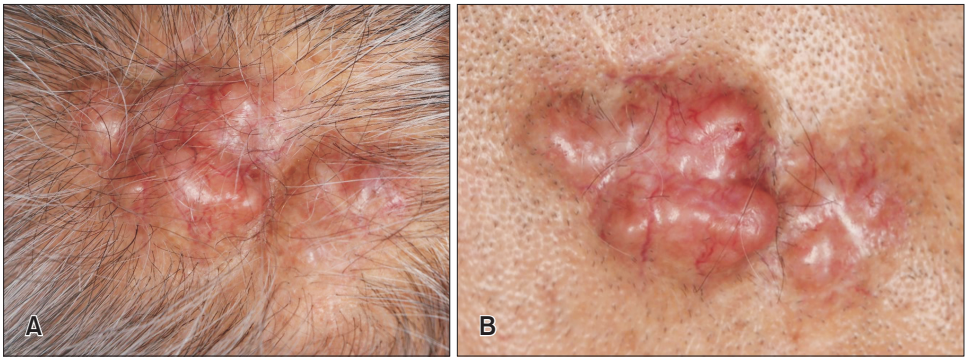

Fig. 1. Clinical photograph of the lesion. (A) Initial photograph showed erythematous nodular mass on scalp. The biopsy was taken at the most protruded lesion. (B) Clinical photograph of the The final surgical defect was filled with a sterile compressed sponge material, and secondary intentional healing was assisted by negative wound pressure therapy.

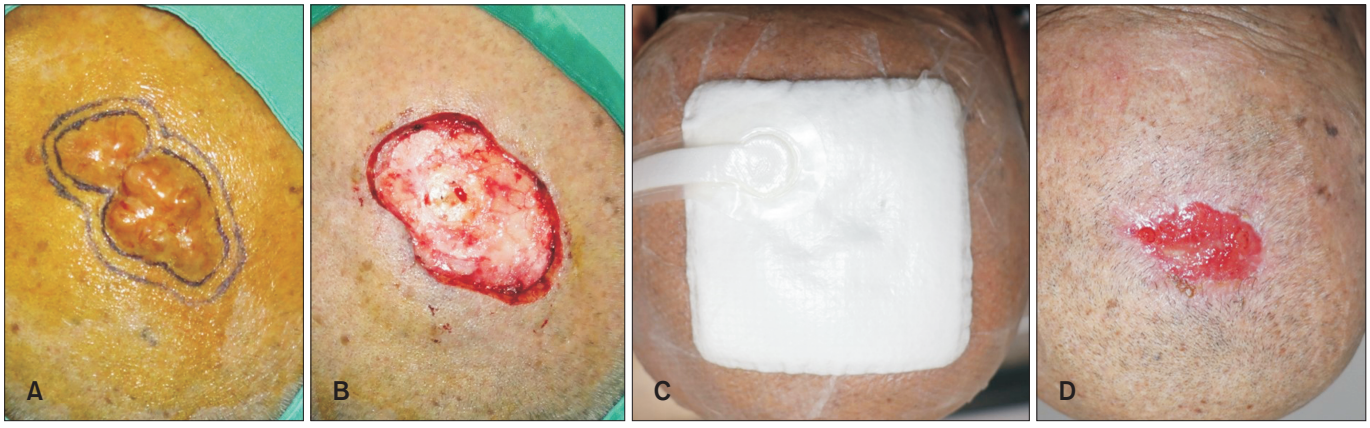

Fig. 2. Clinical progress of postoperative wound. (A, B) The surgical defect was 7.2 cm×4.5 cm after two stages of Mohs surgery. (C) The surgical defect was occluded with negative wound pressure therapy (NPWT) dressing device. (D) The wound showed gradual improvement throughout 7 weeks of NPWT dressing, and there was no recurrence observed.

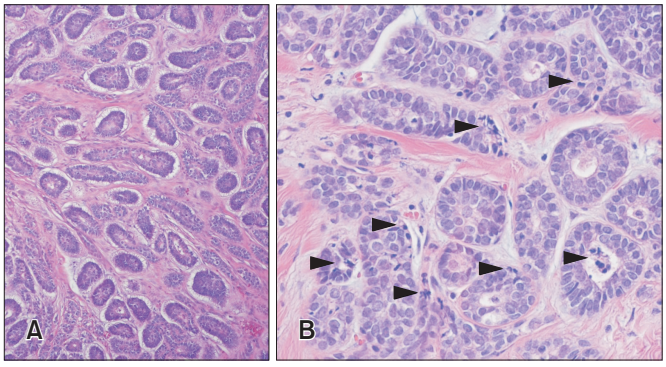

Fig. 3. Histopathologic image of the lesion with H&E staining. (A) Basaloid cell nests and focal stromal infiltration (H&E, original magnification ×100). (B) Lower dermis shows loss of jigsaw pattern, and a few atypical cells with mitosis and pleomorphic nuclei (arrowheads) (original magnification ×400).

tion of cell nests with a loss of the jigsaw pattern (Fig. 3A). In high magnification images, atypical cells with mitosis, apoptosis, and hyperchromatic nucleoli were identified (Fig. 3B). The Ki-67 index increased to 10%~30%, and special staining was performed to rule out other skin appendageal malignancies. These stains were positive for p63 and Bcl-2, focally positive for C-kit and epithelial membrane antigen, and negative for carcinoembryonic antigen. At the five-month follow-up visit, there was no apparent local recurrence.

We received the patient's consent form about publishing all photographic materials.

DISCUSSION

Cylindroma is a rare benign skin appendageal tumor that commonly presents on the head and neck area as a small solitary nodular lesion averaging 0.2 to 0.6 cm in size. Malignant cylindromas—also known as malignant spiradenomas—are large and rapidly growing tumors and normally, malignant transformation more commonly occurs in multiple cylindromatosis rather than in the solitary type. Although some

of these tumors are sporadic, a genetic background such as a *CYLD1* gene mutation may be a causal factor[1,2]. Some studies reported that external factors such as incomplete excision, frequent trauma, chronic irritation, or radiation therapy to contribute to malignant transformation of the tumor[4-6]. In the present case, the lesion was solitary type and the patient had no family history of such disease, but received two times of incomplete excision.

Histopathologically, malignant cylindromas demonstrate cellular pleomorphism, mitosis, necrosis, stromal invasion with loss of the jigsaw pattern and biphase distribution. A review of the literature in PubMed showed fewer than 40 well-documented cases of malignant cylindromas, and only a few cases of well-differentiated malignant cylindromas were reported[1,2,5]. Our patient had a clinical history of tumor recurrence after excision and exhibited an exophytic growth suggestive of malignancy. Histopathologic examination of the deeper margin showed stromal infiltration of the tumor nest and atypical cells with pleomorphic nucleoli and apoptotic features. However, the mitotic and pleomorphic cells were only focally observed, and the Ki-67 index was moderate at 10%~30%. Therefore, the patient was diagnosed with a well-differentiated malignant cylindroma[1,6].

The patient's resection margin was pathologically confirmed as clear and successfully treated using NPWT. Despite of alternative treatment choices such as local flap, skin graft and radiotherapy, the patient preferred secondary intentional healing because these procedures would require an additional large incision, delicate management of the graft site, and frequent visits[7,8]. For surgical defect of skin tumors, dermal substitutes—including NPWT—have been used instead of reconstructive surgery, especially with confirmed clear margin of postoperative wound[9,10]. To assist with secondary intentional healing of the large scalp defect, NPWT was used in this case.

In conclusion, the patient was diagnosed with a well-differentiated malignant cylindroma arising from a solitary-type benign cylindroma. Although pathology showed only focal malignant features, the patient had a clinical history of exophytic enlargement and several recurrences. This case presented a rare case of malignant transformation from solitary cylindroma, suggesting not only the importance of close observation of such lesion but also the complete excision based on pathological confirmation.

CONFLICTS OF INTEREST

The authors have nothing to disclose.

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