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Case Report

Giant Mushroom-Like Cutaneous Cylindroma of the Head

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Abstract

BACKGROUND: Dermal cylindroma is a rare benign skin tumour.

CASE REPORT: We report a giant stalked, mush-room-like cylindroma of the head-and-neck region in a 73-year-old female patient. A tumour was surgically removed, and the defect could be closed by a cheek transposition flap.

CONCLUSION: The mushroom-like growth pattern has yet not been described for cylindroma.

Introduction

Dermal cylindroma is a rare benign apocrine adnexal tumour with a female predominance. Most patients are Caucasians. The slow-growing tumours are usually small. The most common localisation is on the scalp although tumours off the head and neck region have occasionally been observed. Malignant transformation is rare and preferential in patients with multiple tumours [1] [2].

Histologically these tumours consist of rounded islands of basaloid cells which are arranged in a “jigsaw puzzle”-pattern. These islands have a Grenz zone to the epidermis. There is a cellular dualism with the palisading peripheral lining of smaller cells with more hyperchromatic nuclei and an inner population with larger, more differentiated pale cells and nuclei and small duct-like structures. Hyaline droplets can be present. A tumour is surrounded by a hyalinized sheath [3].

On immunohistology, expression of lysozyme and alpha l-antichymotrypsin, cytokeratin, epithelial membrane antigen (EMA) and EGF-receptor has been demonstrated. The presence of intermingled cells with coexpression of keratin and vimentin argues for a partial myoepithelial-like differentiation [4].

The genetics of sporadic and inherited cylindromas demonstrate a molecular heterogeneity. A central role has *MYB* which is an oncogene. *NFIB* is a transcription factor gene. When both of these genes fuse they form an oncoprotein. MYB-NFIB fusion oncoprotein and activated or overexpressed *MYB* are acted as a driver for the proliferation of cutaneous cylindromas [5].

Multiple cylindromas are characteristic in patients with Brooke-Spiegler syndrome, an autosomal dominant disease characterised by the development of multiple adnexal cutaneous neoplasms such as cylindroma, spiradenoma, spiradenocylindroma, and trichoepithelioma. Of these patients, 40% to 85% carry germline mutations in the tumour suppressor gene *CYLD*, but lack *MYB-NFIB* fusion transcripts. However, *MYB* activation has been

demonstrated in 69% of tumours [6].

Sporadic cylindromas may be associated with *MYB-NFIB* fusion transcripts or *MYB* activation in the absence of such fusions [7]. The treatment of choice is complete surgical excision [8] [9] [10].

Case report

A 73-year-old woman presented to our hospital with a slow-growing tumour on the face. A tumour had been there for more than 10 years. The patient had no remarkable medical history, no immunosuppression, and no previous cancer. She took no medications.

On examination, we observed a Caucasian woman with Fitzpatrick's skin type II. She had a 7 cm large, stalked tumour with a mushroom-like growth (Figure 1). A tumour was painless. No enlarged lymph nodes were detected. There was no family history of tumours.

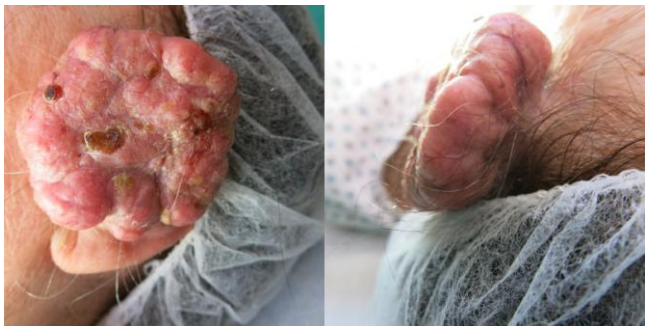

Figure 1: Clinical presentation of the giant cylindroma with a mush-room-like growth pattern

We suspected a non-melanoma skin cancer (keratoacanthoma-like squamous cell carcinoma) and suggested complete surgical removal by delayed Mohs surgery.

A tumour was excised in local anaesthesia. The stalk had pronounced vascularity that needed efficient hemostasis with bipolar tweezers. After resection, the defect was closed by a cheek transposition flap (Figure 2). Healing was unobtrusive.

Histopathology described a dermal tumour with a jigsaw appearance with nests of basaloid cells surrounded by a dense hyaline basement membrane. The basaloid cells with scant cytoplasm and dark nuclei palisaded around the edge of the nests. Larger cells with moderate eosinophilic cytoplasm and lighter staining nuclei were at the centre of the nests. Some of the larger cells were lumen-forming in a ring-like pattern.

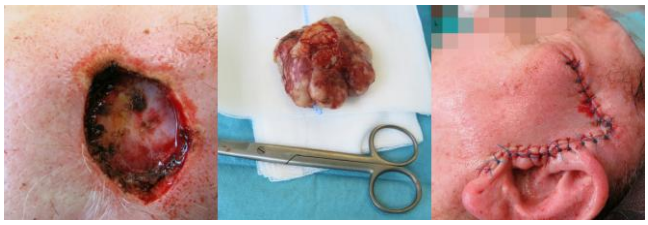

Figure 2: Surgical removal and defect closure: Resulting defect after complete excision (left). Tumor situs, view from the undersurface without the stalk (middle). Defect close by a cheek transposition flap (right)

There was no nuclear pleomorphism and no mitotic figures (Figure 3). The diagnosis of dermal cylindroma was confirmed, the resection status was R0.

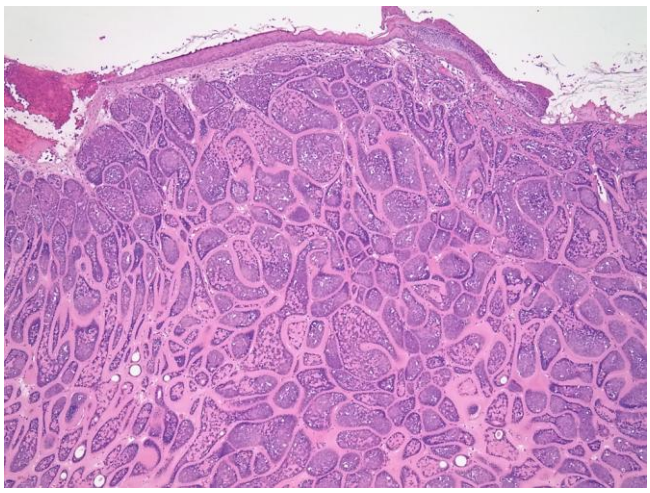

Figure 3: Dermal cylindroma – histopathology with basaloid cell nests in a jigsaw pattern (hematoxylin-eosin x 4)

Discussion

This case is remarkable. Giant tumours such as here are extremely rare. The mushroom-like growth pattern has yet not been described for cylindroma. It was originally described by Alibert for mycosis fungoides [11]. We observed this particular growth pattern with a stalk and exophytic tumour growth in basal cell carcinoma, melanoma, and Merkel cell carcinoma [12]. The present case demonstrates that dermal cylindroma can imitate skin cancer. Complete excision and careful, histopathological investigations are recommended. Fortunately, the true malignant transformation of cylindroma is rare [13] [14].

We had chosen the cheek transposition flap to close the defect after complete surgical removal since this flap offers a robust vascularisation and a perfect match in skin colour and texture. Operation

time is short. In case the tumour had a wider base as the top of the lesion suggested, a meshed graft would be an option for this elderly patient [15].

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