Pinna high grade trichoblastic carcinoma, a report

Elie Yaacoub,[1] Joseph El Borgi,[1] Raymond Challita,[1] Ziad Sleiman,[1] George Ghanime[2]

[1]Plastic and Reconstructive Surgery, Faculty of Medical Sciences, Lebanese University, Beirut; [2]Head, Plastic and Reconstructive Surgery, Lebanese Hospital Geitaoui-UMC, Beirut, Lebanon

Abstract

Trichoblastic carcinoma is a rare malignant hair follicle tumor. It resembles basal cell carcinoma clinically, but different studies emphasize on the importance of differentiating between these two cancers since they have different clinical course which may affect treatment options and follow up. Regardless of its aggressive behavior, no consensus is present for the treatment of trichoblastic carcinoma. We presented the third case of pinna trichoblastic carcinoma, which was surgically excised and followed up for two years post-surgery.

Introduction

The skin is made up of an outer layer the epidermis, an inner layer the dermis, and adnexal structures. Benign and malignant lesions can arise from these structures. Adnexal skin tumors are rare tumors that can arise from hair follicles, sebaceous glands, apocrine, or eccrine sweat glands. Hair follicle tumors can present as benign or malignant lesions. These are mainly differentiated by histological studies.[1] Trichoblastic carcinoma shares a lot of histological similarities with basal cell carcinoma. But different studies emphasize on the importance of differentiating between these two cancers since they have different clinical courses, which may affect treatment options and follow up.[1] No consensus is present for the treatment of these rare tumors.[2] Trichoblastic carcinoma mainly occurs on the scalp region. To the best of our knowledge, only two cases of auricular trichoblastic carcinoma are reported in the literature. So we presented the third case in a 68-year-old male patient who was referred to the Plastic Surgery Department of the Lebanese Hospital Geitaoui-UMC for basal cell carcinoma excision of his right pinna. The patient was then diagnosed as having right ear trichoblastic carcinoma after tumor excision, and required another operation. Surgery was the only treatment received, and the patient was then followed up for 2 years.

Case Report

A 68-year-old male patient with controlled hypertension consulted for a right ear lesion. The lesion started two years ago as a small sub-dermal nodule on the upper third of the helix. This lesion bled easily after minor trauma and then healed slowly. The lesion doubled in size after 1 year and became ulcerated and crusty (Figure 1). The patient did not complain of any pain or pruritus. Physical examination showed a 4×1 cm scaly nodular lesion with ulcerations, which also gave us the impression of basal cell carcinoma. Head and neck lymph node examination showed no palpable lymph nodes. The rest of the examination was normal.

The patient underwent a surgical excision under local assisted anesthesia with macroscopic surgical margins of 0.5 cm. A wedge resection associated with Burrows triangles excision was done followed by approximation of the edges. Two weeks later the pathological result came back as trichoblastic carcinoma with insufficient margins. So patient was re-operated under local assisted anesthesia for larger upper half ear amputation with 1.5 cm margins (Figure 2) followed by retro auricular skin undermining and advancement for closure. The pathological result then came back as the absence of residual neoplastic cells.

Furthermore paraclinical studies were done to rule out loco regional or distant spread. CT scan of the head, neck, and thorax came back normal with no evidence of spread. Abdominopelvic ultrasound was also normal with no evidence of visceral metastases.

Patient was followed up for 2 years with regular 3 months visits. At each visit an extensive clinical exam was done and patient showed no signs of local recurrence or distant metastases (Figure 3). Note that our patient did not receive any adjuvant treatment.

Histopathology

Macroscopically the resected tumor measured 4×1×0.5 cm with central ulcerated lesion. Microscopically, a neoplastic proliferation constituted mainly of two types of cells: a basal type with peripheral palisadic formation and another type of an undifferentiated epidermoid aspect. The latter is made up of cells ulcerating the epidermis and infiltrating the dermis. Intermingled, hyalinized trabecula were noted. Nuclei crowding and mitotic figures were seen. Cellular necrosis was also observed. As a conclusion, the resected tumor is considered as having an aspect compatible with high grade trichoblastic carcinoma (Figure 4). A larger excision was then recommended.

Discussion and Conclusions

Trichoblastic carcinoma is a rare malignant adnexal tumor. No consensus is present for its management, although it is important to differentiate it from other malignant tumors due to its aggressive nature. A lot of controversy is present in the literature and few cases have been studied. Due to this

Correspondence: Raymond Challita, Plastic and Reconstructive Surgery, Faculty of Medicine, Lebanese University, Lebanese University Hospital Geitaoui, Beirut, Lebanon.
Tel.: 096170855498.
E-mail: raychallita@gmail.com

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fact we did a review of literature and compared it with our practice.

With respect to clinical diagnosis and surgical treatment, trichoblastic carcinoma evokes basal cell carcinoma in most cases. It is difficult to distinguish them clinically. In 94% of the cases a diagnosis of Basal cell carcinoma is made.[3] This may explain the high number of reoperations in these patients. Surgeons usually excise according to the Basal cell carcinoma margin's (ideally 0.5 cm). According to a study done by Thomas *et al.* 57% of the patients required revision surgery for inadequate margins.[1] This also occurred in our case where revision surgery was done due to the insufficient margins already taken. Surgery is the mainstay of treatment. Various surgical margins can be seen in the literature. All studies agree on a minimum of 1cm of surgical margin. While some agree that a 1 cm margin is sufficient for all adnexal tumors regardless of severity,[4] others classify these tumors according to their behavior and choose the surgical margin accordingly. Aggressive tumors defined by a diameter greater than 2 cm or histopathological features of neurotropism requires 3 cm surgical margins. Whereas in less aggressive tumors less than 2 cm in diameter and with no histopathological features of neurotropism, a 1 cm margin is sufficient.[5]

Recently, Garcia *et al.* used Mohs micrographic surgery (MMS) to treat a nasal tip trichoblastic carcinoma with good results at 6 months of follow-up. Although more aggressive resection was noted when compared to other tumors operated by MMS, MMS may be an alternative method for the treatment of trichoblastic carcinoma in cosmetically sensitive areas. Thus, larger studies and longer follow-up periods are needed to evaluate the role of MMS in this type of aggressive tumor.[6]

In our case, a 1.5 cm margin in the second operation seemed sufficient with no evidence of local recurrence or distal spread 2 years after surgery. With respect to adjuvant therapy, although radiotherapy has been used in sweat gland tumors where the tumor is unresectable, the patient cannot be re operated, or in the presence of macroscopic residual disease,[7] trichoblastic carcinoma presents low radio sensitivity.[8] However, Laffay *et al.* suggested the use of radiotherapy in selected cases of aggressive trichoblastic carcinoma to improve control.[9]

Chemotherapy can be considered an adjuvant tool for the treatment of patients with trichoblastic carcinoma. Vismodegib is a new chemotherapeutic agent that showed signs of tumor regression especially in locally advanced and metastatic cases. It is a monoclonal antibody that has an inhibitory action on sonic hedgehog pathway. It could represent a new therapeutic tool in the treatment of trichoblastic carcinoma.[8]

Sunitinib is another chemotherapeutic agent that can be used to treat patients with metastatic trichoblastic carcinoma. In one case, partial remission and disease stabilization was observed after 10 months.[10]

With respect to workup and follow up, because of the aggressiveness of the tumor an extensive workup and close follow up are needed. Once the diagnosis is made an extensive clinical workup is needed. A paraclinical workup is to be done in the presence of clinical signs and symptoms.[1] Close follow up every 3 months is recommended for high grade tumors and 6 months for low grade tumors.[1]

So trichoblastic carcinoma is a rare aggressive malignant neoplasm. It mainly occurs in the head and neck region. No consensus is present for its management due to the rarity of cases. Surgical excision with wide margins with a minimum of 1 cm is required. Adjuvant treatment can be provided in poor surgical candidates and in

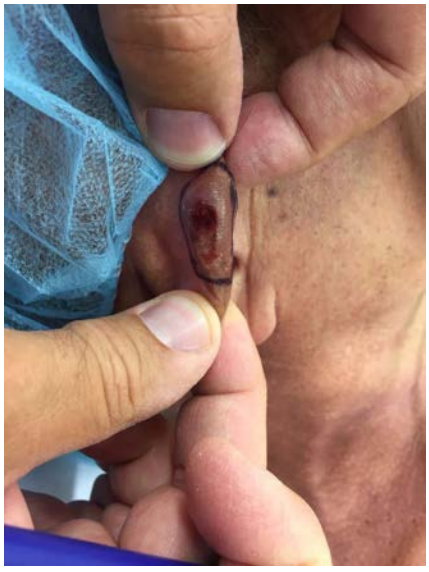

Figure 1. Initial presentation: right ear ulcerated lesion.

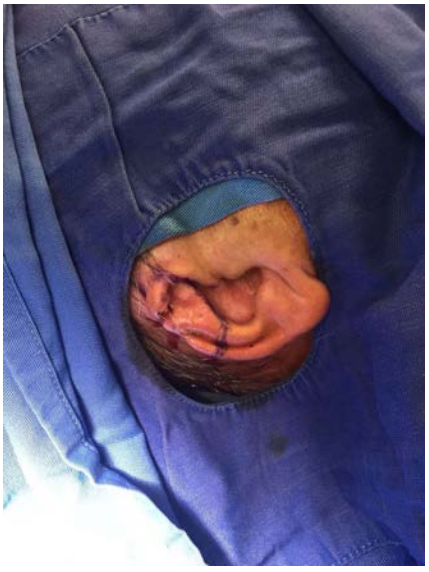

Figure 2. Inadequate margins: markings of the second surgery after histopathology result recommended greater margins of excision.

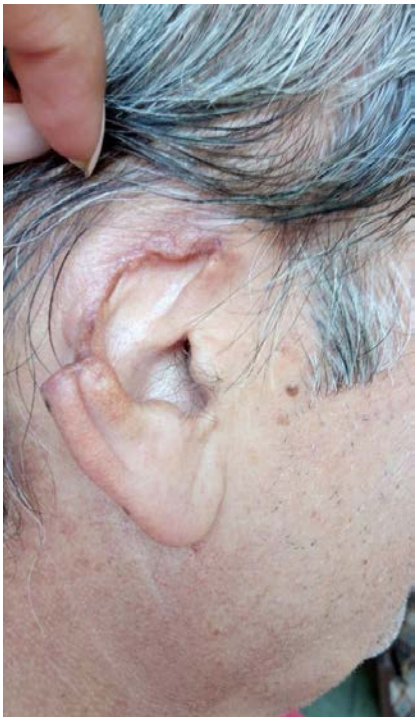

Figure 3. At 6 months follow up: final cosmetic appearance after reconstruction with a retroauricular flap with no local recurrence.

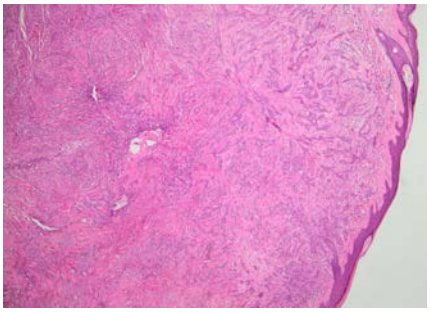

Figure 4. Histopathological result: neoplastic proliferation consisting of two cell types consistent with trichoblastic carcinoma.

patients with distant metastasis. We presented the third case of pinna trichoblastic carcinoma where the only treatment done was wide surgical excision with no evidence of recurrence. Despite all the controversy, wide surgical excision remains one of the major treatments of trichoblastic carcinoma with good results at 2 years of follow-up.

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