

Case Study

Proliferating Trichilemmal Cyst with Lymphadenopathy: A Discussion of Cutaneous Neoplasms Involving the Scalp

Richard T. Klapchar¹, Billie Jean Crigger² and James Paul Dworkin-Valenti^{3*}

¹Department of Otolaryngology, Head & Neck Surgery, Doctors Hospital, USA

²Private Practice, Owensboro, USA

³Department of Otolaryngology, Head & Neck Surgery, Detroit Medical Center, USA

***Corresponding author**

James Paul Dworkin-Valenti, Department of Otolaryngology, Head & Neck Surgery, Detroit Medical Center, 5807 Cedar Ridge Dr, Ann Arbor, MI 48103, USA, Tel: 313-966-9406; 248-444-7396; Email: voicedx@yahoo.com

Submitted: 22 April 2016

Accepted: 29 April 2016

Published: 02 May 2016

Copyright

© 2016 Valenti et al.

OPEN ACCESS

Abstract

This paper introduces a unique case example of a trichilemmal scalp cyst that was initially erroneously diagnosed as a cancerous lesion because of its proliferating cellular morphology. The evaluation and treatment histories of this patient are augmented by a detailed review of the literature on this condition, especially with respect to cases of rare malignant transformation to squamous cell carcinoma. Whereas proliferating pilar or trichilemmal cysts are uncommon tumors that rarely undergo malignant transformation [1]. Squamous cell carcinomas with metastasis to regional head and neck lymph nodes rarely develop from proliferating pilar cysts. However, such degeneration has been reported in the literature [2]. These lesions are fraught with significant morbidity and mortality, and their treatment usually consists of substantial surgical excision. Histopathologic evidence of malignancy often demands adjuvant regimens of radiation therapy (+/-chemotherapy) [3]. The purpose of this case report is to present a patient with an ulcerative, necrotic scalp mass and lymphadenopathy whose initial biopsy demonstrated squamous cell carcinoma. Surgical excision and posterior unilateral posterior neck dissection was performed with final pathology of proliferating trichilemmal cyst. At the close of the case history a detailed review of the literature is presented, particularly with respect to head and neck lesions that have the potential for malignant transformation.

CASE REPORT

A 52 year-old African American male presented initially to an urban emergency room department with the chief complaint of a painful bleeding posterior neck mass and an associated 15 lb weight loss over a 6 month period. His only complaint was scalp pain that inhibited his ability to sleep and wash his hair. On examination, he exhibited a bleeding posterior ulcerative, necrotic scalp mass measuring 10 cm x 9.6 cm x 5.5 cm (Figure 1). Painful lymphadenopathy involving the left posterior and bilateral anterior neck regions was palpated; the largest lymph node measured approximately 3 cm x 2 cm. The patient lived in a shelter for the homeless, but he was healthy otherwise, without a significant past medical history. He denied smoking, alcohol and illicit drug use, head trauma, or exposure to industrial chemicals or radiation. While in the emergency department a biopsy of the lesion was taken for histopathological analysis. Results revealed well-differentiated squamous cell carcinoma. A preoperative

metastatic survey was obtained, including CTs of the chest, abdomen, and pelvis, which revealed a 3.9 cm x 2.7 cm lymph node in the left axilla and a 2.1 cm x 1.5 cm node in the right axilla. There was no evidence of intrathoracic, hepatic, retroperitoneal or pelvic metastatic disease. Wide excision of the scalp neoplasm was performed in conjunction with a left posterior neck dissection under general anesthesia (Figure 2). The surgical defect was repaired with an AlloDerm graft. Deep frozen sections of the galea and pericranium were submitted for analyses. These tissue specimens were negative for carcinoma; all margins were also negative. Because our index of suspicion for carcinoma remained high, notwithstanding these path results, an additional 1-cm cuff of scalp tissue encompassing the lesion was dissected and sent for permanent histopathological analysis (Figure 3). Meticulous hemostasis was achieved, fibrin glue was placed onto the large surgical defect, and a scaffold AlloDerm graft was applied to facilitate healing via secondary intention. The graft was trimmed peripherally, allowed to drape over the tissue edges, and sutured

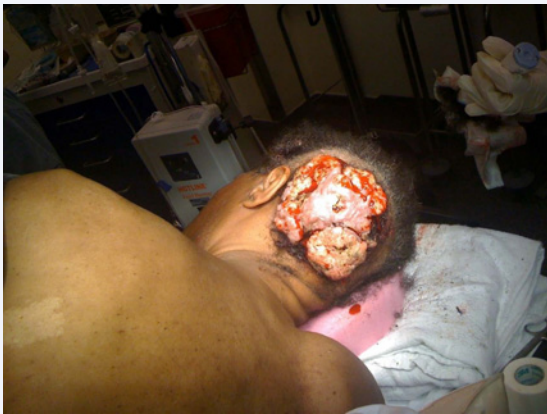


Figure 1 Intra-operative photograph of patient showing large bleeding posterior ulcerative, necrotic scalp mass measuring 10 cm x 9.6 cm x 5.5 cm.



Figure 2 Intra-operative photograph of surgical defect.



Figure 3 Intra-operative photograph of surgical specimen.

onto the recipient site with 2-0 nylon and 2-0 Vicryl. Xeroform gauze was placed over the graft and held in place via retention sutures at the 2, 4, 6, 8, 10, and 12 o'clock positions with 0-0 prolene. A sterile fluff dressing and ABD fluff pad was placed on top of the AlloDerm graft and secured. Final histopathology evaluation reported the occipital scalp neoplasm to be a

proliferating pilar (trichilemmal) cyst with benign fibroadipose tissue. This neoplasm did not extend into the normal cuff scalp tissue; negative margins were obtained without any evidence of squamous cell carcinoma, regardless of the earlier reported biopsy results to the contrary. All 13 lymph nodes dissected were also negative for malignancy. The patient was kept in the hospital for immediate postoperative care. Social services were contacted to help with postoperative care. The patient was followed in our office for serial cleanings and AlloDerm graft debridement. Four months postoperatively, the surgical site re-epithelialization was nearly complete. He was followed serially to monitor maturation of the surgical site. Within one year, he was able to obtain medical insurance and discussions ensued of cosmetic reconstruction of the non-hair bearing surgical area. Repeat CT scans of the neck and chest showed resolution of the axillary lymphadenopathy. Surgical options were discussed with the patient with respect to the non-hair bearing scalp defect. Use of soft tissue expanders were considered to prepare the skin for a pedicle flap rotation to create a hair bearing area.

DISCUSSION

There are many different pathologic neoplasms that need to be considered in the differential diagnostic work-up of a suspected trichilemmal cyst. These include: 1) epidermoid cyst, 2) dermoid cyst, 3) squamous cell carcinoma, (4) steatocystoma, 5) pilomatrixoma, 6) nodular hidradenoma, 7) sweat gland tumor, 8) dermato-fibrosarcoma protuberans, 9) basal cell carcinoma, 10) angiosarcoma, 11) granular cell tumor, 12) giant cell tumor, 13) cylindroma, and 14) cutaneous manifestations of B-cell lymphoma, meningioma or metastasis [6-8]. A discussion of some of the most common pathologies above is presented below.

Epidermoid cyst

Like trichilemmal cysts, epidermoid cysts are true varieties. They can be differentiated from the trichilemmal type by evaluation of the cyst lining [1]. The epidermis cyst lining produces keratinized cellular debris that fills the cyst cavity. It is believed that this type of cyst arises from the follicular infundibulum of hair shafts, and that it may develop insidiously or after trauma to the affected area. If presentation occurs prior to puberty, the cyst may be an indication of Gardner's syndrome. As with trichilemmal cysts, malignant transformation is rare. If the cyst ruptures and becomes infected, pain and scarring can result and cause a cosmetic deformity. Treatment is most effective with complete surgical excision. Some lesions respond to intracystic triamcinolone injections.

Dermoid Cyst

Dermoid cysts are often congenital. Whereas they can occur anywhere on the body, they have a predilection for forming on the face along the lateral eyebrow, orbit and nose. If noted along the nasal root, care must be taken to differentiate it from a nasal glioma. Dermoid cysts arise from inclusion of embryonic epidermis in the embryonal fusion plates [1]. The lining of the dermoid cyst consist of epithelium resembling epidermis. These growths attach to adnexal structures, such as hair and glands (sebaceous, eccrine and apocrine), which aid in differentiation from epidermoid and trichilemmal cysts.

Squamous cell carcinoma

This is the second most common skin cancer. Risk factors are both environmental and genetic, including but not limited to fair skin complexion, chronic sun exposure, smoking, history of skin cancer and HPV infections. If caught early, many squamous cell carcinoma lesions can be cured; a poorer prognosis is associated with perineural invasion and tumor size greater than 2 cm in diameter. Initially, the lesion presents as a scaling, hyperkeratotic plaque that slowly changes into an ulcerative base with a rolling margin that bleeds easily [1]. Treatment typically involves excision with 1-2 cm margins for advanced disease. Alternative approaches to management may include a combination of Mohs surgery, radiation therapy, and neck dissection with positive lymph nodes. Lifestyle preventative measures are encouraged.

Steatocystoma

These are rare intradermal cysts that occur after puberty in an autosomal dominant inheritance pattern. They can frequently be found on the scalp, anterior chest, and face. An exudative yellowish oily fluid with hair is a common feature. Corrugated walls of epithelial cells, with multiple sebaceous gland lobules and keratinized epithelial lining resembling the outer root sheath, are visualized histologically. Infection and scarring are the major complications with this cyst. Dermabrasion, excision, laser (CO₂), and retinoids represent the most efficacious treatments.

Nodular hidradenoma

Benign adnexal tumors resemble epidermoid cysts [7]. They arise from eccrine and apocrine glands. These neoplasms commonly form on the head and neck, giving rise to the appearance of a superficial ulcerative lesion with serous drainage. Because nodular hidradenomas can behave aggressively, they are best treated via surgical excision. Malignant transformation is possible and evident on histopathology; high mitotic activity, angiolymphatic and adjacent tissue invasion are often evident.

Cylindroma

This type of tumor is believed to arise from follicular epithelium, and presents as pink/red dermal nodules of the head, neck and scalp. Multiple tumors on the scalp have been termed, "turban tumors" [1]. Histologically, the epithelial cells are surrounded by hyaline. Cylindromas need to be differentiated from basal cell carcinomas. These lesions have the potential to transform into malignant variants.

Pilar (Trichilemmal) cysts

These growths are true cysts. As previously noted, they are difficult to distinguish from epidermoid cysts because they possess a lining that is analogous to the outer root sheath of a hair follicle. Most trichilemmal cysts occur on the scalp, usually in clusters [8]. Complications occur if the cyst ruptures and becomes infected, which can result in pain and scarring. Trichilemmal cysts can progress to proliferating pilar tumor. But they rarely express malignant transformation into squamous cell carcinoma [1,2,8]. Treatment is complete surgical excision. Pilar cysts can be further classified into simple or proliferating categories with the later being further divided into benign or malignant lesions, with the malignant subset differentiated into low or high grade categories

[2,4]. It is often difficult to distinguish a proliferating pilar tumor from a malignant neoplasm such as squamous cell carcinoma, due to the histological atypia and pleomorphism, as was true in the current patient example. Discrete criteria are lacking to facilitate differentiation between proliferating pilar cysts and squamous cell carcinomas. Consequently, the suspicious lesion must be biopsied and evaluated histologically to determine the underlying pathology and the cellular complexity of the specimen. Occasionally, trichilemmal cysts are ridden with coexisting and overlapping squamous cell malignant infiltrates. When such complicated mixed tumors arise they challenge pathologic identity as well as decisions regarding treatment [5]. When malignant overlay is confirmed tumor reduction may be effectively achieved using chemotherapeutic agents, such as cisplatin and fluorouracil. Radiotherapy and planned neck dissection for a patient with a recurring proliferating pilar cyst with nodal extension have been reported to be successful treatments at seven year disease-free follow-up examinations [8]. For our patient the initial path report demonstrated well-differentiated squamous cell carcinoma. The initial biopsy was derived from large pieces of tumor that were removed with forceps and scalpel. It is not inconceivable that necrotic tissue was included with the specimen for analysis. The necrosed fragments may have produced false positive pathology conclusions regarding the presence of malignancy. We decided to perform a wide surgical excision intervention approach to include large tissue components of normal squamous epithelium and hyaline basement membrane. Final pathology ruled out malignant transformation of this patient's cystic neoplasm. In general, proliferating pilar cysts are benign growths that are curable with simple excision. When such cysts possess low-grade malignant proliferating cellular characteristics, they have a small risk of local recurrence and should be treated with local resection (+/-adjuvant radiation and chemotherapy). High grade malignant proliferating pilar cysts also tend to recur locally, and may metastasize to loco-regional nodes. More aggressive surgical and non-surgical treatments are usually required for control [4]. A neoadjuvant approach to management of such mixed tumors has been reported, whereby chemotherapy (cisplatin, fluorouracil) was administered to reduce the tumor size for successful excision at a later date and for lymph node control [5]. In our patient, there was no role for such a combined treatment plan. Surgery alone was successful in controlling the large cystic mass on the posterior scalp, and posterior neck node dissection arrested the associated, reactive lymphadenopathy. The patient remains disease free at 24 months postop.

Finally, there may be a place for sentinel node biopsy or selective neck dissection in patients that present with trichilemmal cysts without palpable lymphadenopathy. Further research is required to test the validity of this hypothesis relative to long term control of this disease and its propensity toward malignant deterioration.

REFERENCES

1. Cummings. Otolaryngology-Head and Neck Surgery. Fifth Edition. 2010.
2. Eskander A, Ghazarian D, Bray P, Dawson L and Goldstein D. Squamous Cell Carcinoma Arising in a Proliferating Pilar (Trichilemmal) Cyst

- with Nodal and Distant Metastases. *Journal of Otolaryngology-Head and Neck Surgery*. 2010; 39: 63-67.
- Sau P, Graham JH, Helwig EB. Proliferating epithelial cysts. Clinicopathological analysis of 96 cases. *J Cutan Pathol*. 1995; 22: 394-406.
 - Garg PK, Dangi A, Khurana N, Hadke NS. Malignant proliferating trichilemmal cyst: a case report with review of literature. *Malays J Pathol*. 2009; 31: 71-76.
 - Ye J, Nappi O, Swanson PE, Patterson JW, Wick MR. Proliferating pilar tumors: a clinicopathologic study of 76 cases with a proposal for definition of benign and malignant variants. *Am J Clin Pathol*. 2004; 122: 566-574.
 - Weiss J, Heine M, Grimm M, Jung EG. Malignant proliferating trichilemmal cyst. *J Am Acad Dermatol*. 1995; 32: 870-873.
 - Satyaprakash AK, Sheehan DJ, Sangüeza OP. Proliferating trichilemmal tumors: a review of the literature. *Dermatol Surg*. 2007; 33: 1102-1108.
 - Garcia-Zuazaga J, Ke MS, Willen M. Epidermoid cyst mimicry: report of seven cases and review of the literature. *J Clin Aesthet Dermatol*. 2009; 2: 28-33.

Cite this article

Klapchar RT, Crigger BJ, Dworkin-Valenti JP (2016) Proliferating Trichilemmal Cyst with Lymphadenopathy: A Discussion of Cutaneous Neoplasms Involving the Scalp. *J Ear Nose Throat Disord* 1(1): 1002.