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## Case Presentation

### Uncorrectable Ptosis: Primary Cutaneous Signet-ring Cell Carcinoma

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## Abstract

Primary cutaneous signet-ring cell carcinoma (PCSRCC) is a rare but aggressive tumor. Our case highlights a 60-year-old man who presented with eyelid ptosis, for which he underwent multiple surgical procedures over a 3-year period prior to referral to our clinic. These procedures were complicated by scarring, delayed healing, and poor cosmetic outcome. In addition, the patient was noted to develop progressive enophthalmos. These concerning signs led to a CT scan and subsequent eyelid biopsy, which revealed a diagnosis of PCSRCC. Further management has involved an MRI and orbitotomy with biopsy revealing widespread extension of the carcinoma. Exenteration was performed to reduce the likelihood of metastasis.

There are few documented case reports of PCSRCC of the eyelid in the literature. Of the 33 published cases of PCSRCC, 27 cases involve the eyelids and the other 6 cases involve the axilla. The unique clinical features of this case will be discussed, in particular the presentation as ptosis, an otherwise commonplace complaint in the oculoplastics clinic. The surgical course and histopathologic findings will be presented. The literature regarding PCSRCC will be reviewed including demographics, management, and prognosis. Although rare, PCSRCC follows an aggressive course with characteristically delayed diagnosis. Early identification and treatment likely offer a better prognosis. Thus, description of the clinical presentation of this rare tumor may aid in recognition and earlier treatment.

**Keywords:** Signet-ring cell carcinoma, ptosis, primary cutaneous signet-ring cell, carcinoma

## Case synopsis

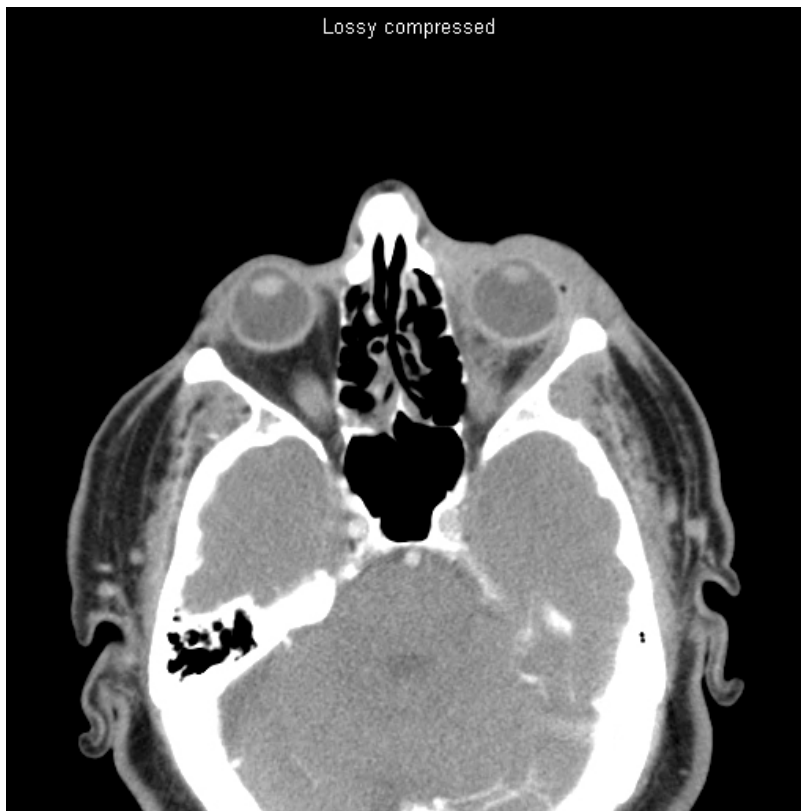
A 60-year-old man initially presented to an oculoplastic surgeon for repair of his unilateral ptosis. He recalled that as a child, he had a cyst over his left eye removed, which was thought to have possibly contributed to his droopy eyelid. He underwent uncomplicated ptosis repair but it was noted in the operative report that fibrotic tissue was encountered. The initial surgery resulted in a peaked eyelid and he desired revision for improved cosmetic appearance. The next surgeon reported an uncomplicated repair, but the surgery was complicated by poor wound healing and chronic discharge.

Three-and-a-half years from his initial surgery the patient presented to our clinic for a second opinion for ptosis revision and chronic ocular discharge (Figure 1). Mild enophthalmos was noted on exam, which is not consistent with involutional ptosis. Thus, imaging was obtained, revealing a tissue-enhancing orbital mass (Figure 2). Subsequent biopsy revealed the classic pathologic findings consistent with signet-ring cell carcinoma. We then performed a map biopsy in order to determine the extent of involvement. The margins were positive in all samples and included "skip areas" throughout the samples examined. A PET scan was negative for alternative primary tumor sites and negative for metastatic spread.

Our patient then underwent complete surgical excision that required orbital exenteration. Frozen sections at the time of surgery all had negative margins except for a small area along the inferior eyelid. The specimen showed a single focus of perineural invasion. Following exenteration, the patient underwent adjunctive radiation therapy with curative intent and has been free of systemic disease for 22 months since the exenteration.



**Figure 1.** External photo with map biopsy markings showing ptosis, peaked upper eyelid and non-healing incision from previous surgery.



**Figure 2.** CT axial cut showing significant thickening of left orbital tissue.

## Discussion

### *Primary Cutaneous Signet-ring Cell Carcinoma*

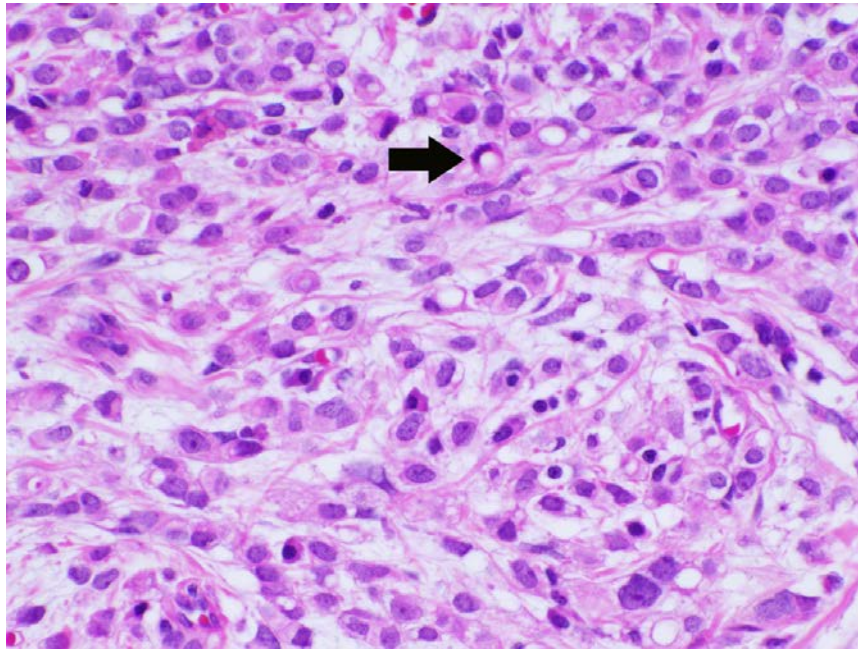
Primary cutaneous signet-ring cell carcinoma (PCSRCC) is a rare but very aggressive tumor [5]. The most common site is the eyelid or axilla and usually involves only the dermis and subcutaneous tissue, sparing the epidermis. The cell of origin remains controversial but usually includes either the eccrine or apocrine cell lines [1,2]. Because PCSRCC is rare, a detailed work-up is warranted to exclude extra-cutaneous metastatic signet-ring cell adenocarcinoma arising from GI, GU, or breast, which is more common than PCSRCC.

Prior to this case, there have been 33 published cases of PCSRCC with the majority, 27, originating in the eyelid and the others from the axilla. PCSRCC has a male predominance and often is initially misdiagnosed as blepharconjunctivitis, chalazion, or other inflammatory disorders causing a delay in diagnosis [4,5].

### ***Pathology***

The histological features of PCSRCC include round cells with histiocytoid and occasional signet-ring features (Figure 3). The cells can diffusely percolate through the dermis as solid collections, in single-file formations, or as individual cells, without involvement of the epidermis.

Mucicarmine stains highlighted the signet-ring cells. By immunohistochemistry the tumor cells were immunopositive for cytokeratin 7, CAM5.2, carcinoembryonic antigen, e-cadherin, p63, and gross cystic disease fluid protein-15 (BRST2). The following immunostains were negative: CDX2, D240, CD68, S-100, BerEP4, mammoglobin, TTF-1, CK20, HMB45/Mart1, and PSA.



**Figure 3.** Histopathology of biopsy with characteristic findings of Signet-Ring Cell Carcinoma (arrow).

### ***Treatment***

The location of PCSRCC determines the treatment modality. PCSRCC arising from the axilla can be locally excised. PCSRCC arising from the eyelid requires excision with wide margins and orbital exenteration is often indicated. For cases in which complete excision is not possible, radiotherapy following surgery has been shown to be useful. Adjunctive chemotherapeutics are still being studied and have shown varied success in advanced cases [8]. PCSRCC invades adjacent tissue and 13 cases have documented metastatic spread to either regional lymph nodes or internal visceral; 2 of those patients died from complications of tumor metastasis [4,5].

### ***Types of Ptosis***

There are many different causes of ptosis. Ptosis can be classified as either congenital, which includes myogenic or neurogenic, or acquired, which includes involuntional (most commonly resulting from dehiscence of the levator aponeurosis), traumatic, mechanical, neurogenic, myogenic, or neuromuscular. The type of ptosis can often be elucidated by a thorough history and examination. Discerning the etiology of the ptosis is critical in deciding the type of repair [3,6].

### ***Ptosis Repair***

Ptosis can be managed in a variety of ways including a more conservative approach and surgical repair. Surgical approach depends on etiology but could involve procedures involving Muller's muscle, levator aponeurosis, or brow/frontalis procedures.

The success rate of ptosis repair varies greatly, with a reported revision rate ranging from 0.6% to 33% depending on the type of repair. There were very few complications in the studies that have been reported [3,6]. Since the success rate of ptosis repair is relatively high, any patient requiring multiple revisions should be re-evaluated to ensure proper management. If any suspicious tissue is encountered, imaging or biopsy is strongly recommended.

## Conclusion

Early diagnosis and initiation of treatment is critical for any patient with a malignant tumor. Therefore, unexpected surgical outcomes or unusual clinical associations should cause the astute clinician to critically analyze the cause of failure, especially in ptosis repair, which typically carries a high surgical success rate. Tissue biopsy or imaging should be performed without delay where indicated. Although the patient in the case presented above does not have any evidence of local spread or metastatic disease at the time of this publication, the patient had indications for biopsy early in his treatment course. Whereas the cutaneous presentation of this patient was subtle, any suspicious findings should prompt a biopsy. Earlier diagnosis and initiation of treatment could have improved the prognosis of our patient.

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