Abstract

Introduction
Pilomatrix carcinoma is a rare cutaneous malignancy of hair matrix cells. They exhibit an aggressive and infiltrative growth pattern and, despite treatment to the local disease, there is a high propensity for recurrence. The literature reports a poor prognosis associated with regional spread occurring with distant metastasis. The patient we report is alive at over two years post-operatively. With only one other case of a head and neck pilomatrix carcinoma ever reported in Canada, our report and review of the literature contributes to what little is known currently about the treatment of this malignancy.

Case report
In previously reported cases, regional lymph node metastasis is always associated with distant metastatic disease on presentation, thus rendering the treatment palliative in nature. We present the unique case of a 63-year-old man with a pilomatrix carcinoma of the left parotid region. He had bilateral cervical lymph node spread but no detectable distant metastasis. The patient underwent an excision of the lesion with a radical parotidectomy, temporal bone resection, bilateral neck dissections, and post-operative radiation.

Conclusion
The treatment of pilomatrix carcinomas involve extensive surgical management with wide margins. Post-operative radiation in patients without distant metastasis may decrease the risk of local recurrence.

Introduction
Pilomatrix carcinoma is the rare malignant variant of pilomatrixoma, a cutaneous tumour of hair matrix cells – also known as the calcifying epithelioma of Malherbe1. A literature review using the Pubmed and Embase databases yielded approximately 90 reported cases of pilomatrix carcinomas. Fifty-one of these occurred in the head and neck region.

Due to the paucity of head and neck pilomatrix carcinoma cases, little is known about the optimal management, particularly in the case of regional spread without distant metastasis. In previously reported cases, regional lymph node metastasis is always associated with distant metastatic disease on presentation, precluding curative treatment. We present the unique case of a 63-year-old man with a pilomatrix carcinoma of the left parotid region. He had bilateral cervical lymph node spread but no detectable distant metastasis.

The literature reports a poor prognosis associated with regional and concurrent distant metastasis. The patient we report has done well and is alive at over two years post-operatively. There is only one other case of a head and neck pilomatrix carcinoma ever reported in Canada.

Case report
A 63-year-old man presented to a community Otolaryngologist with a post-auricular mass of six months duration. At the time of presentation, it was ulcerated and draining. Aside from a recent weight loss of 10 pounds, the patient was essentially asymptomatic in terms of pain and function. A CT scan showed a 3 cm lesion which appeared to be arising laterally in the left parotid gland, with enlarged lymph nodes inferior and deep to the gland. Biopsy revealed an initial diagnosis of squamous cell carcinoma.

The patient was subsequently evaluated at the Tom Baker Cancer Centre by the multidisciplinary head and neck team and further investigations were arranged. On exam, there was a large, fixed mass in the left posterior neck, measuring approximately 6 cm in diameter (Figure 1). Pathology review of the biopsy at a tertiary centre resulted in a revised diagnosis of pilomatrix carcinoma. Additional imaging was undertaken to evaluate the extent of disease. Both the CT and MRI scans showed a large, 5 cm x 3 cm mass in the left suprathyroid neck region with involvement of the overlying skin and bilateral cervical lymphadenopathy.

The tumour extended to the major vessels of the neck, abutting branches of the left external carotid artery as well as the internal jugular vein. There was also gross invasion of the sternocleidomastoid muscle at the mastoid origin of the parotid gland. A CT/PET scan showed bilateral cervical lymph node disease but no evidence of distant metastasis.

The patient underwent an excision of the mass with 1.5-2 cm cutaneous margins, along with a left extended neck dissection, left radical parotidectomy, left lateral temporal bone resection and right modified neck dissection (levels I-IV). He was treated with adjuvant radiotherapy, receiving 4620 cGy of the 6600 cGy that was prescribed because of patient non-compliance. The patient did not attend scheduled regular post-treatment follow-up. Eighteen months later, he presented with an isolated 1.6 cm lymph node in his right supravacular fossa. CT imaging of the neck, chest, and...
abdomen failed to demonstrate other local, regional, or distant disease. Fine needle aspiration of the mass suggested squamous cell carcinoma. Wide local excision of the mass was recommended but the patient declined.

Pathological Findings
On gross examination, the tumour measured 5.8 x 4.2 cm and was found to invade the left parotid gland and surrounding skeletal muscle. There was no extension into the temporal bone. The left neck dissection yielded five positive lymph nodes while there were thirteen nodes positive for metastasis on the right side.

The tumour showed a nodular basosquamous proliferation with areas of necrosis (Figure 2). The advancing tumoural front showed individual cells percolating through a desmoplastic stroma. Tumoural nests composed of pleomorphic basaloid cells were sharply demarcated from central keratinizing areas. Numerous cells were captured in mitoses. Both lymphovascular and perineural invasion were present.

Discussion
Pilomatrix carcinomas (PC) are rare malignant tumours of the hair matrix. There have only been approximately 90 reported cases of pilomatrix carcinomas, half of which occurred in the head and neck region. This family of matrical tumours also includes pilomatrixomas and proliferating pilomatrixomas. It is worthwhile to distinguish these three entities both clinically and histologically (Table 1).

Pilomatrixomas have a bimodal distribution for age of onset with up to 60% diagnosed before the age of 20,2,3,5,6 and a later peak in the 60s-70s.5,6,7 There is a female predominance with a male: female ratio of 2:3.2,3,5,6 Pilomatrixomas are typically smaller in size (<2-3 cm), slow-growing, and are regular in appearance.8,9,10,11 Recurrences are unlikely (3%) with simple excision.12 Histologically, pilomatrixomas are characterized by collections of uniform basaloid cells with shadow cells and the absence of local invasion, central necrosis, and atypical mitosis.

Proliferating pilomatrixomas also have a female predominance but have been mostly reported in older patients from 60-80 years old.7,11 They can grow to upwards of 5.5 cm but are similar to ordinary pilomatrixomas, typically symmetrical in appearance.7,11 Because proliferating pilomatrixomas are infiltrative in their growth pattern, there is a greater likelihood for recurrence after a simple excision (14%).11 Histologically, there is greater atypia and a higher mitotic rate than is seen with a pilomatrixoma.2 Tissue invasion and central necrosis are present. A pilomatrix carcinoma has similar characteristics to a proliferating pilomatrixoma but with significantly greater pleomorphism as well as the presence of perineural and vascular invasion.

Figure 1: Ulcerated mass measuring 5.8 x 4.2 cm in the left parotid region.

Figure 2: Tumoural nests are composed of irregularly shaped basaloid cell aggregates. Centrally, keratotic material, focal necrosis and shadow cells are found.
invasion. Pilomatrix carcinomas occur mostly in middle-aged patients, with an average age of 45 to 60 years old. There is a strong male predominance with a male:female ratio of 3:5 to 3:1. The head and neck region is most often affected and the carcinoma may arise de novo or as a result of malignant transformation of a pre-existing pilomatrixoma.

While it is a low-grade tumour, PCs exhibit an infiltrative growth pattern and are locally aggressive. With inadequate surgical management, there is a great propensity for recurrence, with recurrence rates from 46-60% to 60-70% to 50%. Therefore, the consensus in literature regarding surgical treatment is wide excision with a minimum margin of 5 mm.

The potentially aggressive nature of PCs has been demonstrated in cases of extensive local invasion into surrounding soft tissues and bony structures, at either initial presentation or at the time of recurrence. Treatment involves wide excision of the tumour along with affected bone and soft tissues. Outcomes are good with no evidence of disease at the time of follow-up in the reported cases.

Distant metastasis has also been reported. The most common sites of metastasis are the lungs, spine, and abdominal viscera. Chemotherapy and radiation was ineffective in halting disease progression, suggesting that such treatments are largely futile in cases of distant metastasis. However, there may be a role for adjuvant radiotherapy after surgical excision (both wide and simple), which has been used effectively for local disease control.

Conclusion
In concordance with previous reports, our case demonstrates the capacity for PCs to invade locally and spread regionally. In terms of treatment, our case supports the approach of aggressive and extensive surgical management of the tumour with wide margins. Due to the few number of PC cases, there is little experience in using radiation and chemotherapy as part of the treatment for this malignancy.

Reports to date have shown very little effect in using these therapies in patients with distant metastasis. However, there is some evidence to suggest that radiation may be useful in controlling local disease. Our case can be added to the small body of literature supporting the use of aggressive surgery and adjuvant radiation in patients without distant disease to prevent local recurrence.

Consent
Written informed consent was obtained from the patient for publication of this Case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Abbreviations list
PC – pilomatrix carcinoma

Authors’ Contribution
CCL participated in the literature search and drafting, revising, and approving the final manuscript. MH

Table 1: Clinical and histological characteristics of pilomatrixoma, proliferating pilomatrixoma, and pilomatrix carcinoma.

<table>
<thead>
<tr>
<th></th>
<th>Pilomatrixoma</th>
<th>Proliferating Pilomatrixoma</th>
<th>Pilomatrix carcinoma</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age</td>
<td>Bimodal distribution - &lt;20 yo 5% - 60-70 yo 6-7%</td>
<td>Mostly elderly – &gt;60-70% 11</td>
<td>Middle aged – 45-60 yo 11,13,18</td>
</tr>
<tr>
<td>Gender</td>
<td>M:F ratio 2:3 2-3,5,6</td>
<td>M &lt; F 11</td>
<td>M:F ratio 3:4:1 3-13,14,16,19</td>
</tr>
<tr>
<td>Clinical characteristics</td>
<td>- Single, slow growing, and asymptomatic nodule - Cystic, symmetric 11 - Recurrence rate – 3% 12</td>
<td>- Solid, symmetric 11 - Locally invasive - No metastatic potential - Recurrence rate – 14% 11</td>
<td>- Irregular 11 - Growth may be slow or rapid 3 - Locally invasive 3,10-11,25-26,30 - Can metastasize to lungs, bone, abdominal viscera 4 - Recurrence rate – 46-60% 11,13,20</td>
</tr>
<tr>
<td>Histology</td>
<td>- Collections of basaloid cells, usually at the periphery 11 - Cells are uniform, few mitotic figures 23 - Shadow cells present 2 - No central necrosis 2 - No lymphovascular or perineural invasion 2</td>
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participated in revising and approving the final manuscript.

TWM participated in revising and approving the final manuscript. KG participated in drafting the pathology section of the manuscript, and revising and approving the final manuscript. SC participated in revising and approving the final manuscript.

References

Table 2: Pilomatrix carcinoma cases.

<table>
<thead>
<tr>
<th>Location of tumor</th>
<th>Number of Cases</th>
<th>Treatment (number of cases)</th>
<th>Follow up (number of cases)</th>
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<tbody>
<tr>
<td>Posterior neck</td>
<td>11</td>
<td>Wide excision (5)</td>
<td>NED (4), DOD (1)</td>
</tr>
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<td></td>
<td></td>
<td>Simple excision (4)</td>
<td>NED (2), DH (1), LFU (1)</td>
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<td></td>
<td>Excision, MNS (2)</td>
<td>NED (1), AWD (1)</td>
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<tr>
<td>Lateral/ anterior neck</td>
<td>3</td>
<td>Wide excision (2)</td>
<td>NED (1), LFU (1)</td>
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<td></td>
<td></td>
<td>Excision, MNS (1)</td>
<td>LFU (1)</td>
</tr>
<tr>
<td>Scalp</td>
<td>10</td>
<td>Wide excision (7)</td>
<td>NED (4), LFU (3)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Simple excision (3)</td>
<td>NED (1), DOD (1), LFU (1)</td>
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<tr>
<td>Eye region</td>
<td>6</td>
<td>Wide excision (3)</td>
<td>NED (1), LFU (2)</td>
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<td>Simple excision (3)</td>
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<tr>
<td>Cheek</td>
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<td>LFU (1)</td>
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<tr>
<td>Pre-auricular</td>
<td>5</td>
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<td>Post-auricular</td>
<td>NED (1)</td>
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<td></td>
<td></td>
<td>Ear</td>
<td>NED (1)</td>
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<td></td>
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<td>Nose, lips, forehead</td>
<td>NED (1), LFU (1)</td>
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<td></td>
<td></td>
<td>Scapula, supraclavicular</td>
<td>NED (1)</td>
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<td></td>
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<td>MNS - margin not specified</td>
<td>NED (1), LFU (1)</td>
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<tr>
<td></td>
<td></td>
<td>DOD - died of disease</td>
<td>NED (1), AWD (1)</td>
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<td></td>
<td></td>
<td>NED - no evidence of disease</td>
<td>NED (1), LFU (1)</td>
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<td></td>
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<td>DII - died of recurrent illness</td>
<td>NED (1), AWD (1)</td>
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<td></td>
<td>LFU - lost to follow-up</td>
<td>NED (1), AWD (1)</td>
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</tbody>
</table>

MNS - margin not specified, DOD - died of disease, AWD - alive with disease, NED - no evidence of disease, DII - died of recurrent illness, LFU - lost to follow-up

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Case report


Competing interests: None declared.
Conflict of interests: None declared.
All authors contributed to conception and design, manuscript preparation, read and approved the final manuscript. All authors abide by the Association for Medical Ethics (AME) ethical rules of disclosure.