Case Report

Beyond the skin: A case report of Merkel cell carcinoma involving the external auditory canal and parotid gland

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ABSTRACT

Merkel cell carcinoma (MCC) is a rare neuroendocrine tumour most often arising from sun-exposed skin in elderly Caucasian patients, with high rates of local invasion, distant metastasis, and recurrence. A rare case of infiltrative MCC is described in a 73-year-old Caucasian female with two enlarging lesions in the left external auditory canal (EAC) and parotid gland. Due to this rare clinical presentation, MCC was not considered in the initial differential diagnosis. However, histopathological analysis demonstrated a high-grade neuroendocrine carcinoma with immunohistochemical findings supporting MCC, despite an atypical staining profile. Imaging revealed rapid progression with regional nodal metastases. Despite MCC's grave prognosis, the patient achieved a complete metabolic response to chemotherapy and is currently undergoing adjuvant radiotherapy. This case highlights an uncommon presentation with a wide differential diagnosis, emphasizing the critical role of radiologic and pathologic evaluation alongside a high index of suspicion for MCC in atypical head and neck lesions.

Keywords: Merkel cell carcinoma; Neuroendocrine tumour; Chemotherapy; Radiotherapy; Radiology; Oncology.

Merkel cell carcinoma (MCC) is a rare, aggressive neuroendocrine tumour commonly arising in sun-exposed areas of the skin.[1] Its global incidence is approximately 0.1-1.6 cases per 100,000 annually and is highest in Australia; however, incidence increased by 95% in the United States between 2000-2013.[2] Although the cause of this increase is unclear, proposed explanations, including population aging, increased UV exposure, immunosuppressive medication use, and improved immunohistochemical detection, mirror established risk factors such as sun exposure, Caucasian ethnicity, immunosuppression, and advanced age.[3,11] MCC most commonly occurs on sun-damaged skin of the head and neck in elderly Caucasian patients.[2] Notably, a novel polyomavirus of the Polyomaviridae family was found to be associated with the development of MCC: Merkel cell polyomavirus (MCPyV).[3] A Pan-Canadian Merkel Cell Cancer Collaborative cohort study found five-year overall survival (OS) rates for head and neck MCC ranging from 49.8% for stage 1 to 18.5% for stage 4.[4]

This report describes an extremely rare case of infiltrative MCC involving the left parotid gland and external auditory canal (EAC) in a 73-year-old Caucasian female.

CASE PRESENTATION

A 73-year-old female with a history of hypertension, atrial fibrillation, pacemaker placement, dyslipidemia, hypothyroidism, and recurrent transient ischemic attacks presented to her family physician with a 2-month history of a progressively enlarging palpable mass in the left EAC and a one-month history of firmness in the left preauricular area (anterior to the auricle). She was subsequently referred to otolaryngology. Physical examination revealed a firm 1-cm mass involving the left conchal bowl (central depression of the auricle) and a palpable small firm nodule involving the left parotid gland. Given the rarity of MCC involving the EAC and parotid region, the initial differential diagnosis included cystic basal cell carcinoma (BCC) or unusual-appearing squamous cell carcinoma (SCC). A 4-mm punch biopsy was performed on the left conchal bowl mass, and a fine-needle aspiration was performed on the left parotid gland mass; both specimens were sent urgently.

Histopathologically, both lesions demonstrated features consistent with a high-grade neuroendocrine carcinoma. Immunohistochemical staining revealed strong paranuclear dot-like positivity for pankeratin and neurofilament, as well as positivity for synaptophysin, supporting neuroendocrine differentiation. The Ki-67 proliferative index was elevated, suggesting aggressive tumour behaviour. Patchy weak chromogranin positivity was observed in the parotid sample. Both specimens were negative for cytokeratin 20 (CK20), thyroid transcription factor-1 (TTF-1), cytokeratin 7 (CK7), chromogranin (in the conchal bowl mass), S100, human melanoma black-45 (HMB-45), cluster of differentiation 99 (CD99), and leukocyte common antigen (LCA). Nine days following the two biopsies, contrast-enhanced computed tomography (CT) of the head and neck was performed, and the patient was referred to the local Cancer Centre.

Contrast-enhanced CT of the head and neck demonstrated a 3.0 × 1.6 × 3.4 cm soft tissue mass in the left preauricular region, showing heterogeneous enhancement, extending both superiorly and inferiorly to the external auditory canal (see Figure 1). The medial border of the mass was in direct contact

with the muscular attachments at the mandibular head, raising concern for minimal invasion. Internal neovascularity was noted within the lesion, which extended inferiorly into the superior aspect of the left parotid gland, suggesting direct contiguous spread. Superiorly, the mass was in direct contact with the temporalis muscle, with possible extension into the temporal fossa (superior to the auricle). Laterally, the lesion reached the skin surface.

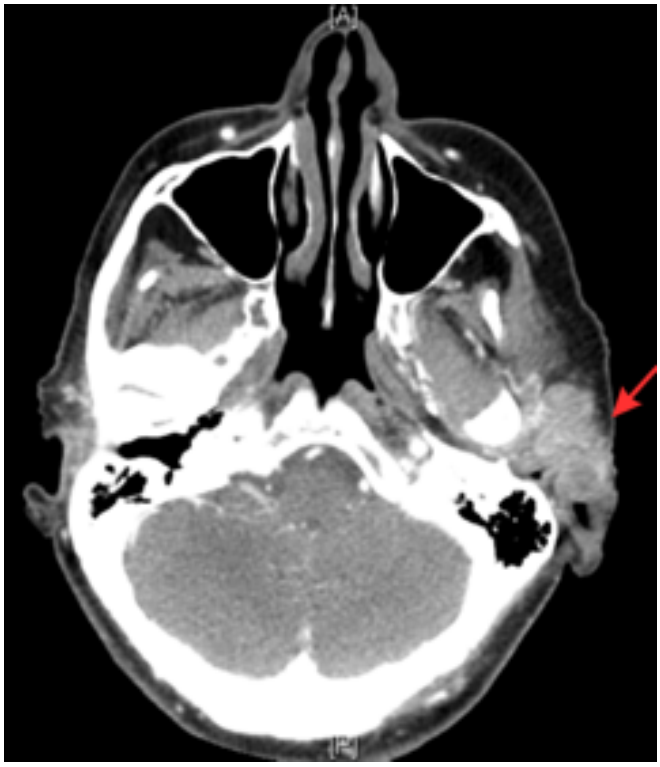

Figure 1. *An enhancing mass is seen within the left preauricular subcutaneous soft tissues, extending both above and below the external auditory canal (EAC), and inferiorly into the superior aspect of the left parotid gland (red arrow).*

A 1.4 x 1.2 cm enhancing, slightly irregularly contoured soft tissue mass was identified within the inferior portion of the left parotid gland and was noted to be suspicious for metastatic lymphadenopathy (see Figure 2). An additional 1.3 x 1.1 cm enhancing lymph node in the left level II cervical region (upper jugular lymph node group) was also concerning for nodal metastasis (see Figure 3).

No additional pathologically enlarged lymph nodes were identified. The remaining salivary glands, upper aerodigestive tract mucosa, cervical spine, and intracranial structures were unremarkable. The differential diagnosis in the radiology report (completed after histopathological diagnosis) considered "basal cell carcinoma of the EAC, squamous cell carcinoma, or even Merkel cell carcinoma", indicating MCC's uncommon presentation in this clinical context.

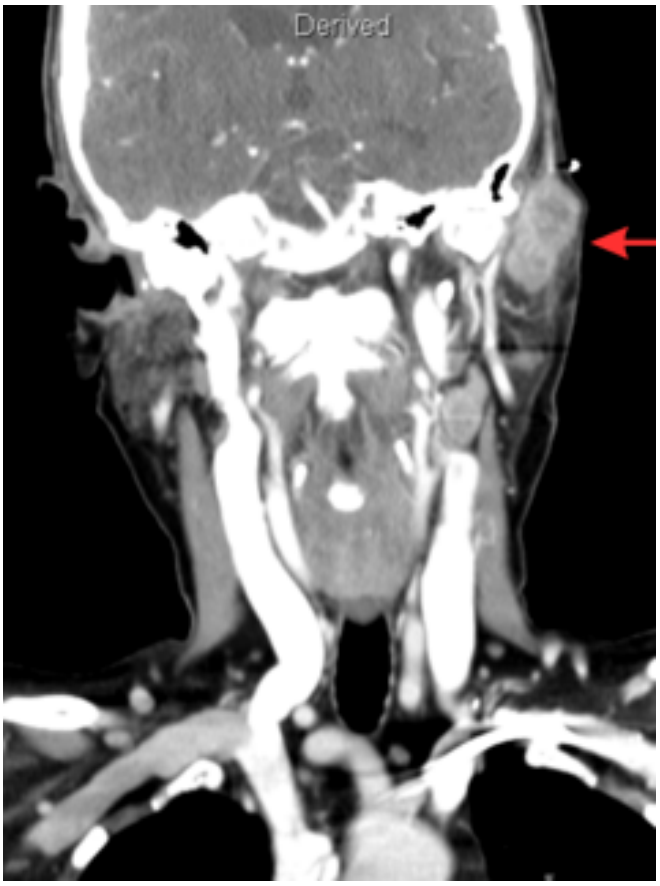

Figure 2. *A slightly irregularly contoured, enhancing soft tissue mass is seen within the left parotid gland (red arrow).*

Following initial discussion with the local Multidisciplinary Tumour Board, it was determined that primary surgery with likely adjuvant radiation therapy would be most appropriate, and a fluorodeoxyglucose positron emission tomography / computed tomography (FDG PET/CT) scan was ordered pre-operatively for staging. However, this imaging demonstrated a rapidly enlarging, intensely FDG-avid left preauricular mass with extension into the superior aspect of the parotid gland, now measuring 2.8 × 2.4 cm. A separate component extending outward from the inferior left conchal bowl region of the left ear measured 1.7 × 1.3 cm. The intense FDG avidity (reflecting increased FDG uptake) indicates high metabolic activity, consistent with aggressive tumour behaviour in this context. Both lesions had increased in size since the initial CT (see Figure 4).

Multiple FDG-avid lymph nodes were identified, including within the left parotid gland and ipsilateral cervical lymph node levels 2A, 3, 4, and 5 in the neck, raising concern for regional nodal metastases. Notably, a right-sided level II lymph node anterior to the styloid process was also intensely

FDG-avid and suspicious for contralateral nodal spread. A small focus of moderate-grade FDG avidity in segment 5 of

the liver, with a corresponding subtle hypoattenuating area on CT, raised concern for distant hepatic metastasis. However, follow-up imaging later demonstrated this lesion to be a stable, longstanding benign finding. Additional FDG avidity was observed in the C1–C3 vertebrae, bilateral acromioclavicular joints, lumbar spine, left hip, and bilateral knees. However, these findings were favoured to reflect degenerative changes, rather than metastatic disease. No concerning pulmonary nodules or aggressive osseous lesions were identified. Given these extensive, rapidly progressive findings, the decision was made to forego surgical intervention and proceed with systemic chemotherapy (4 rounds of carboplatin and etoposide) with palliative intent.

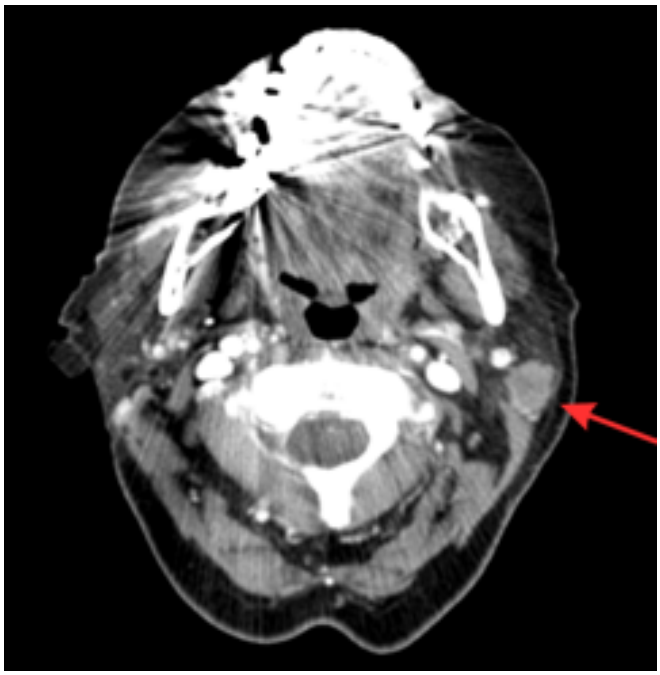

Figure 3*. A pathologically enlarged left level 2 cervical lymph node with mild enhancement is seen, concerning for nodal metastasis (red arrow).*

A follow-up FDG-PET/CT scan performed approximately 3-4 months after the initial diagnosis showed complete metabolic response to therapy, with marked reduction in the size and FDG avidity of the primary lesion in the left ear and previously involved left cervical lymph nodes. Only minimal residual low-grade FDG avidity remained, with no evidence of new or progressive disease. The previously FDG-avid liver lesion was no longer visualized, and a follow-up CT one month later confirmed no evidence of hepatic metastasis. No new FDG-avid lesions were identified in the chest, abdomen, or skeleton. A small, stable right neck lymph node with moderate FDG avidity remained unchanged and was favoured to represent a benign lesion. Adjuvant radiotherapy was recommended, which the patient is currently undergoing. A follow-up FDG-PET/CT scan is scheduled after radiotherapy to assess response and potential need for immunotherapy.

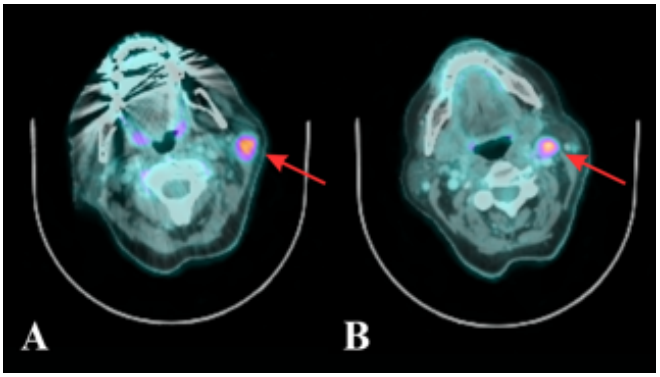

Figure 4. *A very intensely FDG-avid mass is centred within the left preauricular soft tissues, extending into the superior aspect of the left parotid gland, located just below the skin surface (red arrow) (A). A second, smaller, nodular/mass-like component extends outward from the inferior portion of the left ear, just inferior to the external auditory canal (EAC) (red arrow) (B).*

DISCUSSION

MCC is a rare, aggressive neuroendocrine carcinoma typically arising in elderly Caucasian patients.[5] This report details an unusual presentation with infiltrative involvement of the EAC and parotid gland, as MCC normally arises from sun-exposed areas of skin on the head or neck in up to 43% of cases and accounts for less than 1% of skin cancers.[5] MCC involving the EAC, and the ear region more generally, is rare and largely described in case reports.[12-14] A population-based analysis identified 210 cases of external ear MCC between 2000-2019 and found that these tumours retain the aggressive behaviour characteristic of MCC despite their rarity.[8]

MCC can be difficult to distinguish from other carcinomas on imaging alone. Here, the differential diagnosis within the radiological reports also included BCC and SCC, with MCC being considered as the rarest possibility. This reflects the non-specific imaging appearance of MCC, particularly in unlikely areas, and highlights the critical role of radiologists in considering such rare malignancies. Given this, immunohistochemistry is often viewed as the cornerstone of MCC diagnosis. However, this case is also particularly rare given that the tumour was negative for numerous stains that are normally positive. In one report, it was noted that as many as 15% of MCCs may be negative for CK20 (which stains positive in approximately 95% of cases) but are normally still CK7 positive, though the tumour described in this case was double-negative.[6-7] The suspicious liver lesion is an important teaching point to emphasize that some FDG-avid findings may represent benign entities, while still keeping a high index of suspicion for malignancy for truly concerning findings.

In keeping with its aggressive nature, this case revealed findings of metastatic disease and bilateral cervical nodal involvement. Despite this extensive spread, the patient experienced a complete metabolic response to chemotherapy. Generally, surgical excision with clear gross (<1-cm) and wide (>1-cm) margins is considered the optimal treatment for overall survival, especially in localized disease.[8] However, treatments such as chemotherapy or radiotherapy may be considered for advanced metastatic or unresectable cases within the ear, though one population-based analysis showed no significant differences in outcomes.[8] Immunotherapy is becoming an increasingly popular option, with one analysis citing improvement in mean overall survival for patients with stage 4 MCC compared to chemotherapy.[9] First-line options that have been shown to have positive response rates include pembrolizumab, avelumab, nivolumab, and more.[10]

CONCLUSIONS

Given the favourable response in this case, the value of multidisciplinary treatment of advanced metastatic MCC cannot be understated. Early recognition and an index of suspicion by healthcare providers remain essential for rare, aggressive tumours such as MCC - even with atypical imaging or immunohistochemical findings - to yield the best possible patient outcomes.

PATIENT CONSENT

Complete written informed consent was obtained and documented from the patient in this case for the preparation of this manuscript and publication of its contents, as well as the associated images, data, and anonymized information.

DISCLOSURE

All authors agree with the following statement: Neither I nor my immediate family members have a financial relationship with a commercial organization that may have a direct or indirect interest in the content stated, and there are no disclosures on my behalf. There are no funding sources to disclose in association with this content.

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