

A Unique Presentation of Merkel Cell Carcinoma with Metastases to the Peritoneum and Lung

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INTRODUCTION

- Merkel Cell Carcinoma is a highly aggressive cutaneous neuroendocrine malignancy primarily affecting elderly and immunocompromised Caucasian Fitzpatrick skin types (FST) I-III males more than females
- The annual incidence of MCC in the US continues to increase and is projected to rise to approximately 3,250 cases annually by 2025.¹
- Merkel Cell Polyomavirus is associated with MCC, and a vast majority of cases are associated with MCPyV integration into the host genome (80-90% of diagnosed individuals)²⁻⁴ as in our patient.
- Clinically, the solitary lesion can mimic benign or malignant entities, including basal cell carcinoma, lymphoma, and melanoma, so diagnosis requires microscopic examination
- The lesion has a predilection for the sun-exposed areas of the head and neck as opposed to the extremities or trunk, which are much more uncommon.
- Here we present a unique presentation of MCC of the left knee with aggressive spread to the peritoneum and lung with remission on second-line therapy.

CASE DESCRIPTION

- Case: 78-year-old FST II Caucasian male presented with acute-onset reddish rapidly enlarging boil of the left knee (Fig 1A). PMHx notable for melanoma s/p wide excision 3 years ago, COPD and HTN. He had significant sun-exposure since childhood, spending summers on the beach.
- Exam showed a reddish nodule 1.5cm in diameter with a smooth surface and telangiectasias.
- Punch biopsy revealed nests of small blue neoplastic cells with positive CK20 expression showing characteristic “perinuclear dot-like” staining pattern on IHC.
- He underwent wide local excision of the left knee (Fig 1B) with negative sentinel node biopsy in the left inguinal area as well as adjuvant radiation therapy to the thigh. He was clinically diagnosed with stage II MCC.
- Initial FDG PET scan showed high uptake in the right parotid gland, which biopsy revealed to be nonmalignant Warthin’s tumor (Fig 2)
- Surveillance PET CT within one year showed progression of MCC to the right peritoneal lymph nodes and right paratracheal lymph nodes that was biopsy confirmed. (Fig 3A-C, white arrows)
- The patient was placed on avelumab with excellent tolerance for one year (Fig 4A-B)
- He again presented with progression to the right periaortic lymph node and was transitioned to a second line regimen with pembrolizumab.
- This updated regimen resulted in complete remission without evidence of metastases or recurrence to date



Figure 1A: MCC of the left knee; **Figure 1B:** Wide local excision status post linear closure
Figure 2: FDG PET showing uptake in the R parotid gland on initial screen (arrow). Lesion was biopsy proven non-malignant Warthin’s tumor.

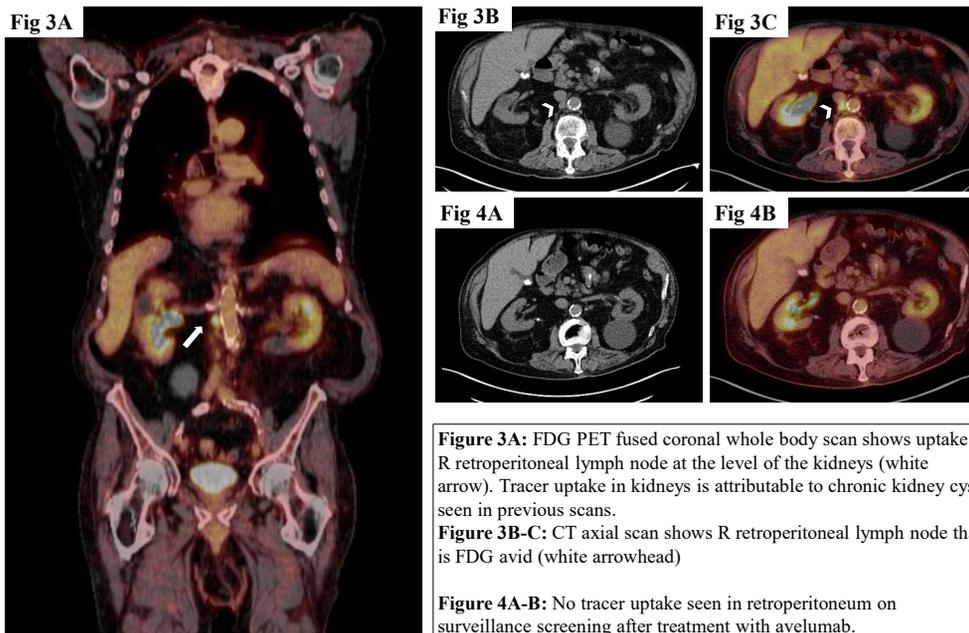


Figure 3A: FDG PET fused coronal whole body scan shows uptake in R retroperitoneal lymph node at the level of the kidneys (white arrow). Tracer uptake in kidneys is attributable to chronic kidney cysts seen in previous scans.
Figure 3B-C: CT axial scan shows R retroperitoneal lymph node that is FDG avid (white arrowhead)
Figure 4A-B: No tracer uptake seen in retroperitoneum on surveillance screening after treatment with avelumab.

DISCUSSION

- This is a unique presentation of MCC of the left knee with aggressive spread to the peritoneum as well as the paratracheal and periaortic lymph nodes.
- Risk factors include old age, seventh decade of life or older, FST I-III, sun-exposure, MCPyV, and association with other malignancies, especially multiple myeloma and chronic lymphocytic lymphoma.
- To our knowledge, this is the first case report to describe MCC dissemination to the peritoneal cavity and lung treated successfully with pembrolizumab as second-line therapy
- He has not had disease progression since starting this treatment
- The role of chemotherapy has yet to be established, especially with patients presenting with advanced disease.
- Clinical trials are currently underway on checkpoint inhibitors, such as PD1, PDL1, and CTLA-4.⁵

CONCLUSIONS

- MCC is an uncommon tumor and is typically found on the sun-exposed areas of the head and neck compared to other areas of the body
- Treatment for local disease is resection, sentinel node biopsy and possible adjuvant radiation therapy
- Chemotherapy trials are underway for advanced progression of disease, and efficacy has not been clearly established.
- Our patient has remained in remission while on a regimen of pembrolizumab for several years.
- It is important to make note of uncommon cutaneous presentations to catch disease early and start treatment promptly.

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DISCLOSURES

The authors have no financial disclosures.

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