



Case Report

Metastatic Merkel Cell Carcinoma of the Pancreas: A Case Report

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Background: Merkel cell carcinoma (MCC) is a malignant primary cutaneous neuroendocrine tumor with high propensity for local recurrence and distant metastasis. It commonly involves skin, brain, bone, liver and lung. Distant metastasis to the gastrointestinal tract, including pancreas, is very rare.

Presentation of Case: In this article we report a case of a 59 year old male with history of left gluteal skin MCC which presented with metastatic involvement of body of pancreas 5 months after the diagnosis of primary tumor.

Conclusion: Although MCC metastases to pancreas is rare, it should be kept in mind in patients with abdominal complaints and past history of MCC.

Keywords: Merkel cell carcinoma; Pancreas; Metastatic involvement

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Introduction

Merkel cell carcinoma (MCC) is a primary cutaneous neuroendocrine carcinoma. It is a rare malignant tumor of the skin and is more common in older ages and in men. MCC has an aggressive nature and a poor prognosis. It can involve regional lymph nodes and it has a high local recurrence and distant metastasis rate.

Distant metastasis occurs in one third of the patients and usually involves skin, liver, lung, bone, and brain. Metastasis to the gastrointestinal tract is very rare and there are very few reports of metastasis to pancreas, rectum, small intestine and stomach. Here, we report a case of metastatic Merkel cell carcinoma of pancreas in a 59 year old male.

Case report

A 59 year old male was admitted to the hospital because of a left gluteal skin mass which had been resected in another medical center and pathologic examination had shown Merkel cell carcinoma with tumoral involvement of all margins. Left inguinal lymph nodes were clinically positive. There was no evidence of distant metastasis in abdominopelvic and thoracic CT scan. A bone scan was done which was normal. With the diagnosis of Merkel cell tumor, the patient underwent wide local re-excision of the residual gluteal tumor and superficial and deep inguinal lymph nodes was dissected. MCC was confirmed histologically and 3 lymph nodes were involved with extracapsular extension. The patient received adjuvant chemoradiation which finished 5 months after the diagnosis of primary tumor. After completion of chemoradiation course, a MRI of the left gluteal region and a PET-CT for detection of local recurrence and distance metastases was done, respectively. MRI showed a region of fluid collection measuring 2x7x10cm, probably due to prior surgery, and no evidence of local recurrence in left gluteal region; But in the PET-CT a mild to moderately increased FDG accumulation measuring approximately 34x42mm was determined in the body of pancreas which was suspicious of malignancy (figure1). An endoscopic ultrasonography was done which reported a round shape malignant type tumor at the body of pancreas (figure2). The reported mass was biopsied with fine needle aspiration.

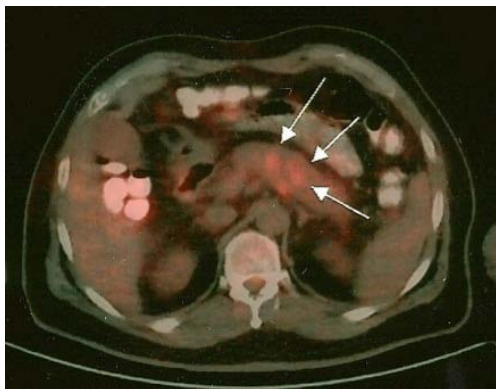


Figure1 Abdominal PET-CT which shows increased FDG accumulation in the body of pancreas.



Figure 2 Endoscopic ultrasonography of the pancreas showing its tumoral involvement.

Pathology report showed adenocarcinoma of the pancreas. With the probable diagnosis of metastatic pancreatic adenocarcinoma the patient underwent exploratory laparoscopy, in which pancreatic mass was confirmed. The mass was not resectable, so it was biopsied. Pathologic examination showed malignant infiltrative tumor with atypical nuclei bearing fine dusty chromatin and high mitotic activity. In IHC examination, tumoral cells showed diffuse and strong reaction for NSE, chromogranin, synaptophysin and CK20, And so metastatic Merkel cell carcinoma was confirmed. The patient was referred to the chemotherapists to receive another chemotherapy course.

Discussion

Metastasis in Merkel cell carcinoma (MCC) is common because of its invasive nature. Skin, liver, lung, bone and brain are usual sites of metastatic involvement; but it rarely involves gastrointestinal tract and there are few reports of involvement of rectum, stomach, small intestine, colon and pancreas in the literature. (1-8). In few reports (2-7) metastatic involvement of pancreas by MCC is reported and in our literature review, we found 6 cases of metastatic MCC which involved pancreas.

Bachmeyer et al have reported a case of a 57 year old male with MCC of left lower eyelid, which underwent wide local excision and adjuvant radiotherapy. 5 months after that the patient was admitted for abdominal pain and jaundice and an abdominal CT scan revealed a large cystic tumor of the body of the pancreas which was biopsied and diagnosed as pancreatic metastatic MCC. The patient did not undergo resection of tumor because of the peritoneal carcinomatosis at the laparotomy and was referred for the chemotherapy but he died before chemotherapy was begun (2).

In another study by Adsay et al, pathology material from 973 surgical specimens and 4955 adult autopsy cases was reviewed to identify metastatic pancreatic tumors. In surgical specimens 38 cases contained metastatic tumors to the pancreas which one of them was MCC (3). Bachman and colleagues, reported pancreatic metastasis of MCC in a 82 year old female with history of right eyebrow MCC 2 years ago which had been resected and received post operative radiotherapy. The patient underwent distal pancreatectomy and splenectomy and resection of splenic flexure of colon and histopathology examination showed metastatic MCC and a concomitant insulinoma (4).

In another report by Dim et al a 79 year old female with history of upper extremity MCC 15 months earlier, presented with a pancreatic mass. She underwent endoscopic ultrasound guided fine needle aspiration and metastatic MCC was diagnosed based on the histologic and IHC features (5). Ouellette et al, also reported a case of a 64 year old male with past history of MCC of finger presented with obstructive jaundice 2 years after the primary MCC. He underwent pancreaticoduodenectomy and the pathology reports confirmed metastatic MCC (6). In 2010 Krejci et al reported a case of a 62 year old male with a history of combined kidney and pancreas transplantation which developed a right gluteal MCC 8 years after the transplantation. The patient

underwent resection without adjuvant radiotherapy (because of the risk of graft rejection), but tumor metastasized to the pancreatic graft leading to its failure and despite chemotherapy the patient died 9 months after the diagnosis (7).

These reports show that despite the scant reports of gastrointestinal and especially pancreas involvement by the MCC, this diagnosis should be kept in mind specially in patients with past history of MCC which presents with abdominal symptoms. Although in our case the patient did not have any symptoms and the diagnosis was made during follow up.

Lymph node involvement is one of the most important factors which predict distant metastasis in MCC and patients with positive LNs are in greater risk of developing distant metastasis (4). This relation is confirmed in this report, as the patient had involved lymph nodes at first disease presentation. The time between the diagnosis of primary MCC and metastatic MCC is reported to be 4 to 24 months in different cases (2, 4-8). In our report, metastasis was diagnosed 5 months after the primary disease, although the patient was not symptomatic and diagnosis was made by follow up imaging. The occurrence of most metastasis in the first 2 years after primary tumor, underscores the need for more intensive surveillance in this period.

Different treatment options, like radical resection or chemotherapy, have been used in cases of metastatic MCC. There are reports which show the effectiveness of both modalities. Some have proposed chemotherapy as an effective modality in the treatment of metastatic MCC; On the other hand, some others have suggested that radical resection should be considered in such cases if possible (4, 6). They suggest that R0 excision of the metastatic MCC can provide survival benefit; However, this needs to be elucidated in future studies.

In conclusion, MCC is a rare malignant primary neuroendocrine tumor of the skin, with high propensity for local recurrence and distant metastasis.

Also metastatic involvement of gastrointestinal tract, especially pancreas, is rarely reported but should be kept in mind in patients with abdominal complaints and past history of MCC. The time between primary tumor and distant metastasis is quite different but some reports show that most of them occur in the first 2 years. Different treatment options are proposed. Some reports suggest radical resection whenever possible, with benefit of increasing survival and some recommend chemotherapy as the beneficial treatment. More investigations are needed to elucidate the best treatment option for metastatic MCC.

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