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Case Report

Cardiac metastases from merkel cell cancer: A case report and review of literature

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Case report

A 61 - year-old physically fit and athletic man presented to his dermatologist with a 10 mm raised, dark lesion on the left side of his neck. A complete skin examination did not show any other abnormal areas of skin. Pathology was found consistent with Merkel cell cancer, and the patient was referred to surgery for a wide local excision and sentinel lymph node biopsy. A PET scan did not show any other areas of concern. At surgery, one of two sentinel lymph nodes was found to be involved with Merkel cell cancer and the patient received postoperative radiation.

After that, he remained in observation and did well but one year later, a PET scan showed a suspicious lesion in the right lobe of the liver. A core needle biopsy confirmed metastatic high-grade neuroendocrine cancer consistent with Merkel cell cancer.

The patient enrolled in a clinical trial using immunotherapy (Ipilimumab and Nivolumab) with a complete clinical and radiological response.

He went on to do well for a year and a half with no treatment. One day, he was out for a morning run when he suddenly got very short of breath and had to be brought to the Emergency Room. He was found to have intracardiac metastases to the right atrium, right ventricle, and pericardial effusion. The pericardial effusion was drained, and the patient went on to receive radiation for the intracardiac lesions. He was given 2000 cGy in 5 fractions. He then restarted immunotherapy again with Ipilimumab and Nivolumab.

Unfortunately, although he did not develop any further cardiac metastases and initially appeared to have good disease control, 6 months into his treatment he developed GI bleeding. Endoscopy was done and stomach and duodenal biopsies were obtained.

These showed widespread infiltration with Merkel cell cancer. He also developed acute renal failure, with a kidney biopsy also confirming the same. He also had marked testicular pain and most likely had testicular involvement as well.

He passed away a few days later.

Review of literature

Merkel cell cancer (MCC) is a rare cutaneous neuroendocrine malignancy that is generally seen in older males with light skin. It generally follows an aggressive course and has a high metastatic potential [1].

MCC has been causally linked to the Merkel cell polyomavirus. This is a double-stranded deoxyribonucleic acid virus and in a meta-analysis, found in 79% of all MCC cases [2]. Carcinogenesis in the absence of Merkel cell polyomavirus is related to ultraviolet light exposure [3].

Data from the Surveillance, Epidemiology and End Results (SEER) database suggests the incidence is rising as the population ages. A tripling of the overall incidence between 1986 and 2000 has been noted [4].

It is predicted that by 2025, 3284 cases will be diagnosed in the United States.

Nearly half (50%) of the patients present with localized disease, but recurrence rates are high and mortality rates are in the range of 33% - 46% [5].

Before the advent of immunotherapy, the treatment for

metastatic MCC was chemotherapy, using regimens generally employed for metastatic small cell lung cancer, with very infrequent (< 10%) durable responses [6].

While lymph nodes, skin, lung, central nervous system and bone are common sites of metastases, cardiac, renal, gastrointestinal, or testicular metastases as seen in our patient are quite rare.

We searched the literature for cardiac metastases from MCC as that is how our patient presented when he developed metastatic disease. Only about a dozen cases of MCC with cardiac metastases have been reported. Interestingly, cardiac metastases from various cancers are not a very rare event in autopsy series. In fact, while primary cardiac tumors are exceedingly rare with incidence rates between 0.001% - 0.28%, cardiac metastases were found to be present in autopsy series in the range of 2.3% to 18.3%. [7], Malignancies with a high prevalence of cardiac metastases include mesothelioma, melanoma, lung cancer, breast cancers, soft tissue sarcomas, lymphomas and leukemias.

Cardiac metastases are most commonly found in the pericardium, followed by the epicardium and myocardium. The most common clinical manifestations are pericardial effusion, tachyarrhythmias, atrioventricular block, and congestive heart failure [8].

Rarely, occlusion of a coronary artery may occur causing ischemia.

The diagnosis of cardiac metastases can be challenging, but worth considering in an MCC patient presenting with cardiac or respiratory symptoms [9]. As pericardial effusions can occur, drainage of the pericardial effusion with cytology may be a good first step. Indeed, this is how we were able to confirm the presence of cardiac metastases in our patient. If the pericardial fluid had not yielded the diagnosis, our plan was to proceed with cardiac catheterization and biopsy for tissue diagnosis.

Since cardiac metastases from Merkel cells have been rarely reported, there is no standard treatment approach. After extensive consultation with outside institutions and experts, we elected to treat our patient with irradiation to the cardiac lesions. Then he was treated with immunotherapy using Ipilimumab 3 mg/kg and Nivolumab 1 mg/kg.

As already mentioned, he initially did well but passed away 8 months after the presentation.

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