Spontaneous Regression of Metastatic Merkel Cell Carcinoma

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Abstract
Merkel cell carcinoma is a rare aggressive neuroendocrine carcinoma of the skin predominantly affecting elderly Caucasians. It has a high rate of local recurrence and regional lymph node metastases. It is associated with a poor prognosis. Complete spontaneous regression of Merkel cell carcinoma has been reported but is a poorly understood phenomenon. Here we present a case of complete spontaneous regression of metastatic Merkel cell carcinoma demonstrating a markedly different pattern of events from those previously published.

Case Report
A 60-year-old Caucasian Irish woman presented in October 2003 with a 3 x 4.5cm lump in her right groin. An excisional biopsy was performed and histology revealed poorly differentiated carcinoma, forming nests suggestive of neuroendocrine origin. Immunostains were positive for synaptophysin, HMB-45, chromogranin and cytokeratin 20, with a characteristic dot like pattern, consistent with metastatic Merkel cell carcinoma. No primary lesion of the skin was identified. The initial computed tomography staging scan was consistent with a previous excisional biopsy and showed an additional lymph node in the right groin. An incidental dermoid cyst was noted in the right ovary.

Figure 1: Lymph node biopsy of poorly differentiated carcinoma, forming nests suggestive of neuroendocrine origin. HE original magnification x 20

Subsequent CT surveillance revealed a gradual increase in size of the right inguinal node. At 15 months post initial diagnosis there was significant disease progression with increased size of the right inguinal node from 1.5 cm initially to 4.5 cm. There were further enlarged pelvic nodes with new right external iliac nodes (the largest measuring 5.0cm), a right common iliac (1.6cm) and a left inguinal node measuring 4.0cm. Surveillance scans continued and in July 2005, 20 months following initial diagnosis, CT showed no evidence of recurrent or metastatic disease. No enlarged lymph nodes were visible in the pelvis consistent with complete spontaneous regression. Her regular prescription at the time of complete spontaneous regression included lisuride and rabeprazole in addition to the previously mentioned psychiatric medications. Four years since spontaneous regression all follow up CT scans show that she remains in complete radiographic remission.

Discussion
This is the 17th case of complete spontaneous regression of Merkel cell carcinoma to be reported. Previous reports have noted a correlation between spontaneous regression and the site of the primary lesion being in the head or neck (94% of cases). In addition, the patients were female in the majority of cases (72%), and all cases except one underwent biopsy of the primary cancer prior to subsequent regression. The clinical features of the current report differ in two significant ways. Firstly, the primary lesion was in the lower body, as opposed to the head or neck. Secondly, the sequence of events leading to biopsy differed from the previous reports. In this case, it was noted that the patient had lymph nodes which regressed following biopsy.

This would mean that complete spontaneous regression of the primary lesion occurred independent of surgical intervention. Furthermore, in this report, following lymph node biopsy, the disease actually progressed further before regressing, whereas in the other reported cases it simply regressed following biopsy.

A further consideration, regarding the absence of a primary at diagnosis, is that it may have been within the dermoid cyst. Although a Merkel cell carcinoma has been reported but is a poorly understood phenomenon. Here we present a case of complete spontaneous regression of metastatic Merkel cell carcinoma, there was no identifiable primary lesion. In addition, the progression of disease subsequent to biopsy in this case would lead us to raise the possibility that complete spontaneous regression occurred here both in the primary cancer and in the nodal disease independently of one another.

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References