


CASE REPORT



Primary malignant melanoma of the lung: a case report of a rare tumor and review of the literature

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ABSTRACT

Primary malignant melanoma of the lung (PMML) is a rare malignancy that exhibits aggressive behavior and has a very poor prognosis. We are reporting on a case of PMML in an otherwise healthy 22-year-old Caucasian male with no significant past medical history and an unremarkable family history. The patient initially presented with a 2-month history of a cough and an unexplained 22-lb weight loss. His initial chest X-ray demonstrated opacification of the right lower lobe (RLL) of his lung and a subsequent computerized tomography scan (CT scan) of his lung revealed a large mass occupying most of his RLL (Figure 1). The patient subsequently underwent a bronchoscopy with endobronchial ultrasound. Biopsies revealed a poorly differentiated carcinoma. A positron emission tomography with low dose CT scan was performed per protocol and revealed an intensely hypermetabolic tumor with no evidence for lymphatic disease or extra-thoracic spread. The patient underwent a surgical exploration and a right lower lobectomy with a thoracic lymphadenectomy. The pathology including immunohistochemical stains demonstrated a malignant melanoma with no lymph node involvement. A physical examination including ophthalmic, mucosal, and skin examinations revealed no evidence for an extra-thoracic site of the disease. The patient had negative margins for resection and did not receive any adjuvant therapy and is alive and well with no evidence for recurrence 3 years after the resection.

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1. Introduction

The most fatal cutaneous neoplasm of the skin is malignant melanoma (MM) and it most commonly arises from a focally uncontrolled growth of melanocytes [1]. MM may arise from multiple extra cutaneous sites including the esophagus, and the liver; however, MM arising from the lung is extremely rare and accounts for only 0.01% of all primary lung tumors [2]. MM can be very aggressive and can metastasize early in the disease process. Before solidifying a diagnosis of primary malignant melanoma of the lung (PMML), other sites of origin such as the skin, the eyes, and the mucosal surfaces have to be ruled out for primary melanoma. The English medical literature reports approximately 40 cases [3].

2. Case

A 22-year-old Caucasian male who had never smoked and had an unremarkable past history presented with a 22-lb unexplained weight loss and persistent nonproductive cough of 3-month duration. He underwent a chest X-ray, which demonstrated subtotal opacification of the right lower lobe (RLL). He was referred to our

hospital. He initially underwent a computerized tomography (CT) scan of the lung with intravenous contrast, which revealed a 7 × 7 cm mass in his RLL that was initially treated as pneumonia (Figure 1). His symptoms and radiological findings persisted despite the treatment. He underwent a bronchoscopy with endobronchial ultrasound (EBUS), which revealed patient's right-sided airways although the RLL segmental orifices demonstrated some degree of what appeared to be extrinsic compression. In addition, endobronchial ultrasonography in this distribution demonstrated a hazy-bordered, heterogeneous echo density. Biopsies sent for pathological evaluation were consistent with a poorly differentiated carcinoma of unknown etiology. A right thoracotomy was performed with a right lower lobectomy and a thoracic lymphadenectomy including stations 4R, 8R, and 9R. CT of patient's chest confirmed the total removal of the lesion and the margins were free of tumor. The pathology revealed a tumor composed of predominantly epithelioid tumor cells. There were junctional changes and mitotic activity with the characteristic nesting of tumor cells beneath the bronchial mucosa. These findings were all suggestive of MM. Immunohistochemical stains supported this diagnosis.

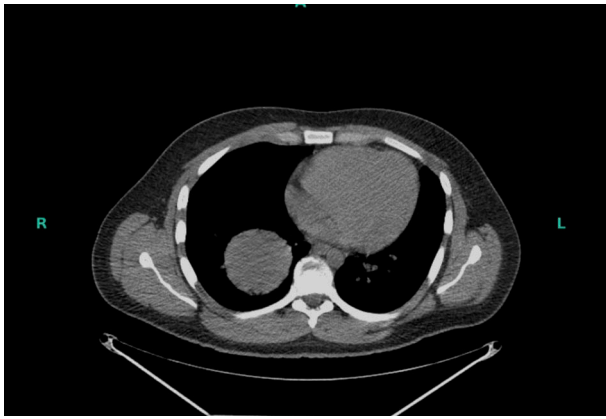


Figure 1. CT of chest shows a mass measuring approximately 7 × 7 cm in right lower lobe.

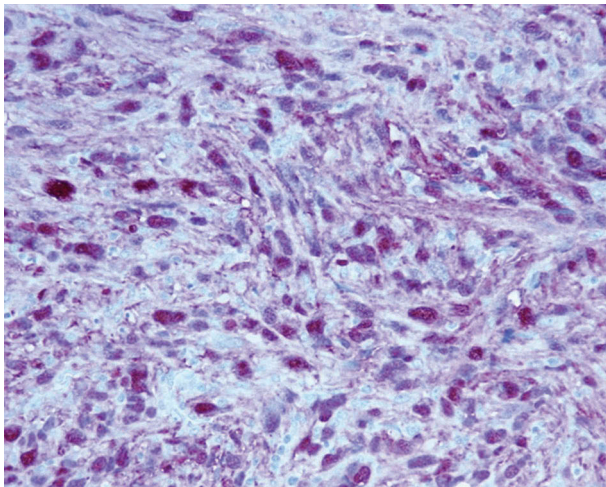


Figure 2. Resected lung tissue stain positive for S-100.

The S-100 (Figure 2), melanocyte-associated monoclonal antibody (MSA), HMB45, and alpha-smooth muscle actin (α -SMA) were all positive. Cytokeratin, epithelial membrane antigen and calponin stains were all negative. The hilar and mediastinal nodes did not have any evidence of lymphatic metastasis. The patient did not have any evidence for cutaneous, ocular, or mucosal melanoma and has been followed for over 3 years with serial CT scan of lung every 6 months and has no clinical or radiologic evidence for recurrence. CT scan of patient's chest done 24 months after resection showed no evidence of disease (Figure 3). Of interest, is the fact that his father recently developed a cutaneous melanoma of the neck.

3. Discussion

Metastatic melanoma of the lung not uncommonly occurs in patients with a primary cutaneous melanoma. Melanoma of the lung is metastatic until proven otherwise. In patients who present with a melanoma of the lung, a detailed ophthalmic, skin, and mucosal examinations need to be performed to exclude the possibility of an extra-thoracic primary

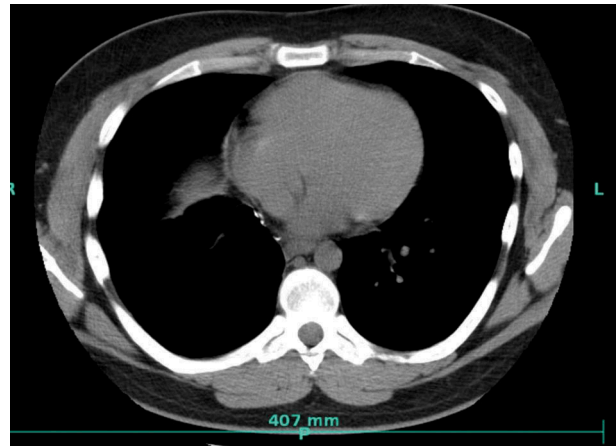


Figure 3. CT of chest shows postsurgical changes (24 months after surgery) with no disease activity.

site. Approximately 0.01% of all primary lung tumors are due to PMML. The etiology of PMML is poorly understood. One of the possibilities is that epithelial cells exist in areas of squamous metaplasia in the lung and undergo differentiation to melanocytes and ultimately develop into MMs [4]. Cigarette smoking is one of the causes of squamous metaplasia of the lung and PMML may in some cases be caused by cigarette smoking. Clinically, patients with PMML present with cough, post obstructive pneumonia, hemoptysis, atelectasis, weight loss, and fatigue [5]. Some patients, however, may be asymptomatic [6]. The clinical and radiologic differentiation between a PMML and a bronchogenic carcinoma is challenging. A thorough physical examination including detailed cutaneous, mucosal, and ocular examinations is a necessity. The Armed Forces Institute of Pathology has recommended the following clinical and pathological criterion: (1) a solitary lung tumor; (2) a MM confirmed by immunohistochemistry or electron microscopy; (3) no past history of excision or fulguration of a cutaneous, mucosal, or ocular lesion; (4) a central pulmonary lesion; and (5) no demonstrable tumor elsewhere at the time of diagnosis [4].

This patient met all of those criteria. Bronchoscopy including EBUS may help in establishing a diagnosis but the small amount of tissue obtained may preclude the establishment of a definitive diagnosis. Whenever possible, an anatomic resection should be performed which will also supply enough tissue for the pathologic determination of melanoma. The exact nature of a cleanly resected PMLL is still unclear and we are recommending a 5-year surveillance period with CT scans of chest performed at 6-month intervals.

There are no clear guidelines regarding treatment of PMML from Chest society or in The National Comprehensive Cancer Network (NCCN) guidelines given the rarity of disease.

A recently published article reviewed 40 cases of PMML from the literature. Of the total cases, 21 of

them did not receive any chemotherapy or radiation while 15 of them received chemotherapy as adjuvant and 4 of them received radiation. In regard to mortality, 26 of them died at various times after surgery, status of one patient was not reported, and remaining were alive at the various time frame after surgical resection [7].

Our patient was young, had clear surgical margins, and had no history of prior melanoma. Thus, he was observed and he has been symptom free and alive 3 years after surgery. Further studies need to be done in order to define better treatment modalities.

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Author's contribution

M. Yunce completed the background research, drafted and edited the manuscript, S. Selinger, W. Krinsky, and DP. Harley edited the manuscript.

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Informed consent

Informed consent was obtained from both patients for educational use of the below mentioned data and no personal patient information has been disclosed.

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