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Case Report
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Primary mediastinal melanoma presenting as superior vena cava syndrome: A case study

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ABSTRACT

The rates of melanoma have increased over the past 30 years. Malignant melanoma most commonly occurs in the skin with secondary involvement of other organs. Here, we present an extremely rare case of malignant melanoma of the mediastinum with presentation of superior vena cava syndrome without clinical evidence of extrathoracic disease. The incidence of this clinical presentation is uncommon, resulting in only a handful of case reports in the literature.

Key words: *Mediastinum, mediastinal tumor, melanoma, superior vena cava syndrome*

Introduction

Melanomas are malignant tumors that are primarily skin lesions and account for approximately 2% of skin tumors. Extracutaneous malignant melanomas (ECMs) accounts for only 4-5% of all primary melanomas, with an incidence of approximately 0.7/100,000 new cases per year [1]. Given the rarity of this disease, ECMs require special consideration within the field of oncology due to the often late diagnosis, which results in a poor prognosis. Furthermore, malignant melanoma presenting as anterior mediastinal mass, with symptoms of superior vena cava (SVC) syndrome is uncommon [2-4]. SVC syndrome occurs in approximately 15,000 persons in the United States each year, with malignant causes being the etiology in more than 90% of the cases. Given the rarity of the disease, the diagnosis, staging, and treatment of ECMs are not well established.

Case Report

A 44-year-old Caucasian male presented to our clinic for evaluation of a chronic cough of a 2-year duration and no prior history of smoking. He was initially treated with inhalers for presumed asthma. Over 1 year prior to diagnosis, a chest X-ray (Figure 1a) showed mediastinal adenopathy for a presumed diagnosis of sarcoidosis. The patient was started on prednisone; however, his orthopnea and dyspnea progressed and he developed voice hoarseness. A computerized tomography (CT) scan demonstrated bulky mediastinal adenopathy, including subcarinal, pretracheal, and

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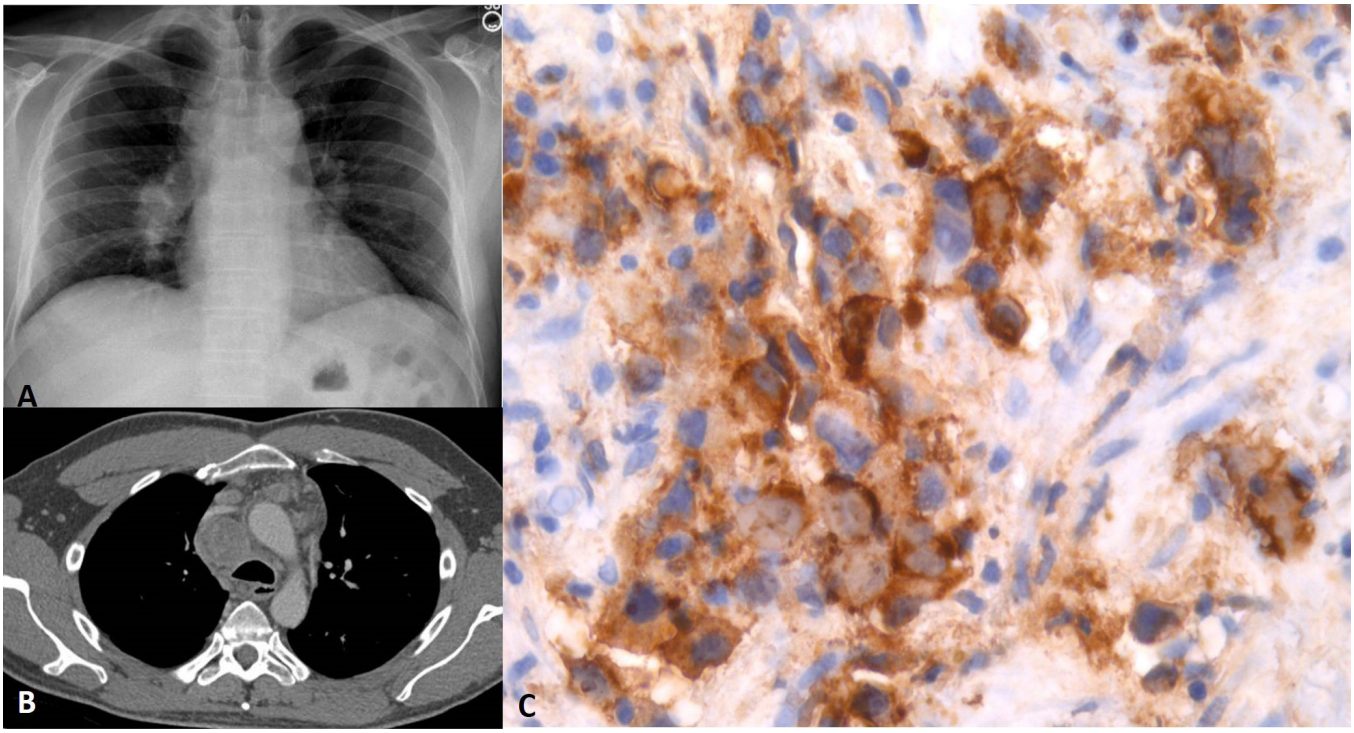

Figure 1. (a) Chest - X-ray with notable hilar adenopathy and mild tracheal deviation, **(b)** Venogram computerized tomography scan noting an anterior heterogeneous mediastinal mass resulting in superior vena cava compression, **(c)** Intense and diffuse cytoplasmic staining with S100 in mediastinal melanoma (×40).

right hilar adenopathy. The patient was evaluated in the thoracic surgery clinic, and diagnosed with SVC syndrome. A CT venogram (Figure 1b) noted significant luminal compromise of the SVC. Following an unsuccessful bronchoscopy biopsy, the patient underwent a mediastinoscopy. His tissue sample was largely necrotic with viable areas demonstrating sheets and nests of discohesive tumor cells with eccentrically placed nuclei, ample eosinophilic cytoplasm, and marked nuclear pleomorphism. The specific mitotic count was unable to be determined given the extensive necrosis. He was treated with emergent palliative radiation. Further investigation for primary or secondary was negative, and dermatology noted no primary skin lesions. Prior to initiation of treatment, his lactate dehydrogenase (LDH), a marker highly predictive of survival, was 202 U/L, which is just outside the high normal range of 198 U/L.

The patient was diagnosed with Stage IV M1a melanoma, as determined by tumor-node-metastasis staging of the mucosal melanoma of the head and neck. The tumor had a p53 mutation with an unknown primary that stained strongly for S100 (Figure 1c) and weakly for tyrosinase. He underwent chemotherapy and radiation with subjective improvement. At 6 months, he had resolution of his SVC syndrome with evidence of disease reduction by CT, as well as a decrease in LDH to 186 U/L. He noted significant improvement in his voice, and a decrease in orthopnea and facial swelling.

Discussion

The incidence of melanoma has been on the rise worldwide. It more commonly presents as a skin lesion. Primary melanoma in the chest is rare with only about 15 cases reported [2-5]; however, there is reportedly a 4% incidence of unknown mediastinal primary lesions [5]. As presented above, melanoma can be seen in the mediastinum, but the differential diagnosis also includes extraadrenal paraganglioma, carcinoid tumor of the thymus, and schwannoma [6].

Despite being uncommon, it is not surprising that the malignant melanoma occurs as a primary lesion in the mediastinum due to embryological development. Melanocytes and autonomic ganglion cells are derived from neural crest cells, with the latter being abundant in the mediastinum. Given a shared progenitor cell, melanocytes can be tracked to within the mediastinum. Both the prognosis and management of primary mediastinal malignant melanomas are unknown due to the rarity of the disease; however, it is considered to be

an aggressive course. The Clark level and Braslow index are commonly used to evaluate and stage cutaneous melanoma, but is not applicable to ECM [6]. The American Committee on Cancer has provided some guidelines on ECMs, particularly mucosal lesions as were present in this case.

Radiotherapy is initiated to control the local spread of a tumor that is often not amenable to surgery; however, when possible, complete surgical excision is the treatment of choice. For this patient, given the aggressive nature of the tumor and the known poor prognosis, non-surgical intervention was initially pursued. Nevertheless, the role of surgical bypass grafting is infrequently used to treat SVC regardless of the underlying etiology. Overall, given the rarity of the disease much progress is needed. First, an appropriate staging system for non-cutaneous locations is needed to improve the planning of treatment and the prognostic factors in patients, and most importantly, to permit meaningful comparison of outcomes for patients and clinicians. The ideal curative approach would involve neoadjuvant chemotherapy, surgical resection and reconstruction, and post-operative radiotherapy.

Conflict of interest statement

The authors have no conflicts of interest to declare.

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