Desmoplastic Malignant Melanoma Presenting as a Lung Mass

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A case of a metastatic desmoplastic malignant melanoma is reported. The patient presented initially with a lung mass and subsequently developed facial swelling and numbness secondary to tumor involvement of the maxillary division of the trigeminal nerve. The pleomorphism, histochemistry, and schwannoid differentiation of these tumors is discussed.

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Abbreviations: DMM=desmoplastic malignant melanoma

We describe a case in which a patient was noted to have a lung mass 2 years prior to his presentation with a trigeminal neuralgia and facial swelling that was subsequently diagnosed as a neurotropic malignant nerve sheath tumor metastatic to the lung. This case demonstrates an unusual presentation of a rare variant of desmoplastic malignant melanoma (DMM).

CASE REPORT

A 63-year-old man without a smoking history was incidentally noted to have a 3-cm left hilar mass on routine chest radiograph when admitted to the hospital for bilateral hernia repair in 1993. Chest CT scan confirmed a smooth mass located in the left upper lobe. Bronchoscopy was performed at that time, which reportedly showed no endobronchial lesions and no cytologic abnormalities. He subsequently presented in January 1995 with complaints of left facial numbness, and he was treated with carbamazepine for trigeminal neuralgia. Four months later, he was admitted to the hospital with nausea, emesis, and severe left facial pain. The facial pain was treated effectively with narcotics. The following month he noted left facial asymmetry and was seen by an ear, nose, and throat physician. Head CT and MRI showed a 9×3×3-cm mass extending along the entire course of the left maxillary division of the trigeminal nerve (Fig 1). This lesion was centered within the skull base at the level of the left cavernous sinus, surrounded by the internal carotid artery within the cavernous sinus, extended into the posterior cranial fossa, and involved the inferior aspect of the orbit. Fine-needle aspirate of the mass revealed a spindle cell neoplasm that could not be further characterized. The patient was referred to Barnes Hospital where another biopsy specimen of the lesion with special stains and electron microscopy revealed the diagnosis of malignant schwannoma (S-100 positive and HMB-45 negative).

On hospital admission chest radiograph, a 4-cm mass was again noted in the left upper lobe. He had no history of tobacco use and had no pulmonary complaints at the time of this hospital admission. His physical examination was remarkable for a left ptosis, left facial swelling, and sensory defect in the trigeminal distribution. His lungs were clear to auscultation and findings from the rest of his examination were otherwise unremarkable. A chest CT scan demonstrated a 4-cm well-defined round mass in the left upper lobe without associated adenopathy (Fig 2). The patient subsequently underwent bronchoscopy that showed a smooth mass bulging into the lumen of the anterior segment of the left upper lobe. Pathologic study proved the mass to be a malignant melanoma, and immunohistochemical stains were strongly positive for vimentin, S-100, and HMB-45. The pathologic condition of the facial mass and the lung mass were reviewed together and were believed to have very similar features (Fig 3), the only difference being the facial mass. Immunocytochemistry was S-100 positive but HMB-45 negative. Although the immunocytochemistry is different between the two lesions, it is believed that this represents a nerve-centered DMM.

Discussion

It is well recognized that malignant melanoma may
express various different phenotypes, particularly in its invasive stage. The spindle cell phenotype may demonstrate neurosustentacular (perineural or Schwann cell differentiation) and/or fibrogenic properties that can mask the true histogenesis of the neoplasm.1

DMM is a rare variant of malignant melanoma that was first described by Conley et al2 in 1971 as an invasive melanoma made up of atypical melanocytes and strands of elongated spindle-shaped cells. Reed and Leonard3 later described neural involvement in 22 cases of DMM. Neural involvement may be of three types: local invasion of neural tissue, distant spread along the perineurium (as seen in this case), or actual differentiation into neural tissue. There is speculation that melanocytes have the potential to differentiate into fibrous and neural tissue because of the common embryologic origin of these cells.4 In a study of 45 cases of DMM by Jain and Allen,5 12 were predominately nerve-centered tumors, 3 of which were previously reported as malignant schwannomas. All were located in sun-exposed areas, and all but one were on the head and neck and involved cranial nerves. Interestingly, the mean age reported was 53 years (range, 42 to 69 years) and there was an 11:1 male predominance.5 Most recently, Skelton et al6 performed a histologic analysis with partial follow-up of 128 cases of DMM derived from the files of the Armed Forces Institute of Pathology, Washington, DC. Clinical features included masses with or without an overlying pigmented epidermal lesion. Histologically there were fusiform spindle cell tumors with abundant collagen and low cellularity. Immunocytochemistry showed all lesions were HMB-45 negative. Factors that correlated with survival included tumor location, sex, tumor depth, and presence of stromal mucin. Overall 5-year survival rate following surgical excision was 61%, compared with 40% for other melanomas. For head and neck DMMs invading deeper than 4 mm, as in this case, the 5-year survival rate was reported as 38%. In this series, 12 cases were available for follow-up; 1 of these patients had metastatic disease to the lung and died within 4 years.1 It is not at all unusual that the metastatic lesion to the lung displays features of the usual melanoma rather than that of the primary neurotropic melanoma.2,3 The pattern of the metastatic lesion is typically the usual melanoma with variable melanogenesis or the desmoplastic variant.

Our patient is currently being treated according to the protocol of McClay et al,6 which includes combination chemotherapy with dacarbazine, cisplatin, carbustine, and tamoxifen. The tumor was never excised.

In summary, to our knowledge, this is the first case noted in the literature in which a patient presents with a lung mass and years later is found to have a malignant nerve sheath tumor metastatic to the lung, consistent with the diagnosis of DMM.

REFERENCES


Cardiopulmonary Bypass as an Adjunct to Pulmonary Surgery*

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Although performance of concomitant open heart and pulmonary operations has been described, there is

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