Dermatology Case Reports

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Case Report

Primary Leptomeningeal Melanoma Presenting in a Patient with Multiple Congenital Melanocytic Nevi

Stanley Skopit, Ramya Garlapati*

Larkin Community Hospital, South Miami, FL

ABSTRACT

Primary leptomeningeal melanoma is a rare and aggressive tumor of the CNS. They are much more common in patients with multiple congenital melanocytic nevi. Congenital Melanocytic Nevi (CMN) are commonly known as moles and are benign pigmented macules that are present at birth or appear within the first few months of life. We report a case of a 28-year-old male with multiple CMN, who was found to have a primary leptomeningeal melanoma. We present this case for its rarity and to review the current association between CMN and neurocutaneous melanosis.

Keywords: Primary Leptomeningeal Melanoma; Congenital melanocytic nevi; Tumor

INTRODUCTION

Neurocutaneous Melanosis or Melanocytosis (NCM) is a rare condition that can occur in patients with CMN. It is characterized by proliferation of melanocytes in the Central Nervous System (CNS). It includes both leptomeningeal melanosis and CNS melanosis [1]. Primary leptomeningeal melanoma is a rare tumor of the CNS that is a malignant manifestation of neurocutaneous melanosis.

CASE REPORT

A 28-year-old male presented with a 3-week history of multiple, intermittent episodes of lightheadedness, disorientation, and right-sided numbness involving the right upper and lower extremities, each lasting 20 minutes-40 minutes. The patient had a history of multiple congenital melanocytic nevi with over a hundred nevi, which have been present since birth. These nevi covered over 50% of his body, varied in size from a few millimeters up to fifteen centimeters in diameter. He has undergone multiple surgical excisions and subsequent skin grafting, with a total of 13 procedures prior to the age of five. He was last seen by a dermatologist over 10 years ago, at which point there was no suspicion of primary cutaneous melanoma.

An MRI of the brain revealed a left parieto-occipital mass with surrounding edema, adjacent to the midline (Figure 1). The patient underwent optic fundi exam, laryngoscopy, endoscopy and colonoscopy, which did not show any significant findings. PET scan reported no abnormal hypermetabolic activity. The patient was also seen by a dermatologist and a thorough skin examination

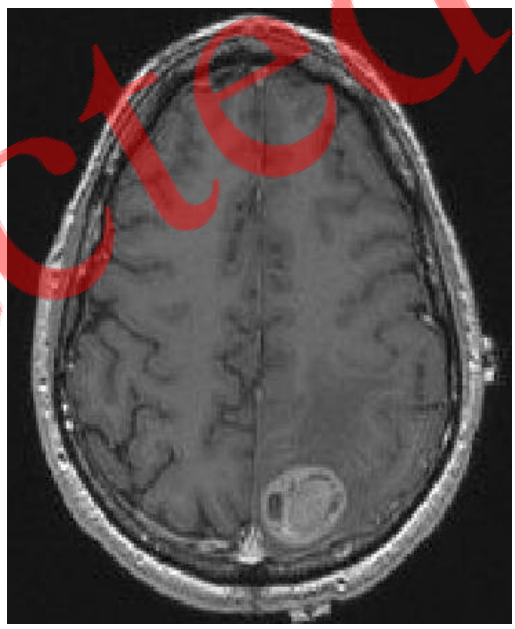

Figure 1: An MRI of the brain revealing a left parieto-occipital mass with surrounding edema, adjacent to the midline.

revealed numerous and diffuse congenital melanocytic nevi, none of which were clinically worrisome to warrant a biopsy. Therefore, work up for a primary melanoma was negative. A diagnosis of primary leptomeningeal melanoma was suspected. The patient was evaluated by neurosurgery and was started on Decadron 4 mg QID and Keppra 500 mg BID.

The patient underwent surgical excision of a 10 mm × 6 mm ×

Correspondence to: Ramya Garlapati, Larkin Community Hospital, South Miami, FL, E-mail: ramya.tripuraneni@gmail.com

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