

Melanocytoma of the Cerebellopontine Angle

Pranay Soni, M.D. Candidate, Faculty Mentor: Lawrence C. Kenyon, M.D., Ph.D.

Pathology Program for Advanced Study, Jefferson Medical College of Thomas Jefferson University, Philadelphia, PA

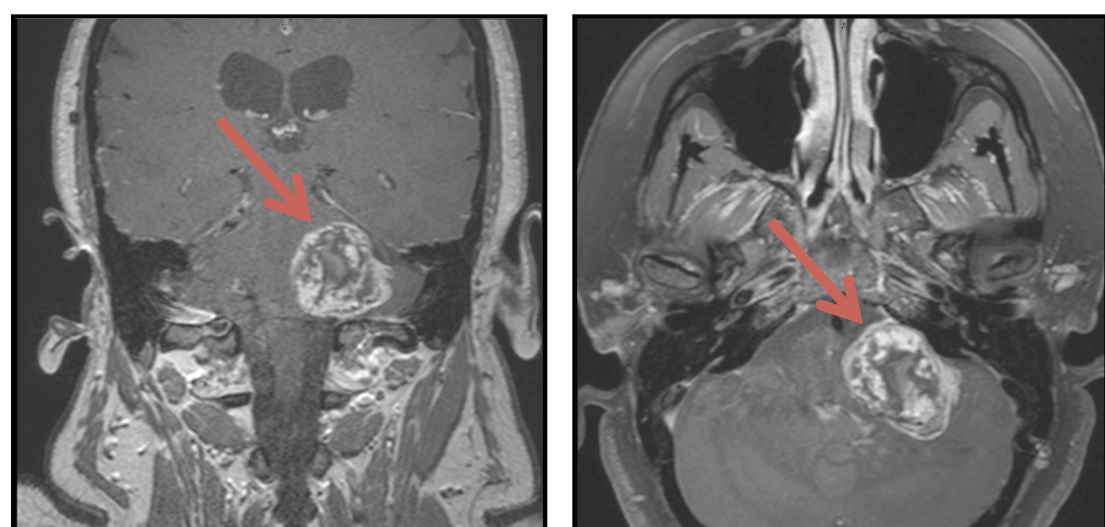
Patient History

Ms. M.G. is a 46-year-old woman with a history of hypertension and prior bilateral laser eye surgery. In 2009, she presented with vertigo, tinnitus, and decreased hearing in the left ear. An MRI scan revealed an enhancing mass in the posterior fossa that was thought to be an acoustic neuroma. In May of 2009, she underwent gamma-knife radiotherapy for the 2.7 cm mass.

In September of 2012, M.G. noted progressive change in her voice quality as well as a swallowing disturbance and left facial spasms. A subsequent MRI showed significant enlargement of the tumor to a maximal size of 3.7 cm with brainstem compression and extension through the jugular foramen. In January of 2013, the patient presented for a neurosurgical consult to discuss treatment options. At this time, a review of systems was also positive for absent hearing in the left ear, vertigo, tinnitus, mild headache, and balance disturbance. On physical exam, the patient was awake, alert, and fully oriented. A cranial nerve exam revealed a hoarse voice, a deviating palate and uvula, absent hearing to finger rubs on the left, mildly decreased sensation of trigeminal nerve in the V3 distribution on the left side, and slight asymmetry in the left trapezius muscle. The remainder of the cranial nerve exam was normal. Motor strength and sensory function were intact in both upper and lower extremities, but she did have a positive Romberg's sign. At this time, the diagnosis of a left posterior fossa jugular foramen schwannoma was made.

In February of 2013, the patient underwent left retrosigmoidal approach to resection of the posterior fossa jugular foramen schwannoma. Because dissection of the tumor from the facial nerve was unsuccessful, a small portion of the tumor was left adherent to the facial nerve superiorly. The patient had a mild left facial palsy postoperatively and was treated with a Decadron taper, but she was subsequently discharged home in a stable condition.

Magnetic Resonance Imaging



Figures 1 & 2. Pre-operative coronal and horizontal T₁-weighted MRI images with contrast show an enhancing mass at the left cerebellopontine angle.

Microscopic Pathology and Immunohistochemical Stains

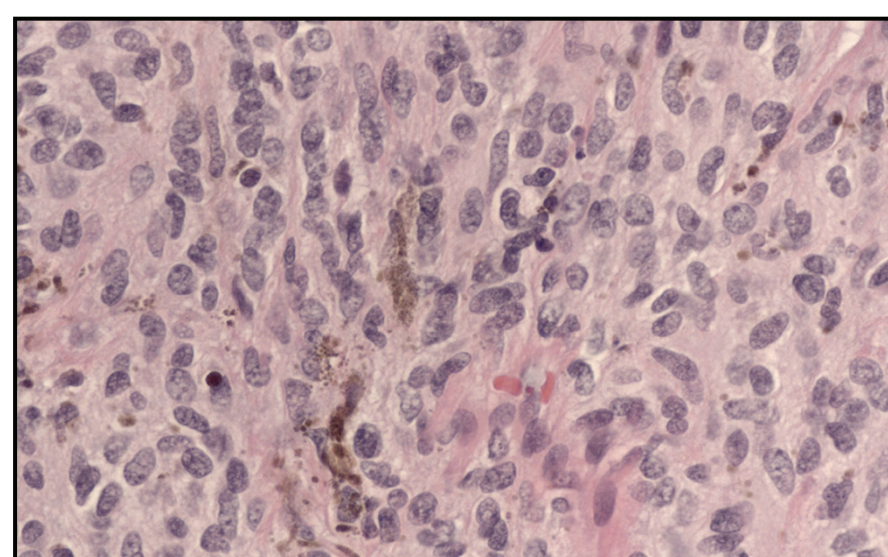


Figure 3. H&E stain of the tumor shows spindle-shaped cells arranged in sheets with cytoplasmic melanin pigment. Nucleoli are small, and there is no nuclear atypia.

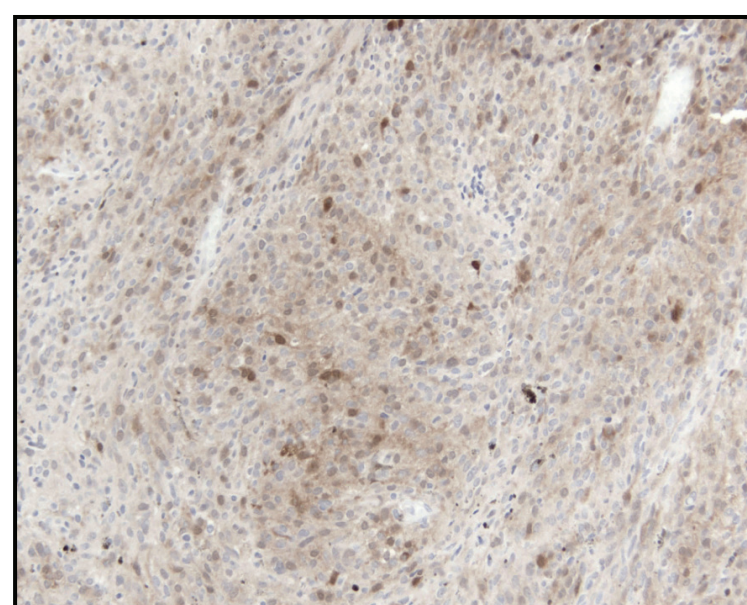


Figure 4. An S-100 stain shows patchy immunoreactivity.

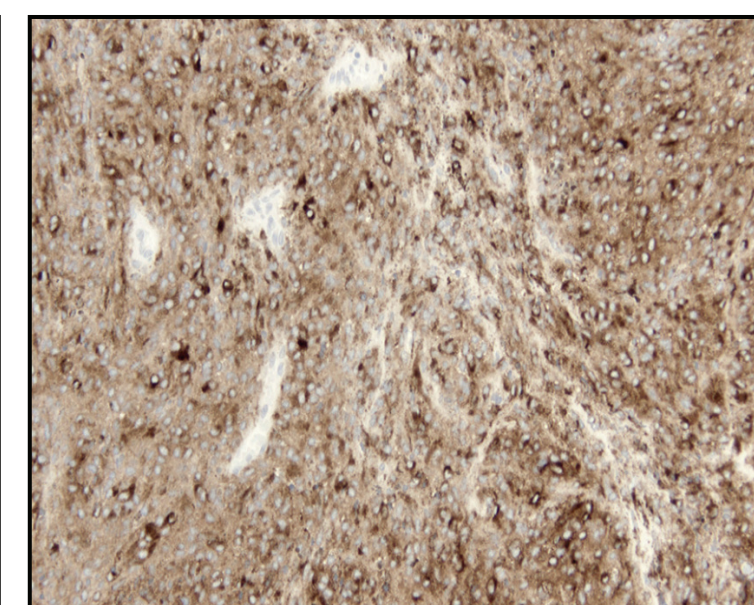


Figure 5. A positively reactive HMB-45 stain.

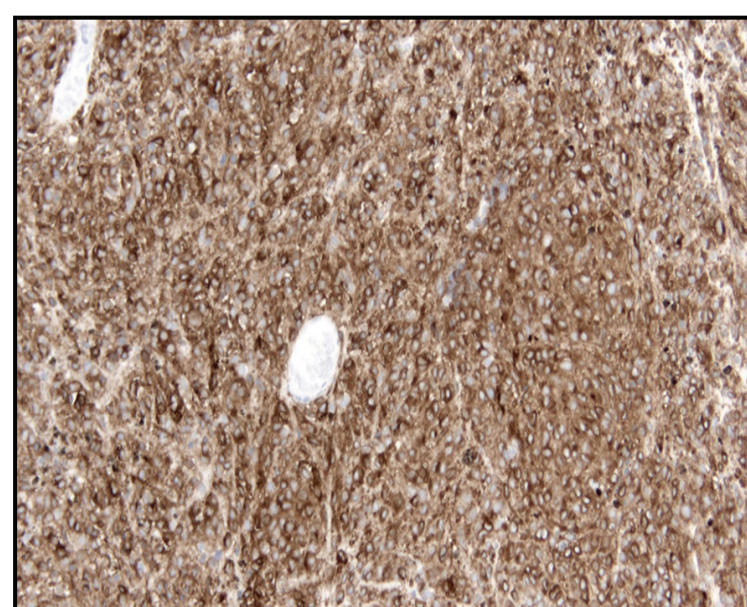


Figure 6. A positively reactive Melan-A stain.

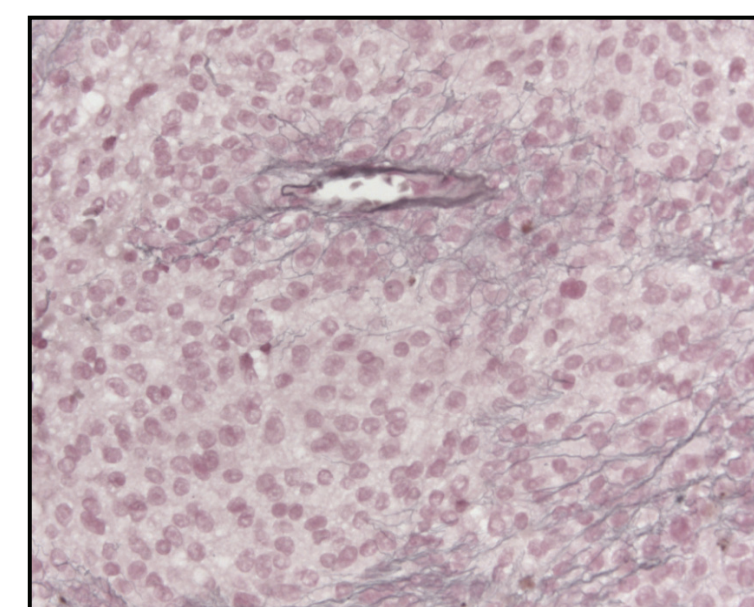


Figure 7. A reticulin stain reveals sparse basal lamina.

Diagnosis

The main differential diagnosis for a pigmented spindle-cell neoplasm includes melanocytoma, primary melanoma of the meninges, melanotic schwannoma, and metastatic melanoma. Immunohistochemical stains are often helpful in distinguishing these tumors.

Human Melanoma Black-45 (HMB-45) is a monoclonal antibody that stains as a marker of melanogenesis, and Melan-A is an antibody that stains as a marker of melanocytic lineage. Strong positive reactivity for these stains along with low atypia and few mitoses on H&E stain narrow the differential diagnosis to melanocytoma and melanotic schwannoma.

The major distinction between these two tumors can be made with the S-100 and reticulin stains. Whereas schwannomas are uniformly strongly reactive for S-100 and thoroughly invested by basal lamina, melanocytomas can be variably reactive to S-100 with individual tumor cells not generally invested by basal lamina. The final diagnosis, therefore, is a meningeal melanocytoma of the cerebellopontine angle.

Discussion

Melanocytes are normal neural crest-derived constituents of the leptomeninges, with the highest density in the ventral portions of the superior spinal cord, brainstem, and inferior surface of the brain.

Melanocytomas are benign, well-differentiated and well-circumscribed tumors with a generally favorable long-term prognosis following surgical excision. There have, however, been case reports of transformation to malignant melanomas, therefore close post-operative follow-up is critical in these patients.

Though melanocytomas are rare tumors, it is important to consider them in the differential diagnosis of certain intracranial neoplasms, particularly at the cerebellopontine angle. Imaging is often inconclusive, and histologic examination is essential in establishing the diagnosis.

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