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Multifocal melanocytoma of the posterior fossa and subcutaneous scalp in the absence of neurocutaneous melanosis

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Abstract

Background: Primary leptomeningeal melanocytic neoplasms of the central nervous system are rare. Multifocal lesions typically occur in the setting of cutaneous melanosis. We present the first report of a posterior fossa melanocytoma and subcutaneous melanocytoma of intermediate grade in the absence of cutaneous melanosis.

Case Description: We present the case of a 22-year-old male with decreased hearing on the right side, ataxia, nausea, vomiting and a scalp mass. Magnetic resonance imaging (MRI) demonstrated occipital and cerebellopontine (CP) angle masses. The patient underwent gross total resection of the scalp mass and subtotal resection of the CP angle mass. Pathologic examination revealed melanocytoma with intermediate grade. The patient underwent stereotactic radiosurgery to the residual CP angle tumor. This case represents, to the author's knowledge, the first report associating a posterior fossa melanocytoma with a subcutaneous melanocytoma of intermediate grade in the absence of cutaneous melanosis.

Conclusion: This case introduces the first report of a new variant of multifocal melanocytoma which is not confined to the central nervous system.

Key Words: Multifocal melanocytoma meningeal melanosis, radiosurgery, subcutaneous melanocytoma

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INTRODUCTION

Primary leptomeningeal melanocytic neoplasms of the central nervous system (CNS) include a spectrum of entities, which include diffuse lesions (leptomeningeal melanocytosis and melanomatosis), as well as circumscribed lesions (meningeal melanocytoma and primary malignant melanoma). Meningeal melanocytomas are derived from the melanocytes of the leptomeninges in the CNS, and were first described as benign focal lesions by Lima and Tio.[11] They usually present as solitary lesions in the CNS and can occur in association with cutaneous melanosis. Meningeal melanocytomas have an estimated incidence of 1 per 10 million with a slight female predominance.[12,21] They are most commonly found in the posterior fossa and spine.[12,24] They are mostly intradural extra-axial lesions and usually grow in a solitary, expansile rather than infiltrative fashion. Despite their benign nature, they are known to show a tendency for local recurrence. Leptomeningeal spread and malignant transformation to melanoma has been

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described months to years after diagnosis and treatment of a melanocytoma.[4,7-9,15,19,22,23]

Multifocal meningeal melanocytomas at the time of diagnosis have been reported in five cases in the published literature. All described cases were contained to the CNS in the form of posterior fossa and spinal lesions. We present the case of a young male with multifocal disease in the form of a cerebellopontine intradural meningeal melanocytoma as well as a subcutaneous occipital melanocytoma. Intraoperative leptomeningeal hyperpigmentation of the surrounding dura was noted but did not meet the criteria for melanocytosis. The patient did not have cutaneous anomalies. This is the first case report of a multifocal CNS meningeal melanocytoma with a subgaleal extracranial melanocytoma described without a context of cutaneous melanosis.

CASE REPORT

A 22-year-old male presented to our institution's emergency room with decreased hearing on the right side, ataxia, nausea, and vomiting. On physical examination, an occipital painless subcutaneous mass was noted and audiogram revealed non-serviceable hearing on the right side. A magnetic resonance image (MRI) of brain with contrast demonstrated two separate lesions with similar signal characteristics. A subcutaneous occipital mass measuring 4.8 × 2.9 × 4.1 cm was noted as well as an intradural extraxial 2.6 × 2 × 5.2 cm CP angle mass. There was extension of the CP angle mass into the enlarged jugular foramen and invasion of the internal jugular vein. Both lesions were T1 hyperintense enhancing lesions with a mixed iso and hypointense signal on T2 [Figures 1 and 2]. A cerebral angiogram confirmed obstruction of the right sigmoid sinus and jugular vein at the level of the jugular foramen with recanalization distally [Figure 3]. Imaging of the spine by MRI did not reveal other lesions.

Surgical resection of the two lesions was performed at our institution. The posterior fossa mass was approached through a right translabyrinthine approach and the subcutaneous lesion was resected in the same setting with a separate incision. Electrophysiologic monitoring with cranial nerve monitoring was performed. The extracranial lesion was resected first and did not show any continuity to the intracranial lesion.

A posterior petrosal translabyrinthine approach was utilized for the intracranial lesion which permitted presigmoid access as well as exposure of the sigmoid sinus and jugular bulb. The dura showed extensive patchy areas of melanocytic coloration over the entirety of the surgical exposure. The intradural lesion was readily visible with invasion of the sigmoid sinus. The sigmoid sinus was opened and found to be occluded with tumor. The sinus was ligated on either side of the tumor invasion and sacrificed. The mass was debulked using a combination of ultrasonic aspiration and suction. The capsule was dissected-off of the lateral cerebellum, brainstem, and cranial nerve 7 despite its adherence. Dissection of the tumor to the lower cranial nerves could not be completely achieved and a subtotal resection was performed. Closure was performed with a dural onlay

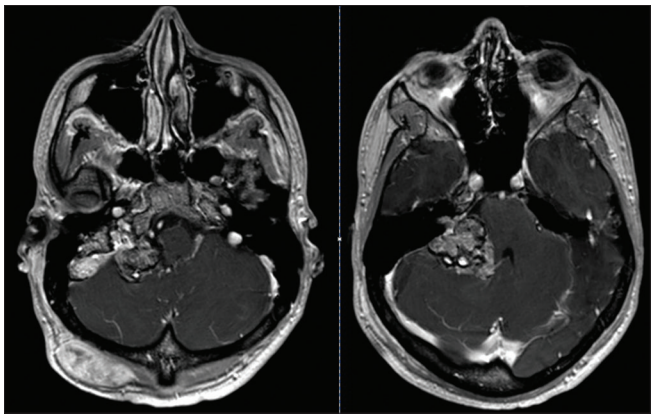

Figure 1: Preoperative contrast enhanced magnetic resonance imaging

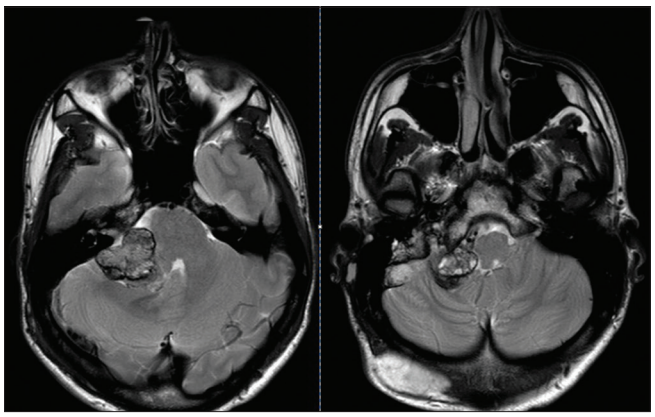

Figure 2: Preoperative T2 magnetic resonance imaging

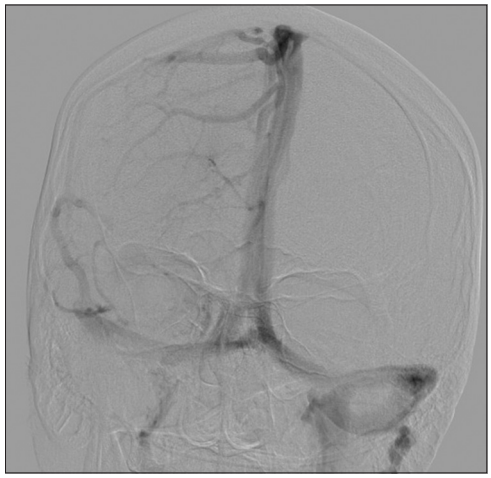

Figure 3: Preoperative angiogram

followed with the use of abdominal fat graft after waxing the surrounding bony structures. A lumbar drain was used postoperatively for 5 days. The patient postoperatively was noted to have nystagmus and a facial nerve function consistent with a House–Brackmann grade 2 which recovered within a month. The patient was discharged home on postoperative day 6. The patient was treated with stereotactic radiosurgery at 6 months (22 Gray in three fractions) and has been stable on MRI imaging for 12 months [Figure 4].

Pathology

Gross and microscopic examination of both of the lesions demonstrated similar histomorphological features and immunohistochemical profile confirming the melanocytic origin. The tumor cells were arranged in a nested pattern and showed mild pleomorphism, although rare mitotic figures were noted. Marked intracellular and extracellular melanin deposition was noted [Figure 5]. No evidence of high grade features were noted and both the lesions were classified as melanocytoma with intermediated grade.[3]

DISCUSSION

CNS melanocytomas are tumors that arise from leptomeningeal melanocytes which are embryologically derived from neural crest cells. They are classified as benign circumscribed lesions within the spectrum of primary leptomeningeal melanocytic neoplasms. They occur across all age groups with a slight predilection for females. In the CNS, they are usually intradural extra-axial lesions that may occur intracranially as well as in the spine. The two most common intracranial locations are the posterior fossa and Meckel's cave. Spinal melanocytomas typically occur in the cervical spine followed by the thoracic spine.[24] Meningeal melanocytomas outside the spectrum of cutaneous melanosis are usually diagnosed because of their compressive effect on the surrounding nervous structures (cranial nerve palsies, myeloradiculopathy, headaches, hemorrhage, or seizures). In cases of cutaneous involvement in the form of cutaneous melanosis, naevus of Ota or Ito, asymptomatic lesions can be diagnosed examining their known relationship.[12]

Meningeal melanocytomas are usually benign lesions that are encapsulated and do not present with direct invasion of the surrounding CNS. They are classically described by MRI imaging as hyperintense on T1-weighted images, hypointense on T2-weighted images, hyperintense on fluid-attenuated inversion recovery (FLAIR) images, and enhance homogeneously with gadolinium, as seen in this case [Figures 1 and 2].[12] Macroscopically, they appear black or brown due to their melanin content. Microscopically, abnormal mitoses and atypical cytology are usually absent. Melanocytoma cells are distinguished from melanocytic meningioma on electron microscopy from their lack of developed desmosomes and interdigitating cytoplasmic processes.

The benign nature of melanocytomas has been questioned based on the description of an intermediate grade. An intermediate grade is defined microscopically with the presence of mitoses (1–3 mitoses per 10 HPFs and MIB-1 LI ranging 1–4%) and/or microscopic CNS invasion.[2] The clinical behavior of an intermediate lesion is uncertain, as observed in this case.

Treatment strategies in meningeal melanocytoma should aim for a complete resection of the lesion when feasible. If a subtotal resection is accomplished due to CNS invasion and adherence to critical structures, observation or radiosurgery should be offered. Patients should be followed closely with serial imaging of the entire CNS. It is now suggested that melanocytomas progress to melanoma within 3 months to 12 years.[8,15,19,22,23] Such patients experience a very poor prognosis despite the use of chemotherapy and radiation.

Multifocal melanocytomas of the CNS was first described as an entity in 2009 by Ali *et al*. when reporting a case

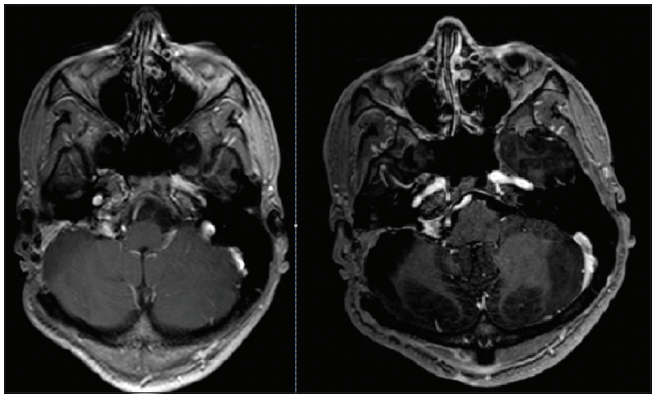

Figure 4: Postoperative contrast enhanced magnetic resonance imaging

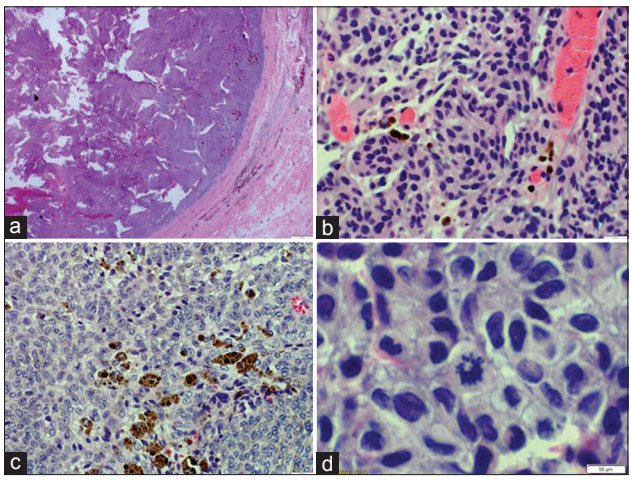

Figure 5: Pathology slides, (a, b) from the scalp lesion and (c, d) from the cerebellopontine angle lesion

Table 1: Review of published cases of multifocal melanocytoma involving the CNS

Author	Year published	Sex/ age	Presenting Symptoms	Location	Treatment(s)	Outcome
Present study		M/22	Hearing loss, nausea/ vomiting	CP* angle and scalp	GTR** of scalp lesion, subtotal resection of intracranial lesion plus SRS†	No recurrence or progression at 9 months (present)
Ali *et al*.[1]	2009	M/31	Hearing loss, headache	Bilateral CP* angle lesions and thoracic spine lesion	VP shunt and resection of spinal tumor	Death secondary to brain stem compression several weeks postoperative
Franken *et al*.[7]	2009	M/26	Headache, nausea/ vomiting	Two posterior fossa lesions and spinal drop metastases	GTR** of cranial lesions and craniospinal radiation	No recurrence or progression at 18 months
Merciadri *et al*.[13]	2011	M/68	Back pain and lower extremity weakness	Conus medullaris and three posterior fossa lesions	GTR** and SRS† to cranial lesions	No recurrence or progression at 12 months
Reddy *et al*.[18]	2012	M/43	Neck Pain	Two cervical spine lesions	GTR**	No recurrence at 6 months
Foit *et al*.[6]	2013	M/43	Head, neck, and arm pain	One cervical and one thoracic lesion	Subtotal resection of both lesions plus stereotactic EBR	No recurrence or progression at 18 months

*CPA: Cerebellopontine angle, **GTR: Gross total resection, †SRS: Stereotactic radiosurgery

of bilateral CP angle melanocytomas with a thoracic melanocytoma at the time of diagnosis. A total of five cases have been described in the combination of a posterior fossa mass with a spinal lesion or multiple spine lesions.[1,6,13,18,20] The resected lesions were all benign on pathology except in the case described by Foit *et al*., where one of the two lesions had an intermediate grade [Table 1].

This entity is thought to be different from cases that have demonstrated leptomeningeal spread after total or subtotal resections of a melanocytoma.[4,7-9] Some of the melanomas thought to be derived from degeneration of a melanocytoma have demonstrated leptomeningeal spread.[8,15,22,23] Koenigsmann *et al*. have described the only case that involves hematogenous spread with the presence of liver metastasis 8 years after resection of a melanocytoma. Their case also had leptomeningeal spread within the CNS at the time of the diagnosis of liver metastasis.[9] Our case report is the first description of a CNS melanocytoma combined with a subcutaneous melanocytoma without malignant degeneration. Furthermore, we believe this represents multifocal disease observing the lack of continuity between the two lesions and their exact similar appearance and grading on pathology.

Case reports of melanocytomas have sometimes also commented on the presence of diffuse leptomeningeal hyperpigmentation of the surrounding dura, as seen in our case.[1,5-7] These are not considered to be the cases of melanocytosis, which usually occurs with underlying cutaneous melanosis. Of all the case reports describing multifocal disease, Ali *et al*. and Foit *et al*. also mentioned the presence of surrounding leptomeningeal hyperpigmentation.[1,6] The other authors did not specifically comment on the status of the surrounding dura. The clinical significance of underlying leptomeningeal hyperpigmentation in terms of the formation of a de novo melanocytoma, recurrence, leptomeningeal spread, or degeneration to melanoma is not known. Table 1 compares other cases of multifocal meningeal melanocytomas with our case.

Melanocytomas are known to recur locally even after complete resection. Five year overall survival rates are 83% after complete resection and 40% in incomplete resection without radiation therapy.[14,16] Radiation therapy increases survival in subtotal resection to an overall survival of 91–92% at 5 years.[10,17] Our case was treated with radiosurgery to the posterior fossa residual lesion, and serial imaging showed stability at 15 months.

This is the first case report describing multifocal disease with an intradural lesion and a subcutaneous mass at the time of diagnosis. Their relationship is not understood but strongly suggests multifocal disease.

Our findings support the concept of multifocal melanocytomas, which may not be restricted within the CNS, as previously described.

CONCLUSION

We introduce a new variant of multifocal melanocytoma in a patient which was not confined to the CNS. This is the first report associating a posterior fossa melanocytoma with a subcutaneous melanocytoma of intermediate grade in the absence of cutaneous melanosis. Its clinical behavior appears to be similar to other cases of multifocal disease described in the literature.

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Conflicts of interest
There are no conflicts of interest.

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