Intracranial Meningeal Melanocytoma mimicking Meningioma: A Case Report

Primary melanotic neoplasms involving the central nervous system are extremely rare. Radiographically, it may be indistinguishable from intracranial meningioma. Here, we present a 52-years-old male, histopathologically-diagnosed as having posterior fossa meningeal melanocytoma with extension into the upper cervical area, who underwent successful radical surgery and postoperative radiotherapy. Details of the intraoperative and histopathological findings, imaging characteristics and clinical features are also described in correlation with literature research.

**Keywords:** Meningeal melanocytoma, MRI, Radiotherapy, Tumor excision

Meningeal melanocytoma is a melanotic neoplasm of benign nature that arises from the melanocytes of the meninges and can occur anywhere along the meninges of the neural axis. Primary intracranial melanotic neoplasms are rare and usually occur as malignant melanoma.

However, meningeal melanocytoma is considered as a benign type of primary melanotic neoplasm and is said to be less common than the malignant types. Using radiographic modalities even with MR Imaging, preoperative differentiation of meningeal melanocytoma from the malignant melanoma and meningioma is difficult. Histopathological examination is essential to confirm the diagnosis. The prognosis of the tumor is not always favorable with occasional local recurrence. Total resection is the treatment of choice in most of the cases. Local recurrence is best irradiated by gamma knife radiosurgery.

We report a case of intracranial meningeal melanocytoma arising in the posterior cranial fossa extending into the upper cervical canal. Its intraoperative and histopathological findings, imaging characteristics and clinical features are described in detail.

**Case Report**

A 52-year-old old man, recently diagnosed with diabetes mellitus, who had generally been in good health, visited our hospital with a three-month history of occipital...
headache on and off, occasional vomiting and dizziness, particularly on standing position. On neurological examination, the patient was found to have nuchal rigidity and increased deep tendon reflexes with ataxic gait. The cerebellar signs, namely past pointing, dysmetria, were profound on the right side.

There were no melanotic pigmentations of the skin, mucous membrane or the eyes. On radiological imaging, non-contrasted T1-weighted MR images demonstrated iso- to slightly hyperintense signal lesion in the posterior fossa extending into the upper cervical canal up to C2 level (Figure 1A and 1B) and the same lesion was homogenously enhancing on postcontrast images (A) Precontrast sagittal (B) Precontrast coronal (C) Postcontrast sagittal (D) Postcontrast coronal sequences.

Operative findings revealed a large black-colored, soft, gelatinous, well-delineated, moderately vascular, extraaxial tumor within the dura mater, occupying the cisterna magna extending up to floor of fourth ventricle rostrally and downwards to C1 lamina level caudally. There was a small attachment to the right lateral rim of Foramen of Magnum. The cerebellum was pushed superiorly and cord was pushed anteriorly.

The postoperative course was uneventful. On one-month follow-up, there were still some minimal residual cerebellar signs. The patient was then subjected to postoperative radiotherapy at Bhaktapur Cancer Hospital. On two-month follow-up after radiotherapy, the patient was neurologically intact and in high spirit.

Histopathological examination (HPE) revealed tumor cells in diffuse sheets. These tumor cells are round to oval, intermediate sized, with ill defined cytoplasmic borders, vesicular nucleus with small conspicuous nucleoli. Several foci with melanin pigmentation within the tumor cell is visualized. Mitosis and necrosis is absent. (Figure 6 A and B).
Discussion

Meningeal melanocytoma is also misnamed as pigmented meningioma or melanotic meningioma because of the light microscopy characteristics - which is almost identical to those of meningioma. Electron microscopy demonstration of the melanocytic ultrastructure of these tumors help coin the term “meningeal melanocytoma” based on the melanocytic origin. Melanocytes are derived from the neural crest during early embryonic development and occur in normal leptomeninges. Scattered melanocytes are most frequently found in the recesses of the sulci around the base of the brain and in the upper cervical spinal cord. Meningeal melanocytomas probably arise from these cells and mainly occur in the posterior fossa and the spinal canal.

The differential diagnosis of a primary pigmented tumor of the leptomeninges includes melanoma, melanoblastosis, melanotic schwannoma, pigmented primitive neuroectodermal tumor, and meningeal melanocytoma. It must be noted that the World Health Organization’s classification of CNS tumors does not recognize a separate pigmented variant of meningioma and, instead, classifies meningeal melanocytoma as a primary melanocytic lesion based on the early work of Masson. They usually present with symptoms during their fifth or sixth decade, but can present anywhere from 9 to 71 years. The females are affected more than the men (male-to-female ratio, 2:1). In our case, the patient was a 52-year-old male. Duration of symptoms can range from 4 weeks to 14 years. In our case, the patient presented with three-month history of symptoms prior to seeking medical advice. The radiological appearance of meningeal melanocytoma is well described. These tumors are usually iso- to hyperdense and enhance homogeneously on CT scans. These tumors are usually found in the posterior fossa and the cerebello-pontine angle and tend to occur as solitary lesions. Unlike the MRI appearance of most other CNS tumors, melanocytomas are usually isointense on T1-weighted images and hypointense on T2-weighted images, and enhance homogeneously after addition of gadolinium. It is believed that the degree of melanization attributes to the imaging finding of these tumors. On MRI, the signal intensity of these tumors is strongly related to the amount of melanin pigment. More melanin means more shortening of T1 and T2 relaxation times. It is difficult to differentiate meningeal melanocytoma from malignant melanoma solely on the basis of the MRI findings. On the other hand, meningiomas are often difficult to differentiate from these tumors owing to their capability of displaying similar imaging features. Because the imaging appearance of these tumors may be remarkably similar to...
A)  B)

**Figure 4: HPE showing the tumor in diffuse sheets. Note the cytoplasm pigmented with brown granules (A) low-power microscopic view (B) high-power microscopic view.**

those of other melanin-containing tumors, melanocytomas are ultimately differentiated by microscopy. In this present case, the radiological diagnosis of meningioma was made due to the iso- to slightly hyperintensity on T1, mixed intensity in the T2-weighted sequences and homogenous enhancement on Gadolinium contrasted images.

Microscopically, these tumors are hypercellular and composed of uniform spindle or fusiform cells arranged in whorls, sheets, bundles, or nests, often surrounded by a fine network of reticulin. Individual cells contain prominent nucleoli, cytoplasmic melanin, and, rarely, mitosis. Other possible features include intratumoral hemorrhage, increased vascularity, calcification, and a lack of necrosis or brain invasion. As in some cases, evidence of intratumoral hemorrhage may present. In contrast, malignant melanomas tend to be more densely cellular and greater degrees of pleomorphism and nuclear atypia are generally present. Because meningeal melanocytomas are histologically benign tumors, the goal of treatment is complete surgical removal. However, despite gross tumor removal recurrence may occur. Malignant transformation has also been described.

Generally, radiotherapy is reserved for symptomatic residual, progressive, or recurrent tumors that are not amenable to further resection and in this regard gamma knife is probably rewarding. In our case, the patient was subjected to postoperative radiotherapy due to the unpredictability of this tumor even with total resection. The prognosis is highly variable, but overall, it is significantly better than the median survival time of 2.5 months predicted for patients harboring a cerebral metastatic malignant melanoma. Postoperative survival time in patients harboring intracranial meningeal melanocytomas has ranged from 1 to 28 years. In our case, the patient is still being followed up periodically even after 3 months of surgery.

**References**

11. Ray BS, Foot NC: Primary melanotic tumors of the meninges: resemblance to meningiomas. Report of two cases in which operation was performed. Arch Neurol Psychiatry 44: 104–117, 1940