Nepal Journal of Neuroscience 1: 113-16, 2011

Intracranial Meningeal Melanocytoma mimicking Meningioma: A Case Report

Pradip Prasad Subedi, MBBS
Neurosurgery Unit, Department of Surgery
Tribhuvan University Teaching Hospital
Maharajgunj, Kathmandu, Nepal

Gopal Sedain, MS
Neurosurgery Unit, Department of Surgery
Tribhuvan University Teaching Hospital
Maharajgunj, Kathmandu, Nepal

Mohan Raj Sharma, MS
Neurosurgery Unit, Department of Surgery
Tribhuvan University Teaching Hospital
Maharajgunj, Kathmandu, Nepal

Sushil Krishna Shilpakar, MS
Neurosurgery Unit, Department of Surgery
Tribhuvan University Teaching Hospital
Maharajgunj, Kathmandu, Nepal

Address for correspondence:
Dr. Sushil Krishna Shilpakar
Professor, Head of Neurosurgery
Neurosurgical Unit, Department of Surgery
Tribhuvan University Teaching Hospital
Kathmandu, Nepal
E-mail: sshilpak2001@yahoo.com

Received, 23 November, 2010
Accepted, 11 December, 2010

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[1, 2]. 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

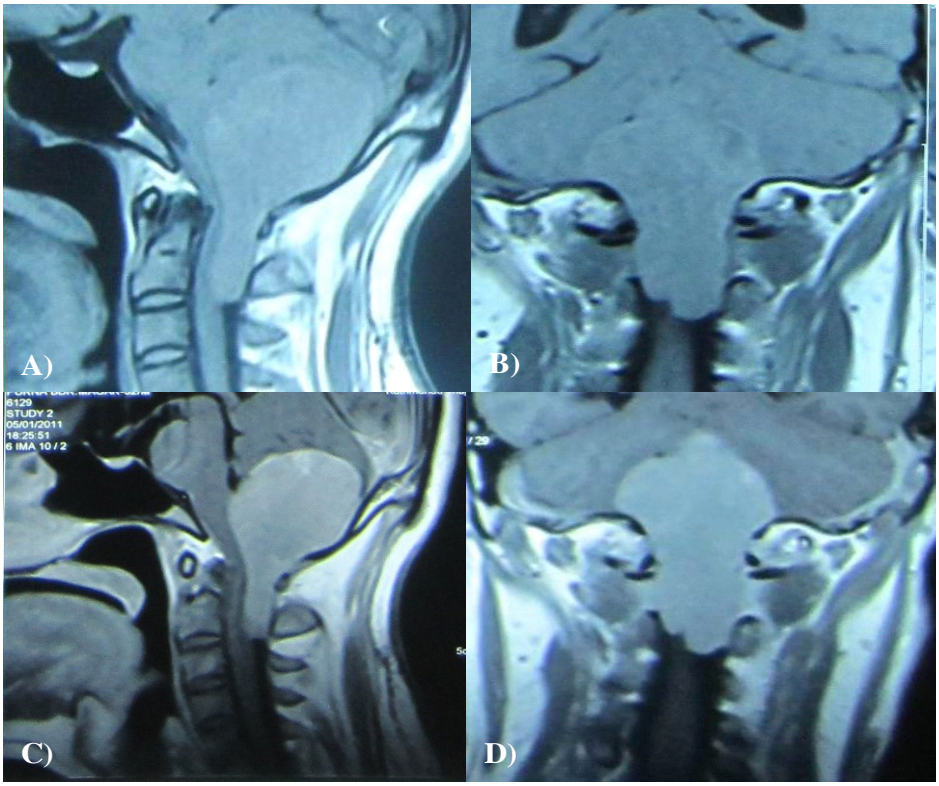

Figure 1: Non-contrasted T1-weighted MR images demonstrating iso- to slightly hyperintense signal lesion in the posterior fossa extending into the upper cervical canal up to C2 level and the same lesion showing homogenously enhancement on postcontrast images (A) Precontrast sagittal (B) Precontrast coronal (C) Postcontrast sagittal (D) Postcontrast coronal sequences.

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 T_1-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 the postcontrast images **(Figure 1C and 1D)**. T2-weighted MR images revealed heterogeneous hyperintensity in the lesion. **(Figure 2)**. Due to its homogeneity of signal intensity, radiological diagnosis of posterior fossa meningioma was made preoperatively. On January 14, 2011 (Poush 30, 2067 B.S.), the patient underwent standard suboccipital craniectomy with C1, C2 laminectomies under general anesthesia for total excision of the tumor **(Figure 3)**.

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 C_2 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,intermidiate sized,with ill defined cytoplasmic borders,vesicular nucleus with small conspicuous nucleoli.Several foci with malanin pigmentation within the tumor cell is visualized.Mitosis and necrosis is absent. **(Figure 6 A and B)**.

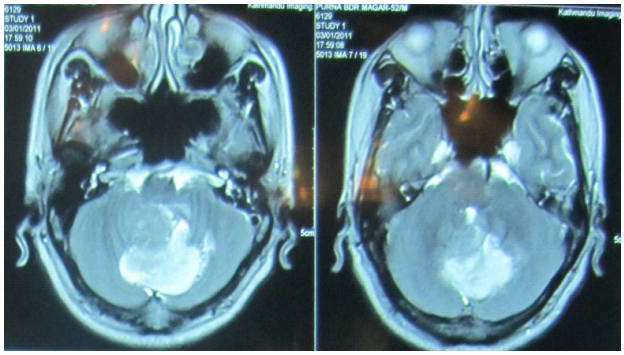

Figure 2: T2-weighted MR images showing the heterogeneous hyperintense lesion.

must be noted that the World Health Organization's classification of CNS tumors[6] 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[7]. 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

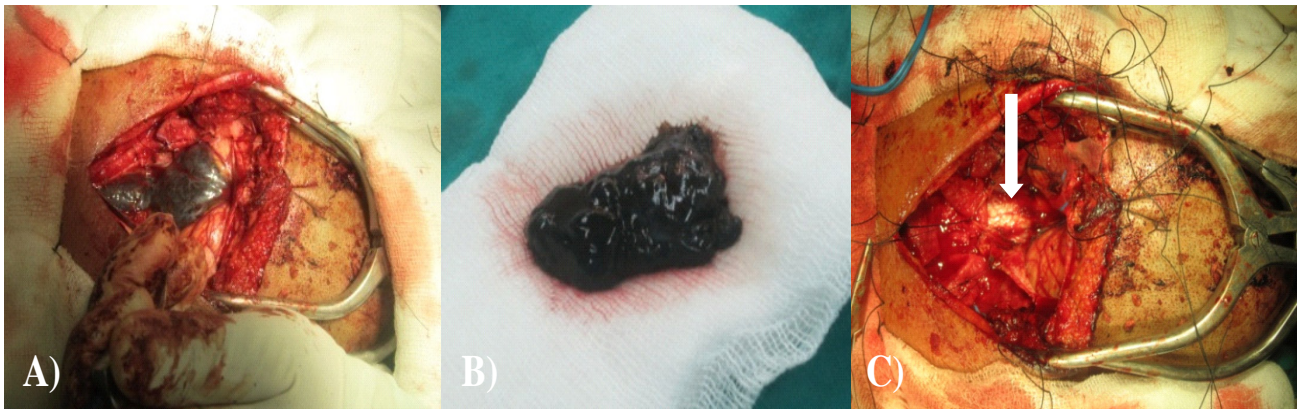

Figure 3: Intraoperative images of the tumor (A) gross appearance showing the black-colored tumor within the dura after suboccipital craniectomy and laminectomy of C1, 2. (B) the tumor specimen after excision, note the gelatinous nature, (C) complete excision of tumor and exposure of dorsal surface of medulla as indicated by white arrow.

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[3]. Electron microscopy demonstration of the melanocytic ultrastructure of these tumors help coin the term "meningeal melanocytoma" based on the melanocytic origin[4]. 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[5]. 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 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[8]. It is believed that the degree of melanization attributes to the imaging finding of these tumors[9]. 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

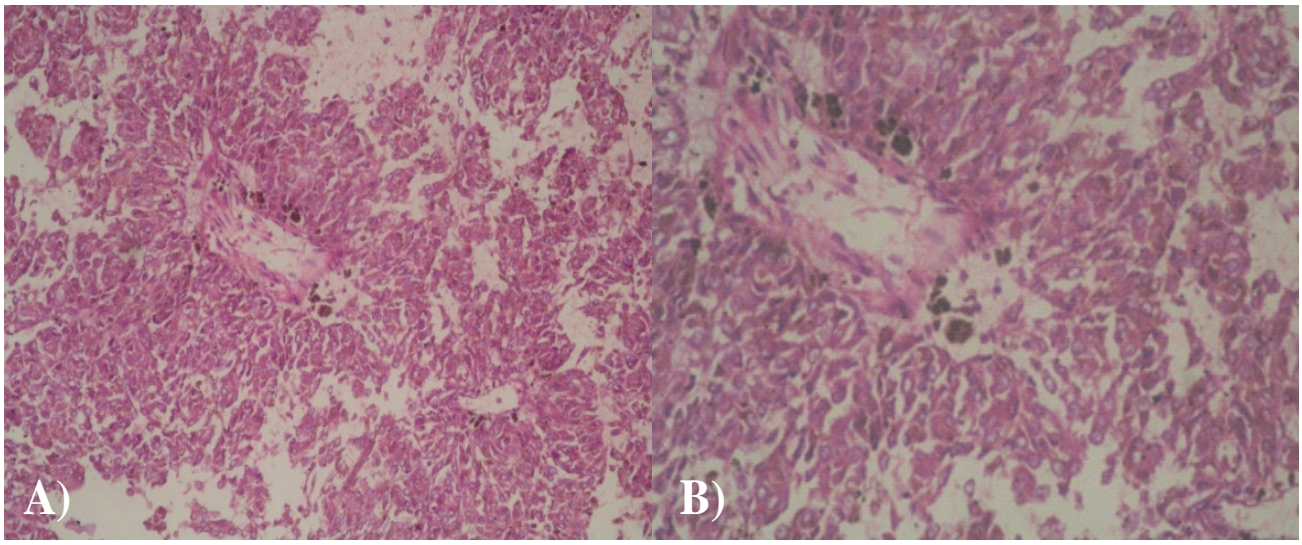

Figure 4: HPE showing the tumor in diffuse sheets. Note the cytoplasm pigmented with brown granules (A) low-power microscopic view (B) high-power microscopic view.

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[10]. Because meningeal melanocytomas are histologically benign tumors, the goal of treatment is complete surgical removal. However, despite gross tumor removal recurrence may occur[11]. Malignant transformation has also been described[12].

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[13, 14]. Postoperative survival time in patients harboring intracranial meningeal melanocytomas has ranged from 1 to 28 years[14]. In our case, the patient is still being followed up periodically even after 3 months of surgery.

References

1. Brat DJ, Giannini C, Scheithauer BW, et al: Primary melanocytic neoplasms of the central nervous system. **Am J Surg Pathol 23:**745–754, 1999
2. Clarke DB, Leblanc R, Bertrand G, et al: Meningeal melanocytoma. Report of a case and a historical comparison. **J Neurosurg 88:**116–121, 1998
3. Goldgeier MH, Klein LE, Klein-Angere S, Moellmann G, Nordlund JJ: The distribution of melanocytes in the leptomeninges of the human brain. **J Invest Dermatol 82:** 235-238, 1984
4. Keegan HR, Mullan S: Pigmented meningiomas: an unusual variant. Report of a case with review of the literature. **J Neurosurg 19:** 696-698, 1962
5. Kleihues P, Burger PC, Scheithauer BW: The new WHO classification of brain tumors. **Brain Pathol 3:** 255–268, 1993
6. Limas C, Tio FO: Meningeal melanocytoma ("melanotic meningioma"). Its melanocytic origin

as revealed by electron microscopy. **Cancer 30:** 1286-1294, 1972

7. Lach B, Russell N, Benoit B, et al: Cellular blue nevus ("melanocytoma") of the spinal meninges:
8. Masson P: Les neavi pigmentaires, tumeurs nerveuses. **Ann Anat Pathol 3:** 417–453, 1926
9. Matsumoto S, Kang Y, Sato S, et al: Spinal meningeal melanocytoma presenting with superficial siderosis of the central nervous system. Case report and review of the literature. **J Neurosurg 88:** 890–894, 1998
10. O'Brien TF, Moran M, Miller JH, et al: Meningeal melanocytoma. An uncommon diagnostic pitfall in surgical neuropathology. **Arch Pathol Lab Med 119:** 542–546, 1995
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
12. Retsas S, Gershuny AR: Central nervous system involvement in malignant melanoma. **Cancer 61:** 1926–1934, 1988
13. Russell DS, Rubinstein LJ: Pathology of Tumors of the Nervous System. Baltimore, Williams & Wilkins, 1989, pp 794-797
14. World Health Organization: Classification of tumors, in Kleihues P, Cavanee WK (eds): Pathology and Genetics. Tumours of the Nervous System. Lyon, IARC Press, 2000, pp 193-195