INTRODUCTION

Primary intracranial melanoma accounts for approximately 1% of all cases of melanoma. It usually presents in the same way as other intracranial masses. Histologically, tumours of melanocytic origin range from benign melanocytomas to malignant melanomas. It is generally difficult to diagnose this rare entity on clinical grounds alone; radiological appearance with biopsy usually confirms the diagnosis. Aggressive surgical resection is recommended, as it has overall good prognosis when compared with the common lesions of that particular location.

CASE REPORT

A 28 years old woman presented with left sided headache, numbness of the left side of face and diplopia. On examination, there was absent sensations in all three territories of 5th nerve, absent corneal reflex and deceased masseter muscle mass on the left side. Left lateral rectus muscle was also paralyzed.

MRI showed a dumbbell lesion in left temporal lobe, abutting ipsilateral cavernous sinus and extending posteriorly into the basal cisterns causing pressure on brain stem at left side (Figure 1). On T1-weighted image, it was hyper intense; on T2-weighted and FLAIR images, it was mixed hypo as well as hyper intense. Due to its typical location and extension, it was initially considered as a 5th nerve neuroma, but its typical hyperintense signals on T1WI, favoured the melanoma. She underwent left temporal craniotomy. Inside the lesion, dark blood contained material was found. Since it was not infiltrating into brain parenchyma and could be removed easily so gross total removal was achieved. The immunohistochemical profile was positive for S-100, HMB-45, and Melan-A (Figure 2).

Her metastatic evaluation included clinical examination for intraocular and dermal primary melanoma and was negative. Patient remained symptom-free till the last follow-up that was four years postoperatively.

ABSTRACT

Primary intracranial melanoma is an extremely rare lesion of the brain. A 28 years old lady, without any co-morbid conditions, presented with headache and left side facial numbness. MRI of brain showed a mass lesion in left middle cranial fossa extending into posterior fossa, with signals characteristics of melanoma. Histology and immunohistochemistry confirmed the diagnosis after surgical excision of that lesion. Patient remained symptom-free till the last follow-up that was four years postoperatively.

Key words: Intracranial lesions, Malignant melanoma, Prognosis.
found negative. Skeletal scintigraphy ruled out any bony metastasis, whereas CT scans of the chest, abdomen and pelvis were found normal. MRI of spine was also done to see secondaries and was normal. No adjuvant therapy was given. The patient is symptoms and recurrence free for 4 years.

**DISCUSSION**

Primary melanoma of the central nervous system (CNS) arises from melanocytes, which are derived from precursor cells (melanoblast) originating in the neural crest. In the craniospinal axis, melanocytes are predominantly located in the leptomeninges at the anterior and lateral surfaces of the spinal cord, the brain stem and the base of the brain. This distribution of melanocytes closely correlates with the reported distribution of these rare tumours. Females are affected twice as common as males. Clinical manifestations vary widely.

Primary melanocytic tumours of the CNS can be classified into diffuse or localized disease. Diffuse disease, also referred to as leptomeningeal melanosis, represents the diffuse infiltration of the subarachnoid space of the brain and spinal cord by melanocytes. Localized disease suggests the melanocytes are limited to a circumscribed part of the CNS. They range from benign melanocytomas to malignant melanomas. Diffuse leptomeningeal melanomas mainly appear in children and may be part of neurocutaneous melanosis complex or phakomas. These may present with features of raised intracranial pressure, cranial nerve palsies and meningism. But focal melanomas, as in this case, presents as leptomeningeal or dural based neoplasms and are more common in adults. Cerebellopontine angle tumours typically manifest clinically with audiovestibular symptoms and facial nerve palsy. Although rare, it may be considered in differential diagnosis of CP angle tumours.

These tumours have distinctive appearance on MRI scans. In the typical melanotic melanoma, the melanin has a paramagnetic effect that derives from the presence of stable organic free radicals inside. The unpaired electrons of these free radicals interact with the water protons, resulting in shortening of both T1 and T2 relaxation times and producing hyper intensity on T1 and hypo intensity on T2 weighted images. Intratumoural haemorrhage produces a heterogeneous signal on T1 and T2 images. Finally, amelanotic melanoma and melanoma without a haemorrhagic component appear iso to hypointense on T1 and moderately hyper intense on T2 images. In this case, the tumour was hyper intense on T1 and mixed to hypo intense on T2 weighted images.

HMB-45 is an antibody with higher specificity for melanocytic tumours. In the literature, 86 – 97% of melanocytic tumours were positive for HMB-45 antigen, however, S-100 is found in almost all melanocytes but it is also present in other neural crest derived tumours. Thus, it has high sensitivity and low specificity. Melan A is a differentiation antigen expressed in all melanocytes, and is reported to be positive in most cases of melanoma. In this case, the tumour was typically positive for Melan-A, HMB-45 and S-100.

Primary melanoma of CNS must be differentiated from the metastasis to brain. Primary extra cranial tumour can be found in as many as 90% of patients while it remains unknown in rest of the 10% of patients. In this patient, the absence of melanoma outside the CNS and the presence of single intracranial lesion support the diagnosis of primary CNS melanoma, moreover, immunohistochemistry does not help to distinguish between primary tumour and secondary metastasis.

Complete surgical excision in focal intracranial lesion can be curative. Radiation is considered in patients in whom either surgery is not possible or those cases where complete excision cannot be attempted. The prognosis is quite good in cases of totally excised melanomas. The mean survival depends upon the presence of neuroological deficit and signs of raised intracranial pressure at the time of presentation. It also varies with the extent of tumour removal and use of adjuvant therapies. Postoperative survival time has ranged from 1 to 28 years. This patient underwent complete excision of tumour without any adjuvant therapy and remains disease-free for 4 years and still alive and healthy with recent MRI showing no pathology. CNS melanoma without any cutaneous manifestations is difficult to diagnose pre-operatively. The clinical and radiological findings are the most important clues in the diagnosis but it is a diagnostic dilemma. The preferred treatment option is surgical excision. The role of adjuvant therapy is not well known.

### REFERENCES


