

Case Report

A Rare Case of Primary Malignant Melanoma in Brain

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	International Archives of Integrated Medicine, Vol. 6, Issue 7, July, 2019.	
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	Available online at http://iaimjournal.com/	
	ISSN: 2394-0026 (P)	ISSN: 2394-0034 (O)
	Received on: 20-06-2019	Accepted on: 27-06-2019
	Source of support: Nil	Conflict of interest: None declared.
How to cite this article: Ravikumar Mansuriya, Jigna Patel. A Rare Case of Primary Malignant Melanoma in Brain. IAIM, 2019; 6(7): 87-90.		

Abstract

Malignant melanoma arises from either melanocytes or their precursor cells, melanoblasts. As melanocytes are found in normal leptomeningeal tissue, it is not surprising that primary melanomas can grow within the central nervous system. Primary intracranial melanoma is uncommon and accounts for only approximately 1% of all cases of melanoma. It is difficult to diagnose a primary CNS melanoma upfront. Here, we are presenting a rare case of primary malignant melanoma in left frontoparietal area in 65 years old male patient. This case provides us with a good learning opportunity, which is to increase recognition and awareness of rare entity of primary malignant melanoma.

Key words

Primary Malignant melanoma, Intracranial, Leptomeningeal tissue.

Introduction

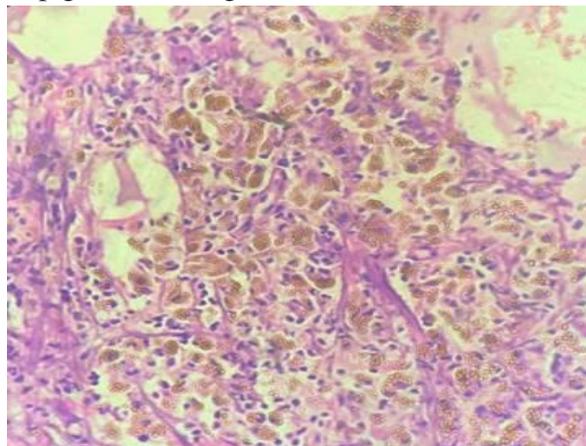
Malignant melanoma arises from either melanocytes or their precursor cells, melanoblasts [1]. As melanocytes are found in normal leptomeningeal tissue, it is not surprising that primary melanomas can grow within the central nervous system. Primary intracranial melanoma is uncommon and accounts for only approximately 1% of all cases of melanoma [2]. Primary Malignant melanoma is often

complicated to diagnose. Radiological investigation e.g. CT scan and MRI can help, but histopathological examination is the final tool for the diagnosis. Here, we are presenting a rare case of primary malignant melanoma in left frontoparietal area in 65 years old male patient. This case provides us with a good learning opportunity, which is to increase recognition and awareness of rare entity of primary malignant melanoma.

Case report

A 65 years old male patient presented to Neurosurgery OPD with chief complaint of progressive weakness of all four extremities and vomiting since 2 months. On examination patient is not able to move his upper and lower limbs. He underwent MRI Brain which showed large extra axial heterogeneously enhancing mass lesion arising from base of anterior cranial fossa mostly in the left frontoparietal area suggestive of meningeal metastasis. Patient underwent craniotomy surgery and the tumor tissue was sent to the histopathology laboratory for examination. After processing of the tissue multiple sections were taken. On microscopic examination, there was presence of hyper cellular sheets, nests and clusters of malignant spindle and epithelioid cells. The individual cells showed hyper chromatism, high N:C ratio, abnormal mitosis and abundant melanin pigment (**Photograph - 1, 2**). On IHC examination, the tumor cells showed S100, HMB 45, EMA and vimentin positivity (**Photograph - 3, 4, 5**). From overall histomorphological and IHC findings the diagnosis of Malignant Melanoma of brain was given.

Photograph – 1: Hyper cellular sheets and nests of pigmented malignant cells (H&E Stain, 10 X).

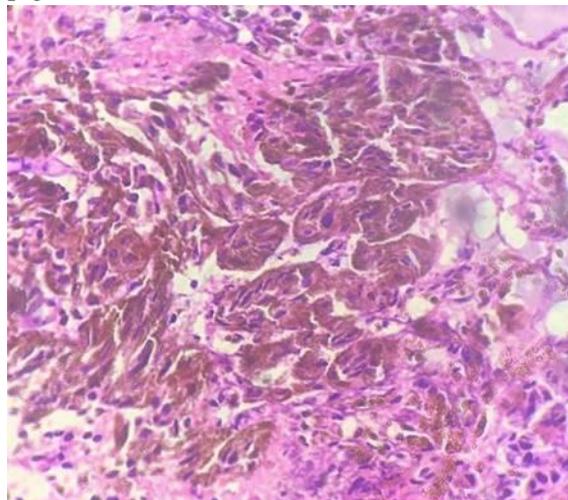


Discussion

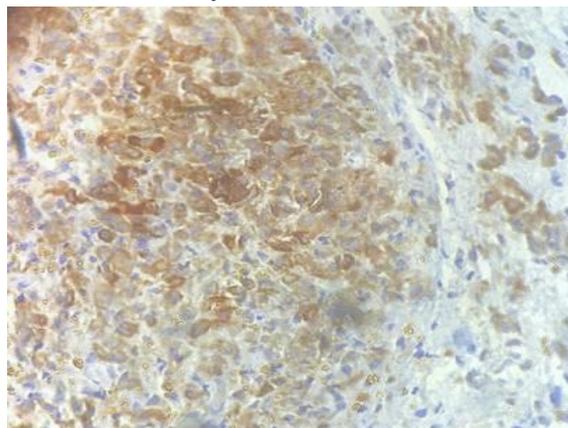
Melanocytes are of neural crest in origin and they migrate during development to the skin, eyes, oral cavity, and the leptomeninges. Primary melanoma of the CNS originates from the melanocytes of the leptomeninges. Primary

malignant melanoma is very rare, and the other sites of possible primary melanoma in the body should be excluded by clinical and radiological examination [2]. Primary melanocytic tumors of the central nervous system (CNS) form a rare entity which is histologically and clinically distinct from metastatic cutaneous or retinal malignant melanoma. It is difficult to diagnose a primary CNS melanoma upfront.

Photograph – 2: Clusters of malignant spindle and epithelioid cells with abundant melanin pigment (H&E Stain, 40X).



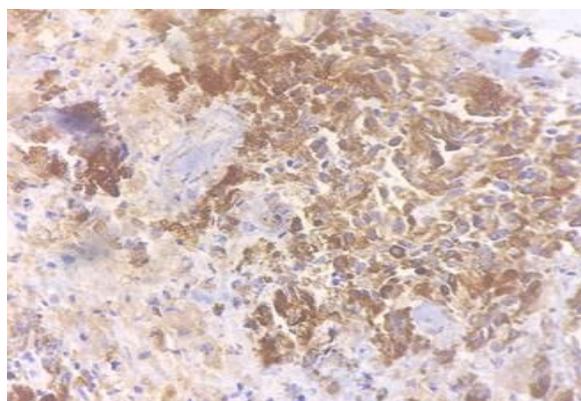
Photograph – 3: Immunohistochemical stain, HMB45 Positivity.



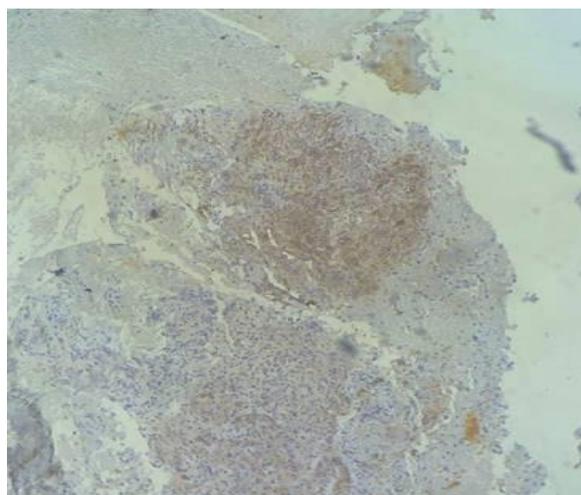
Majority of patients with intracranial melanomas present with features of raised intracranial tension (43%), neurological deficits (35%), features of subarachnoid haemorrhage (14%), or convulsions (12%) [3]. There is a documented male predominance in primary CNS melanomas,

same as our patient. The preoperative diagnosis of primary CNS melanoma is difficult. The CT findings of intracranial melanomas, including high density mass on precontrast scans, homogeneous enhancement, and marked peritumoral oedema, are not specific [4–6]. MRI brain with gadolinium is the imaging of choice for a patient with suspected CNS melanoma. The MRI findings of a CNS melanoma are so typical that they are hyperintense on T1 weighted images and hypointense on T2 weighted images due to presence of melanin [7].

Photograph – 4: Vimentin positivity in Immunohistochemical stain.



Photograph – 5: S100 positivity in Immunohistochemical stain.



The differential diagnosis is more difficult in some cases of meningioma with cells containing melanin pigment, ectopic meningioma, or hemorrhagic meningioma [8]. In our case also

the diagnosis by MRI showed possibility of Meningioma.

Hayward [9] proposed the following factors for establishing a diagnosis of a primary CNS melanoma: (a) No malignant melanoma outside the CNS, (b) leptomeningeal involvement, (c) intramedullary spinal lesions, (d) hydrocephalus, (e) tumor location in the pituitary or pineal gland, and (f) a single intracerebral lesion. HMB-45 is an antibody with a higher specificity for melanocytic tumors. According to the literature, 86–97% of melanocytic tumors are positive for HMB-45 antigen [10, 11]. In our case also the tumor cells showed HMB 45 positivity. Tumor bleeding can be detected by positive Prussian blue stains for iron.

The prognosis of the patient with solitary primary intracranial melanomas depends on the degree of mitosis, leptomeningeal dissemination, extent of surgical excision, and location of the tumor [12]. The clinical outcome of patients with primary CNS melanoma is reported to be better than that of patients with metastatic disease [13].

Options are limited for the treatment of Malignant Melanoma of the brain. Standard treatment models include surgical resection of the tumor, whole brain radiotherapy, stereotactic radiosurgery and chemotherapy [14, 15, 16, 17].

Conclusion

We have presented a rare case of primary malignant melanoma in left frontoparietal area in 65 years old male patient. This case provides us with a good learning opportunity, which is to increase recognition and awareness of rare entity of primary malignant melanoma.

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