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Case Report

Primary Cerebral Malignant Melanoma: A Rare Entity in a Female Patient

Authors:
Dr. M. R. Vishwateja[1*], Dr. K. Kiran Kumar[2], Dr. Nadia Mateen[3], Dr. Avilash Banerjee[4], Dr. Surjali Roy[5]

[1]Consultant Radiation Oncologist, Department of Oncology, Yashoda Hospitals, Somajiguda, Hyderabad, Telangana.
[2]Senior Consultant Radiation Oncologist, Department of Oncology, Yashoda Hospitals, Somajiguda, Hyderabad, Telangana.
[3]Associate Consultant Radiation Oncologist, Department of Oncology, Yashoda Hospitals, Somajiguda, Hyderabad, Telangana.
[4]Radiation Oncology Resident, Department of Oncology, Yashoda Hospitals, Somajiguda, Hyderabad, Telangana.
[5]Radiation Oncology Resident, Department of Oncology, Yashoda Hospitals, Somajiguda, Hyderabad, Telangana

Corresponding Author*:
Dr. M. R. Vishwateja, Consultant Radiation Oncologist, Department of Oncology, Yashoda Hospitals, Somajiguda, Hyderabad, Telangana.

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ABSTRACT:

Background: Primary cerebral malignant melanoma is a rare type of brain tumor, typically found in adults. These primary melanomas have an aggressive nature and have the potential to spread to other organs. In the pediatric demographic, a limited number of cases only documented in the existing literature. The main objective of this report was to delineate the anticipated imaging features and underscore the significance of adopting a multidisciplinary strategy in the treatment of this uncommon condition.

Case Presentation: We present a rare case of the primary cerebral malignant melanoma in a 16-year-old young female. Computed tomography (CT) scan showed an extra-axial lesion seen in the right frontal region with perilesional edema and mass effect. Patient underwent right fronto-parietal craniotomy surgery. An irregularly arranged brownish-black coloured mass present in the right frontal region was completely excised and malignant melanoma was identified by histopathology. Subsequently, the patient proceeded to undergo radiotherapy and chemotherapy. After receiving radiotherapy and chemotherapy, the patient was asymptomatic, but on her follow-up after 5 months showed disseminated intracranial melanoma metastasis.

Conclusion: The primary treatment of choice for primary cerebral malignant melanomas is complete resection combined with radiotherapy, chemotherapy and immunotherapy. These treatments in combination can improve the prognosis.

***Keywords:** Malignant melanoma, Metastasis, whole brain radiation therapy (WBRT)*

INTRODUCTION:

Malignant melanomas are frequently developed in the uvea, brain parenchyma, leptomeninges, mucosal linings and epidermis.[1] Nearly 12 to 17 percent of all cases of intracranial malignancies are malignant melanomas, with the majority of them are metastatic.[2] Approximately 1% of cases of melanoma are primary cerebral melanomas.[3] Even though preoperative anamnesis and computed tomography (CT) patterns closely resemble those of meningiomas, the prognosis of these tumors varies depending on the location of the tumor and the degree of resection; for this reason, it's critical to make an accurate diagnosis and determine the precise extent of the tumor area.[4] Based on the clinical presentation, possible treatment options include surgical resection, whole-brain radiation therapy (WBRT), stereotactic radiosurgery, chemotherapy, immunotherapy and systemic therapy.[5] Our aim is to highlight the importance of employing imaging methods in the initial diagnosis and treatment of primary brain tumors through the presentation of this case report.

CASE PRESENTATION:

A 16-year-old female presented with a history of headache for one week and an episode of seizure. She has not reported experiencing a fever, vomiting or dyspnea and Glasgow coma scale (GCS) examination was normal. MRI and contrast CT scan showed a well-defined extra-axial lesion measuring 3.0 x 2.5 x 2.6 cm with perilesional edema in right high frontal para-falcine

region and a high-density mass on pre contrast scans with homogenous enhancement and marked peritumoral edema. Heterogeneously hyperintense T1W signals and heterogeneously hypointense T2/FLAIR imaging signals were seen on the MRI scan. The patient's mass effect has been observed as a leftward shift in the midline and effacement of the right lateral ventricle. The patient had surgery after meningioma was suspected based on CT and MRI features.

She underwent right frontal parietal craniotomy and her brain tumor was completely removed. During surgery, an irregular mass of brownish-black color was noticed in the right frontal region. Complete excision of the frontal lesion was done and the tissue was sent to the surgical histopathology laboratory. Microscopic analysis of melanoma block showed solid sheets of highly pleomorphic plasmacytoid to epithelioid cells with intracytoplasmic brownish black pigment and macro nucleoli had

infiltrated the brain parenchyma. On Immunohistochemistry, the tumor cells exhibited positivity for HMB-45 (Fig 1), S-100 (Fig 2) and Vimentin (Fig 3) and negative for GFAP (Fig 4) and Dopa oxidase reaction was found positive. These characteristics point to malignant melanoma. Microscopic analysis showed that solid sheets of highly pleomorphic plasmacytoid to epithelioid cells with intracytoplasmic brownish black pigment and macro nucleoli had infiltrated the brain parenchyma.

For additional care, patient was directed to a radiation oncologist. We did whole body PET scan from vertex to foot after injecting 8.3 mCi of F18-FDG and CT images were acquired after injecting IV contrast to check for any extra cranial disease status (Fig 5). PET CT scan findings revealed that non FDG avid enhancing lesion at the postoperative site in the right frontal lobe (Fig 6). There was no evidence of any metabolically active primary lesion present in rest of the body.

Treatment:

She was planned for external beam radiation with a WBRT of 30 GY in 10 fractions, sequentially boost given to the tumor area with a dose of 15 GY in 5 fractions. After the tumor board discussion with medical and neuro oncologist's opinions were sought, and the patient was kept on oral temozolomide therapy. We explained about the immunotherapy option to the patient, as the patient was not financially fit for it, then she was kept on Oral TMZ chemotherapy. Patient was asymptomatic after radiotherapy and chemotherapy, but her follow-up MRI scan after 5 months showed multiple altered signal foci along the cortical margins appearing hyperintense in T1 showing post contrast enhancement involving right frontal, parietal and bilateral temporal lobes, left insular cortex, postcentral gyrus and pons- showing disseminated melanoma metastasis.

DISCUSSION:

Primary brain melanoma represents a rare condition, with a limited number of documented cases in the medical literature. It is responsible for approximately 0.005 cases per 100,000 individuals, constituting about 0.1% of all brain tumors (6).

The MRI evaluation of melanoma was challenging, because melanin pigment is paramagnetic, the amount of melanin pigment and blood volume in the tumor are the two main factors affecting the results of the MRI. Isiklar et al., categorized the MRI presentations into four distinct groups: The melanotic group, exhibiting hyperintensity on T1 and hypo intensity on T2; The amelanotic group, exhibiting exhibiting iso-/hyperintensity on T1 and iso/hyperintensity on T2;The mixed group not meeting either of the two criteria and the hemorrhagic group exhibiting traits of intra/peritumoral hemorrhage(7). In the present case the most common lesions are characterized by a hyperintense on T1- weighted sequences and hypointense on T2-weighted sequences

Figure 1: HMB-45 Positivity

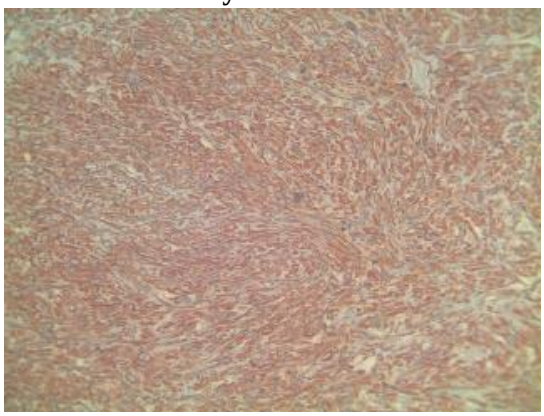


Figure 2: S-100 Positivity

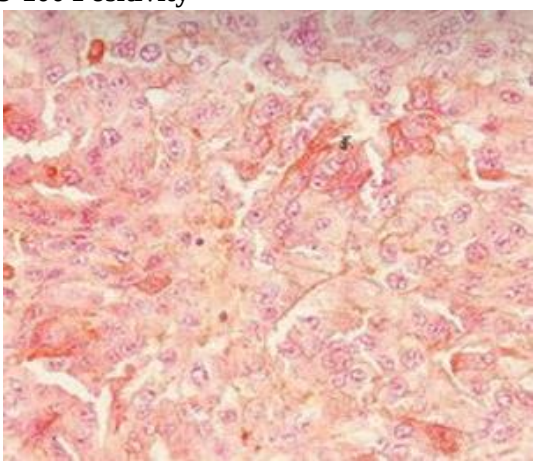


Figure 3: Vimentin Positivity

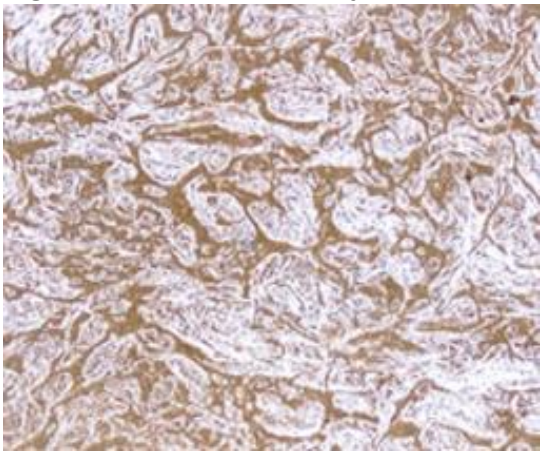


Figure 4: GFAP Negative

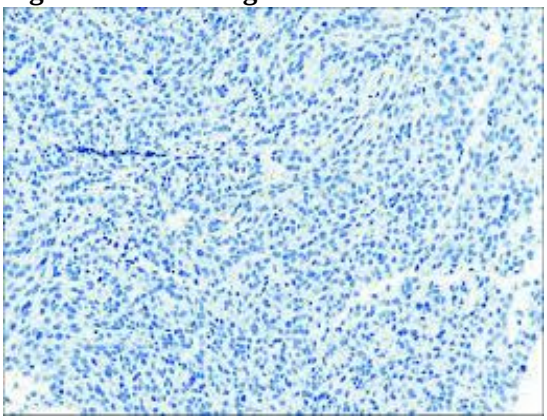


Figure 5: PET Scan from vertex to mid-thigh to cheek for any extracranial disease status

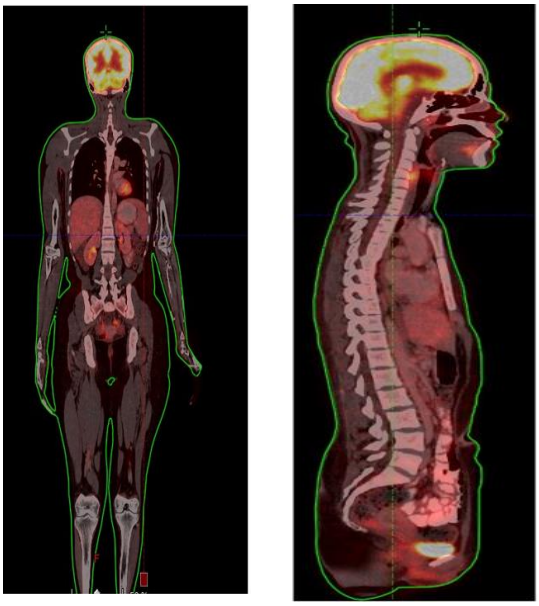
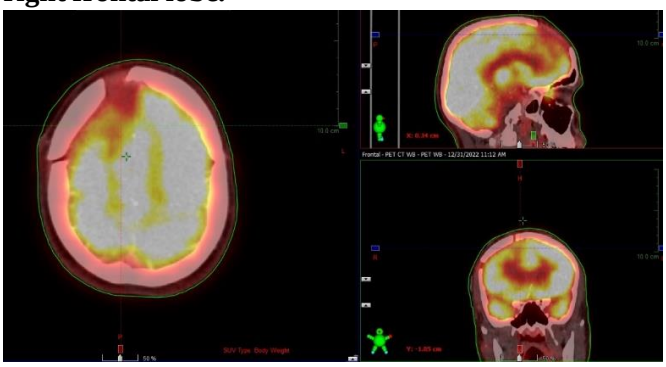


Figure 6: PET Scan in the post operative site in the right frontal lobe.



Strong post contrast enhancement on a CT scan [8]. iso-/hyperintensity on T1 and iso-/hyperintensity on T2; The mixed group, not meeting either of the two Cerebral melanomas can be detected with non-specific CT imaging. A frequently observed characteristic of primary solitary cerebral melanomas on CT scans is a visibly situated hyperdense mass that significantly improves in contrast after enhancement. Though they frequently mimic the presence of meningioma, these results may point to the diagnosis of melanoma rather than glioma.[9]

In the current case, CT scan showed a high-density mass with homogenous enhancement on pre contrast scan.Total body F-18 FDG PET/CT and post-hoc PET/MRI images may aid medical professionals in choosing the most effective course of treatment for individuals suffering from primary meningeal melanoma.[10] According to Yunfeng Ma et al., whole-body F-18 fluorodeoxyglucose (FDG) position emission tomography (PET)/CT helped to rule out the possibility that the melanoma lesions had an extra cranial origin.[11] In our case we also used F-18 FDG PET/CT scan to check for any extra cranial disease status. We didn't find any active primary lesion present in the body.

Radiation therapy has traditionally been viewed as a palliative treatment for advanced cases or widely disseminated diseases.[12] However, according to certain publications, radiation therapy for leptomeningeal disease and residual tumors is linked to better survival .[13]

It can be challenging to diagnose primary cerebral melanomas before surgery. These melanomas usually appear hyperdense and exhibit

In such cases, Radiation therapy can be given to the surgical cavity after surgery as fractionated doses or a (whole brain radiation therapy (WBRT).[14] Due to the severity of the disease at the time of initial diagnosis, our patient received whole brain radio therapy (30 GY in 10 fractions) and sequential boost given to the tumor area to a dose of 15 GY in 5 fractions.

Treatment for primary cerebral melanomas is thought to involve total resections along with efficient postoperative radio chemotherapy.[15] Recently, Dacarbazine which has an efficacy rate of 16-20% has emerged as the most popular and effective chemotherapy drug. It can be used post-radiation therapy or surgery.[16] In our case, after

radiation therapy, our patient received oral Temozolomide as chemotherapy drug.

According to Salpietro et al. certain immunotherapies with low toxicities were essential adjuvant treatments for small residual malignant melanomas. Although it has been discovered that high doses of interferon (IFN) b or IFN a-2b can prolong survival and improve disease control, the ideal dosages remain debatable.[17] In this case, we informed to the patient about immunotherapy, but the patient was not financially fit for it.

The majority of patients with metastatic tumors have an average survival time 2-3 months. Patients who survive for longer than a year are extremely rare.[18] With Adjuvant radiotherapy and oral Temozolomide (TMZ) chemotherapy, our patient had no symptoms, but after five months MRI scan revealed a disseminated melanoma metastasis.

CONCLUSION:

The primary cerebral melanoma is a very uncommon condition, particularly seen in pediatric and adolescent age group. Their nonspecific presentation and imaging results make a definitive diagnosis difficult. The most reliable way to identify primary cerebral malignant melanomas and distinguish them from other tumors is to correlate immunohistochemical staining with imaging results. To manage such a rare entity, a multidisciplinary neuro-oncology tumor board should be formed, with members including neurosurgeons, radiologists, pathologists, and oncologists. Therapeutic advancements in immunotherapies, chemotherapy, and targeted therapies may lead to more effective management of primary cerebral melanomas.

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