Primary Intracranial Malignant Melanoma: A Case Report And Review Of Literature

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Case report

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Abstract

Background

Primary Intracranial Malignant Melanoma (PIMM) is a very rare neoplasm and account for 1% of all melanomas and 0.1% of all intracranial tumors. It carries a poor prognosis with overall poor survival.

Case Presentation:

We here present a case of PIMM in a 35-year-old male presenting with sign and symptoms of a left cerebellar hemisphere tumor with leptomeningeal spread.

Conclusions

Diagnosis on mere imaging findings of an intracranial melanoma is a daunting task. Multimodality treatment with surgery followed by chemotherapy and radiotherapy have proved to be effective in improving clinical outcome in these patients.

Introduction

Primary Intracranial Malignant Melanoma (PIMM) is a very rare neoplasm and account for 1% of all melanomas[1]. It accounts for 0.1% of all intracranial tumors[1]. PIMMs are derived from melanocytes that are normally present in leptomeninges. It is more common in males and have a poor survival rates over all. Pathologically it is of two types, diffuse type which infiltrates pia-mater and the subarachnoid space (SAS); and the solitary type which is present as nodular mass. Treatment of choice is complete resection followed by radiotherapy and chemotherapy. We here present a case of PIMM in a 35-year-old male presenting with sign and symptoms of a left cerebellar hemisphere tumor with leptomeningeal spread.

Case Discussion

A 35-year-old male presented to our institute with complaint of headache, decreased hearing in left ear, difficulty in walking and diminished vision bilaterally for the past 2 years. Non-contrast computer tomography (CT) head revealed hyperdense lesion in left cerebellar hemisphere with associated perilesional edema (Figure 1,A). Contrast enhanced MRI brain revealed a 4.5cm x 5cm x 3.5cm homogenous mass lesion in the left cerebellar hemisphere which was hyperintense on T1 and hypointense on T2 with mass effect and perilesional edema (Figure 1,B,C,D). Radiographic features were suggestive of meningioma. No evidence of hemorrhage on GRE sequence was seen.

Patient was taken for surgery and complete excision was done through left retro-mastoid suboccipital approach. Meningeal spread of the tumor was evident from the dark colored dural after removal of the

bone flap (Figure 2,A). Intraoperative observation revealed a firm, well-demarcated, highly vascular, dark brown, 5cm x 4cm sized lesion with dural attachment (Figure 2,B,C).

Histopathological examination confirmed melanocytes on H and E stain. Tumor consisted of polygonal to spindle shaped cells disposed in sheets with abundant dark melanin pigment deposits in the cytoplasm. These cells stained positively with human melanin black-45 (HMB-45) antibody, S-100 and Melan A. It was reported as malignant melanoma.

Patient was evaluated for alternative site of melanoma but no such sites were found. Postoperative period was uneventful and patient was discharged on tenth postoperative day.

Discussion

Primary Intracranial Malignant Melanoma (PIMM) is a very rare neoplasm and account for 1% of all melanomas[1]. It account for 0.1% of all intracranial tumors[1]. PIMMs are derived from melanocytes that are normally present in leptomeninges. It is more common in males and have a poor survival rates over all[2].

In humans melanocytes are found in skin, mucous membrane, uvea, etc. There are many theories for the origin of primary intracranial melanomas. Endodermal theory states that some aberrant embryonic ectodermal cells in central nervous system produces melanin pigment. Neurogenic theory states that neural crest gives rise to pigment cells. Neural crest later develops into mesodermal and neural elements. Mesodermal theory proposes that the mesodermal pigment cells reach the central nervous system through pial vessels.

Hayward proposed few features of a primary intracranial melanoma[3]. He suggested that for a PIMM, there should no malignant melanoma outside the CNS, evidence of leptomeningeal involvement, hydrocephalus, intramedullary spinal lesions, tumor located in pituitary or pineal region and a single intracerebral lesion.

Gibson et al divided intracranial melanomas into two pathological types[4]. Diffuse type which infiltrates pia-mater and the subarachnoid space (SAS); and the solitary (discrete) type which is present as nodular mass. Diffuse type was more prevalent in younger patients (mean age 26-years). It presented with clinical features of intracranial hypertension, cranial nerve deficit or meningism. Solitary type is more common in adults (mean age 44-years). Most of the solitary tumors are supratentorial. Leptomeningeal involvement is frequently seen in solitary melanoma. Our patient has a solitary lesion.

Symptoms included headache, vomiting (especially due to raised intracranial pressure), hydrocephalus (43%), focal neurological deficit (due to mass effect, 35%) and convulsion or subarachnoid hemorrhage (16%). Headache and focal neurological deficit were present in our case. Intracranial hypertension and hydrocephalus is more commonly seen in diffuse type. Presence of congenital melanocytic nevus

increases risk of malignant melanoma. 25% of patients of primary intracranial melanomas have congenital nevus. This was not the case in our patient. Extracranial metastasis of PIMM is rare.

It is difficult to differentiate primary central nervous system (CNS) melanomas from metastatic melanomas on the basis of imaging alone. Melanin cells in cerebrospinal fluid (CSF) analysis helps in the diagnosis. 86-97% melanocyte tumors are positive for HMB-45 Antigen. Other commonly used tumor markers are S100 and Melan A. All three were positive in our patient.

Treatment of choice for intraparenchymal melanocytic tumor is complete resection and postoperative chemoradiotherapy. Survival in adults can be up to 17 years and 8 months in pediatric patients[5]. Mean survival was found to be better (19.6 months) in patients in whom complete resection was done as compared with those where partial resection was done (9.3 months). Also prognosis of secondary melanoma was poorer than PIMM. There is 95% mortality in secondary intracranial melanomas.

Initially, radiotherapy (RT) was not offered to patients of PIMM as melanomas were radioresistant tumors but recent studies showed that it was effective in controlling tumor growth. Stereotactic Radiosurgery (SRS) along with whole brain radiotherapy (WBRT) has been shown to reduce recurrence and improve survival. High dose of 5000 centigray as SRS in small lesions (size less than 3cm) gave better tumor control in almost 80% of cases. Hydrocephalus caused by leptomeningeal spread of the disease has been shown to respond well to WBRT.

Dacarbazine is commonly used in the treatment of PIMMs after surgery or radiotherapy. It has 16-20% effectiveness. Another agents are temozolomide and Dimethyl-triazeno-imidazole-carboxamide which is common in use.

Despite optimum treatment, median survival is less than a year. Leptomeningeal spread have a median survival of 10 weeks.

Conclusion

Primary Intracranial Malignant Melanoma (PIMM) is a very rare neoplasm and account for 1% of all melanomas and 0.1% of all intracranial tumors. It carries a poor prognosis with overall poor survival. Diagnosis on mere imaging findings of an intracranial melanoma is a daunting task. It is best diagnosed intraoperatively due to its characteristic appearance as well as during preoperative clinical examination. Postoperative chemoradiotherapy plays an important role in preventing recurrence as well as improve overall survival. Collaboration of large scale data from many such small case series from centers all over the world can confirm findings suggested in this paper and formulate guidelines to improve clinical outcome.

Declarations

- Ethics approval and consent to participate: Taken

- Consent for publication: Taken
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Figures

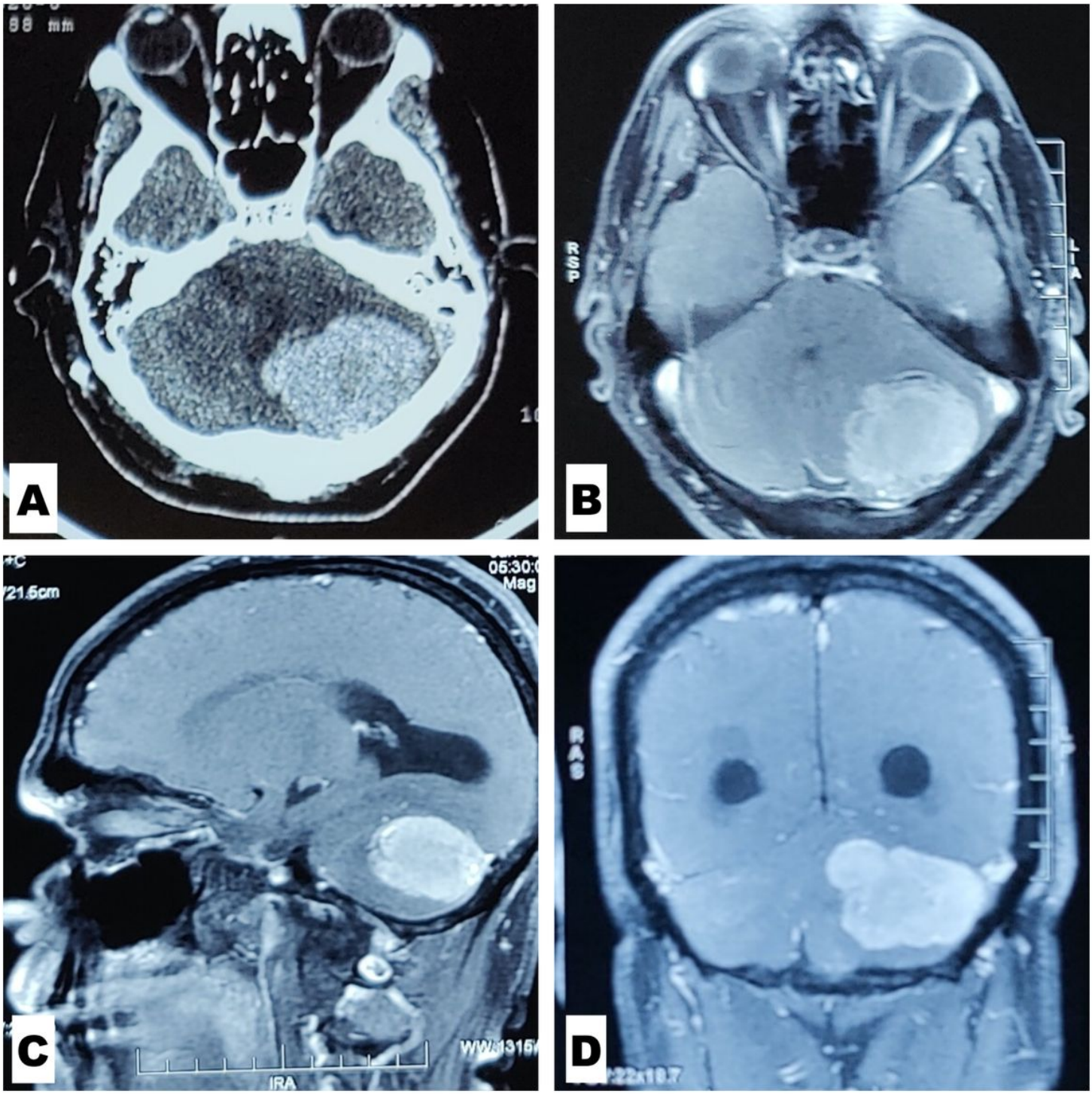

Figure 1

Preoperative CT Non-contrast (CT) and Contrast Enhanced MRI images. (A) CT images revealed hyperdense lesion in left cerebellar hemisphere with associated perilesional edema. (B) Axial view, contrast enhanced T1WI MRI. (C) Sagittal view, contrast enhanced T1WI MRI. (D) Coronal view, contrast enhanced T1WI MRI. Contrast enhanced MRI brain revealed a 4.5cm x 5cm x 3.5cm intra-axial heterogenous mass lesion in the left cerebellar hemisphere which was hyperintense on T1 and hypointense on T2 with mass effect and perilesional edema.

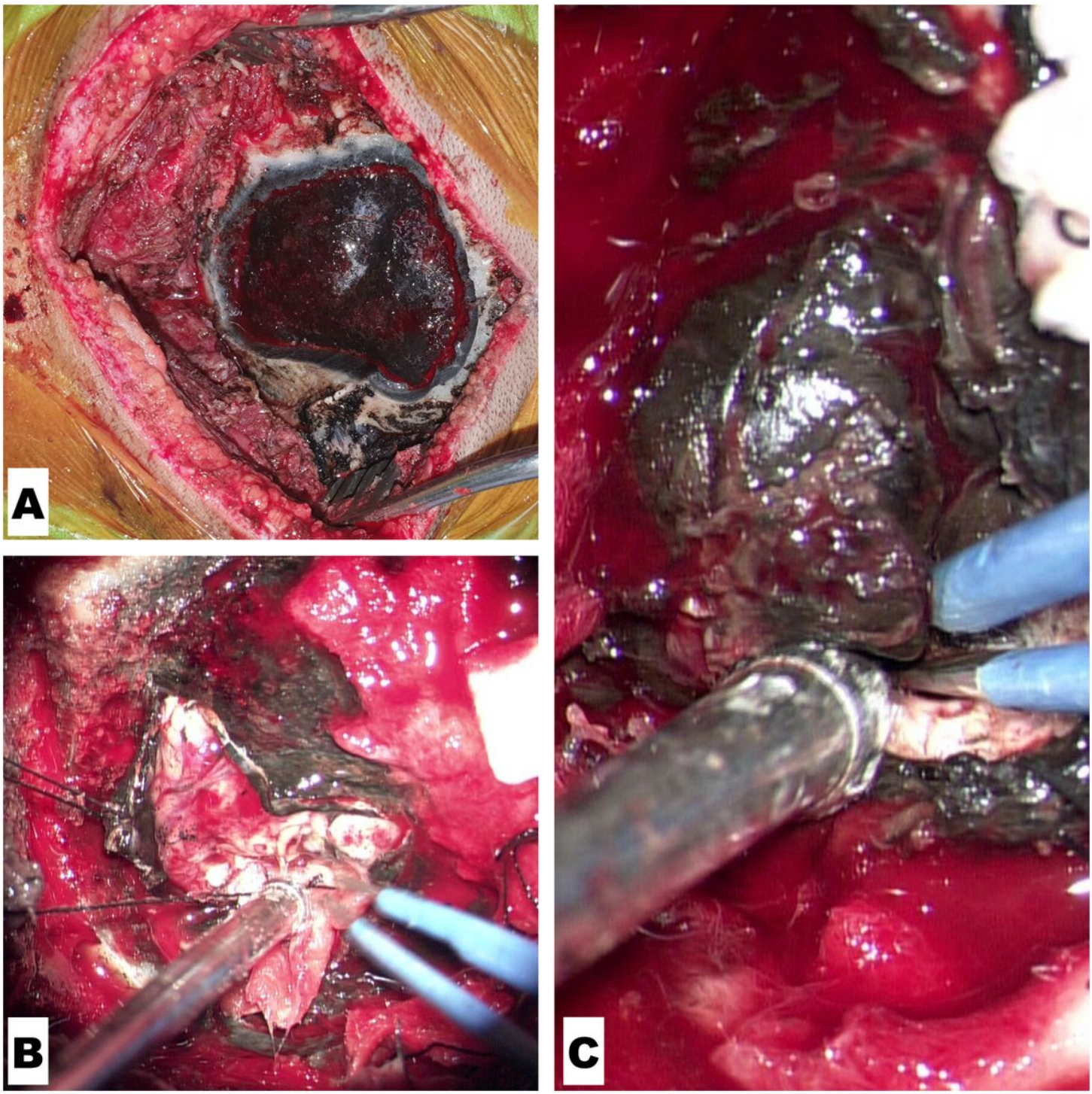

Figure 2

Intraoperative images showing gross total resection of dark colored lesion with leptomeningeal involvement and dural attachment.