Primary Malignant Melanoma of brainstem medulla mimicking as cavernoma – Case Report

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Abstract

40-year-old male presented with vertigo, headache, dizziness for 1 month. MRI showed pear shaped T1 hyperintense lesion at medulla oblongata and predominantly hypointense on T2 with focal area of hemorrhage. Lesion showed diffuse enhancement on postcontrast images. On plain CT lesion was Hyperdense. It was initially reported as Cavernoma. Surgical excision of lesion was done with per-op findings of solid, dark maroon colored lesion with hemorrhage. Histopathology showed neoplastic lesion with abundant melanin pigment deposition. The lesion was finally diagnosed as Malignant neoplasm with features favoring Malignant Melanoma.

Aim

Aim of this case report is to present a rare case of primary malignant melanoma of brain stem at the region of medulla oblongata mimicking as cavernoma.

Introduction

Primary malignant melanoma of central nervous system is very rare and accounts for 0.07% of all brain tumors [2]. It has very low incidence, estimated at 0.9 per 10 million inhabitants [3]. Primary CNS melanoma arises from melanocytes which has been developed from melanoblasts in the neural crest. Melanoma of brainstem is difficult to diagnose and distinguish from cavernoma radiographically. Clinical picture is same but treatment and clinical management of these two diseases differ significantly. We report the case of malignant melanoma mimicking as craniocervical junction cavernoma.

Case Report

40 Years old male with no known comorbid presented with complain of vertigo, dizziness and headache for 1 month. On CNS Examination: GCS 15/15, with no neurological deficit. Rest of the clinical examination was unremarkable and routine laboratory tests were normal.

CT and MRI scan of brain with contrast were performed. On plain CT there was pear shaped hyperdense lesion at medulla oblongata of brainstem. Focal area of increased hyperdensity was seen in left posterolateral aspect of the lesion suggestive of focal hemorrhage.

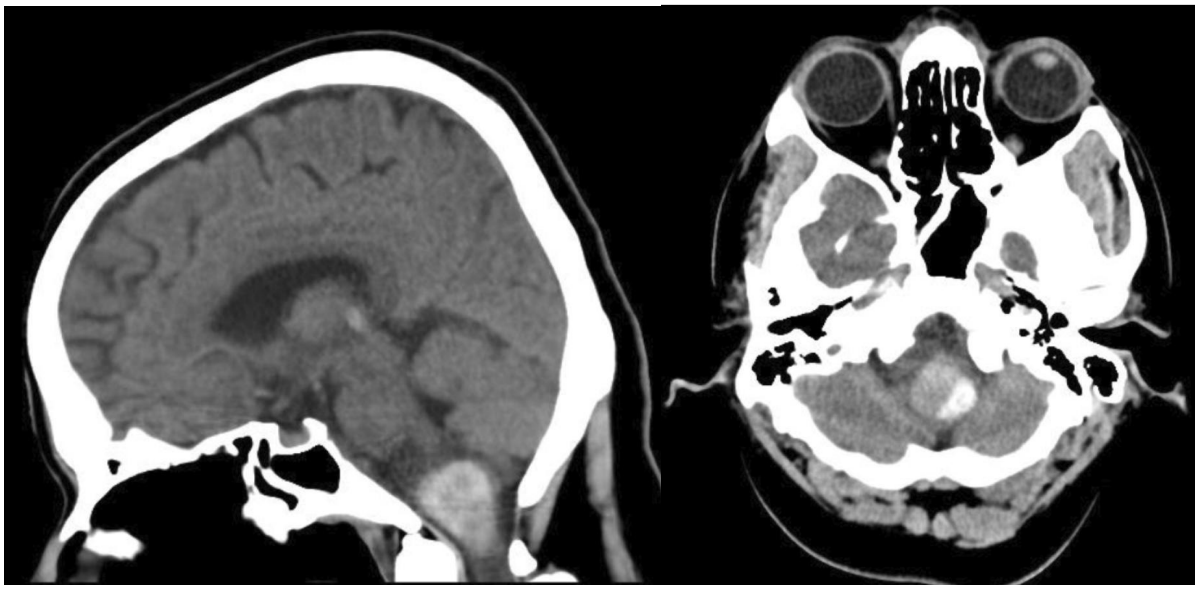

Fig 1. A. Coronal and B. sagittal non-contrast CT images shows well-defined hyperdense lesion in medulla and craniocervical junction with Focal area of increased hyperdensity in left posterolateral aspect of the lesion suggestive of focal hemorrhage.

On MRI, the lesion was hyperintense on T1-weighted images and predominantly hypointense with mottled hyperintensity on T2-weighted images. No diffusion restriction is seen. Areas of susceptibility dropout was noted along the left posterolateral aspect of the lesion representing hemorrhage. Lesion showed diffuse enhancement on postcontrast images. Appearance of lesion raised the possibility of craniocervical cavernoma with focal hemorrhage.

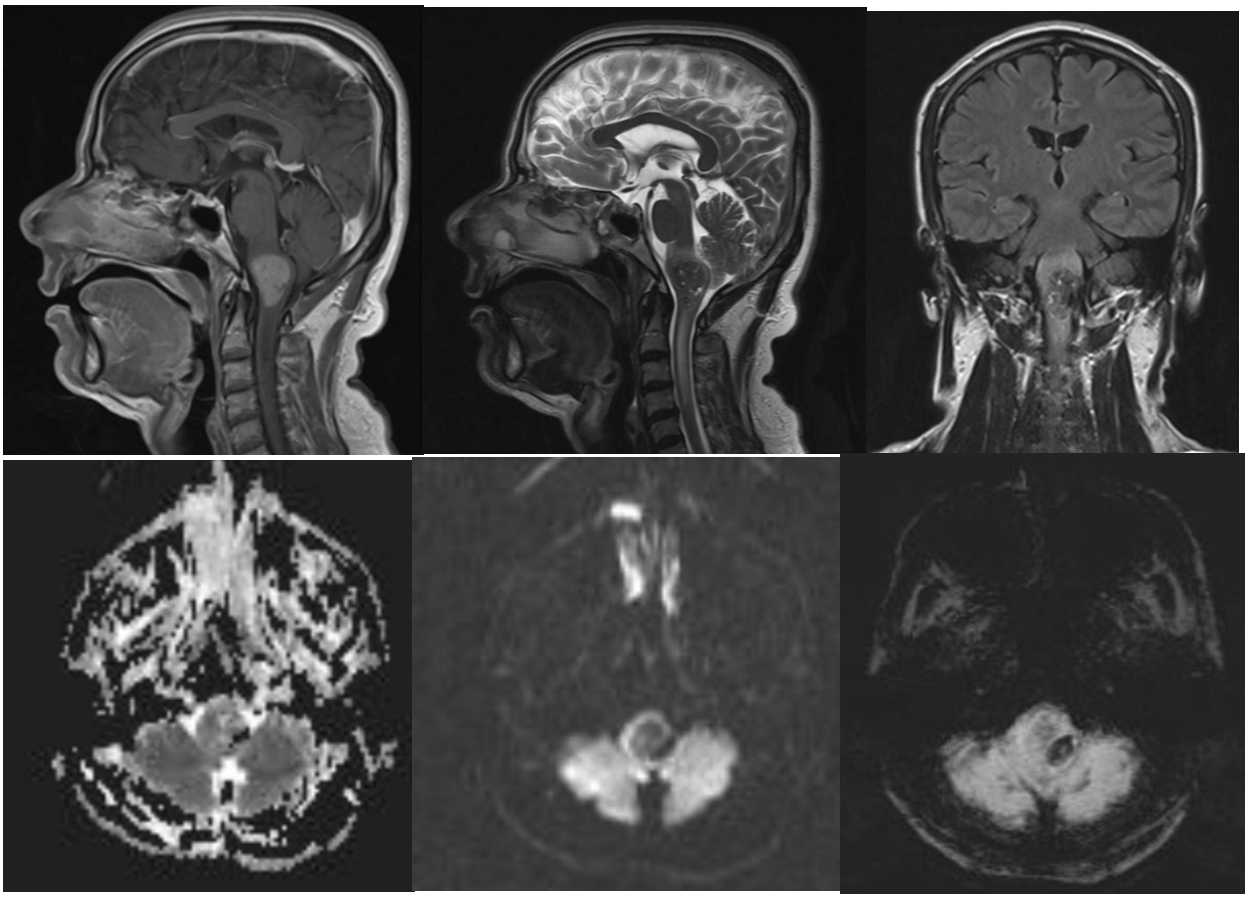

Fig 2. MRI images shows hyperintense lesion at brainstem medulla on T1 (A) which is hypointense on T2 with mottled hyperintensity. No diffusion restriction is seen in lesion. Areas of susceptibility dropout was noted along the left posterolateral aspect of the lesion representing hemorrhage.

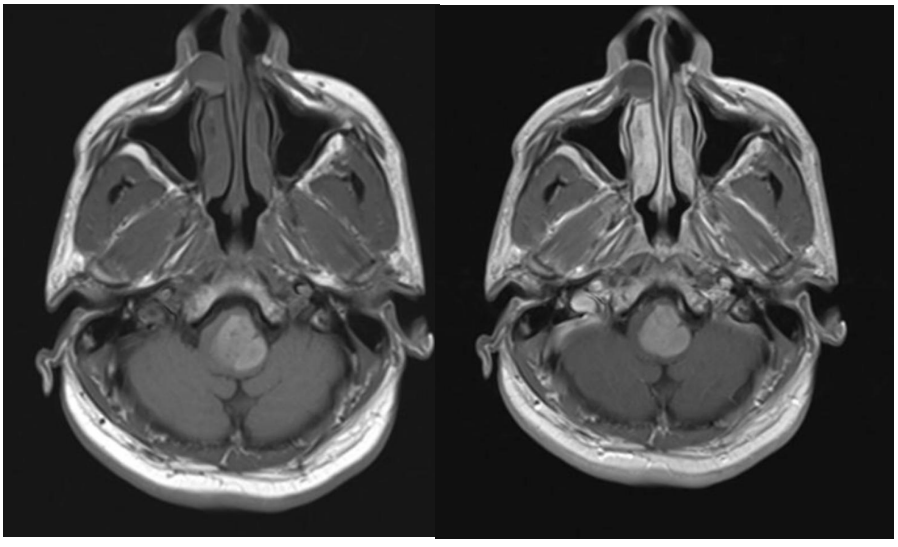

Fig. 3. Axial T1 pre and post contrast images diffuse shows post contrast enhancement of the lesion.

Patient was then admitted for elective surgery after 1-week and neuronavigation guided Craniotomy, Excision of SOL and Lumbar drain placement was done. Per-op findings were of a solid, non-suctionable dark maroon colored lesion with hemorrhage. Obtained specimen was sent for histopathology

Post-op the patient remained vitally and hemodynamically stable. He was first shifted to Special care and then to regular bed, he was slowly progressed to regular diet and he was also ambulated which he tolerated well. After that the patient is stable enough to be discharged home.

Histopathology revealed multiple fragments of a neoplastic lesion infiltrating into the glial tissue. The lesion was arranged in nests of epithelioid neoplastic cells with abundant melanin pigment deposition. These neoplastic cells had abundant amount of eosinophilic cytoplasm. The nuclei were oval, moderately pleomorphic, vesicular with pseudo-inclusions and prominent nucleoli. Increased mitotic activity was appreciated in these neoplastic cells. Immunohistochemical stains were performed which show the following reactivity pattern:

S100 Positive, HMB-45 Positive, Melan-A Positive, Mib-1 (Ki-67) High (approximately 15%).

Based on above histopathologic features, lesion was diagnosed as Malignant neoplasm with features favoring Malignant Melanoma.

Since finding for melanoma other than this lesion was negative in the whole body, it was labelled as primary malignant melanoma.

Discussion

The most frequent occurrence of melanoma in the central nervous system (CNS) is through metastasis. [5]. Primary melanocytic tumors of the CNS, are much rare and should only be considered primary after a thorough evaluation with absence of cutaneous, mucosal (GI) and retinal disease [4]. Up to 20% of melanoma patients with CNS involvement also have brainstem involvement [6]. Metastatic melanoma has a median survival of 113 days [4].

Treatment options for CNS malignant melanoma includes surgery, chemotherapy, radiotherapy and immunotherapy however no standard therapy is present due to poor prognosis [7].

Obtaining the correct diagnosis remains the foremost challenge for brainstem melanomas that can be mistaken for brainstem cavernomas when the lesion is associated with hemorrhage.

Melanomas are typically hyperintense on T1 and hypointense on T2 weighted imaging, however it is not always necessary, T1 hyperintensity of melanoma depends upon the amount of melanin in the lesion [1] [6], if lesion low melanin amount, it will be hypointense on T1. Post contrast imaging shows contrast enhancement within the lesion. T1-weighted, T2-weighted, and T2* or susceptibility-weighted sequences are used to assess hemorrhage [8]

In case of cavernoma usually there is subacute hemorrhage with degraded blood products within the lesion producing a halo of signal hyperintensity around the lesion on T1-weighted images, a useful finding for differentiating cavernous malformations from melanoma [9]. But still recent hemorrhage from a cavernoma may be indistinguishable from other acute or early subacute hemorrhagic lesion making the diagnosis challenging.

The final diagnosis is made after histopathologic examination. Histopathologic features of malignant melanoma includes nests of epithelioid neoplastic cells with abundant melanin pigment deposition. Neoplastic features includes hypercellular sheets or nests of spindled or epithelioid cells, significant pleomorphism, atypical mitoses , invasion of adjacent structures, prominent nucleoli [10].

In addition to histology, the presence of positive S100 and HMB-45 are confirmatory features of malignant melanoma on immunohistochemical staining [11].

Based on histopathology, melanocytoma is considered in differential diagnosis since it also shows melanin pigmentation but it usually do not invade adjacent structures and contains nests of relatively uniform cells with bland, oval nuclei with eosinophilic nucleoli.

Conclusion

Awareness of the unusual presence of melanoma within the brain stem is important and the possibility of presence of Malignant melanoma must be considered when above described MR images depicted. The final diagnosis, however, is based on the results of pathologic examination.

Acronyms

CT=Computed Tomography, MRI=Magnetic Resonance Imaging, CNS=Central Nervous System, HMB=Human Melanoma Black

Ethical disclaimer:

- Exemption for ethical approval regarding this case report has been obtained from ethical review committee of Aga khan university hospital after thorough review and requirement of signed consent form is waived.

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آغا خان یونیورسٹی

THE AGA KHAN UNIVERSITY

24-Jan-2019

Dr Fatima Mubarak
Department of Radiology
Aga Khan University
Karachi

Dear Dr Fatima Mubarak

2019-0966-2438. Dr Fatima Mubarak: Primary Malignant Melanoma of Craniocervical junction and medulla mimicking as cavernoma - Case Report

Thank you for your application for exemption from ethical approval regarding the above mentioned case report.

Your case report was reviewed and approved as exemption. Please ensure that all AKU standards are followed when reporting this case.

Thank you.

Sincerely,

Dr. Khabir Ahmad

Chairperson
Ethics Review Committee

To approve a single suggestion, mouse over it and click "✔"
Click the bubble to approve all of its suggestions.