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Case Report

A Unique Case of Primary Intracranial Melanoma

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Abstract

Primary intracranial melanoma is an uncommon entity and only case reports have been published in the literature. We report a case of an elderly male who was operated with a preliminary diagnosis of meningioma, but it proved to be a histological surprise as it came out to be melanoma with no primary anywhere in the body.

Keywords: *Central nervous system, melanocytes, melanoma*

Introduction

Primary intracranial melanoma is uncommon and accounts for only approximately 1% of all cases of melanoma.[1] Intracranial melanomas are often complicated to diagnose with differential diagnosis of other pigmented lesions such as pigmented meningioma, schwannoma, medulloblastoma, choroid plexus papilloma, astrocytoma, and pituitary tumors.[2] In this report, we present a case of primary intracranial melanoma in the left frontal area; the clinical, neuroradiological, and histological findings are discussed along with a review of the literature.

Case Report

The patient is a 48-year-old man who presented to us with the history of rapidly worsening symptoms of headache and nausea. A left frontal lesion with variegated appearance was detected on noncontrast computed tomography (CT) head [Figure 1]. A contrast-enhanced magnetic resonance imaging brain was done demonstrated a heterogeneous mass lesion, (hyperintense on T1-weighted image [T1-WI] and relatively hypointense on T2-WI) [Figure 2] with mass effect with perilesional edema was seen in the left frontal area. A provisional diagnosis of meningioma was made preoperatively. Patient underwent gross total resection of this lesion. The lesion was large about 7 by 5 cm, blackish, vascular with no dural attachment, and confined to the left frontal lobe. Histopathological examination revealed that the tumor consisted of epithelioid cells, arrayed in sheets, with abundant melanin pigment deposits in the cytoplasm. The tumor was hypercellular with some signs of pleomorphism. Mitoses were present. Foci of hemorrhage and necrosis were seen [Figures 3 and 4]. Both melanin and hemosiderin pigment were identified by Masson-Fontana stain and iron stain, respectively. The neoplastic cells were stained positively with human melanin black-45 (HMB-45) antibody. It was reported as malignant melanoma. Clinical and radiological search for any other site of melanoma in the body was negative. Investigations included chest radiograph, ocular examination, and ultrasonography of abdomen. Postoperative period proved to be uneventful and patient was discharged on the seventh postoperative day with complete excion confirmed by post opertative NCCT Head and MRI Brain [Figures 5 and 6].

Discussion

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.[1] 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. These account for only approximately 1% of all cases of melanoma. During embryonic development, precursor melanocytes originate in the

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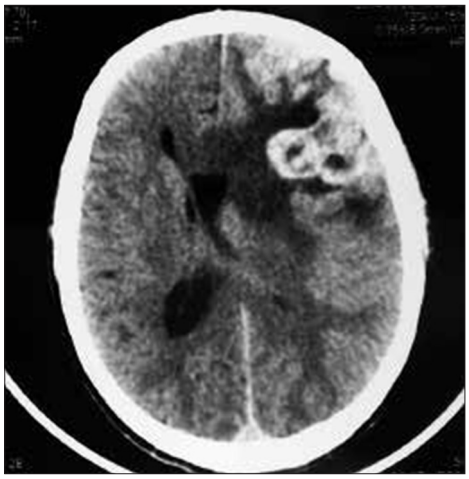

Figure 1: Noncontrast computed tomography head showing a left frontal lesion with variegated appearance, mass effect, edema, and midline shift

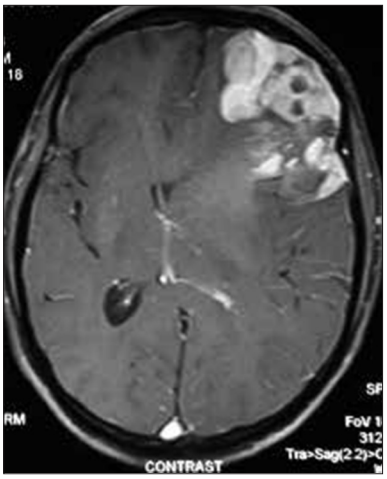

Figure 2: Contrast-enhanced magnetic resonance imaging brain showing a left frontal contrast enhancing mass lesion

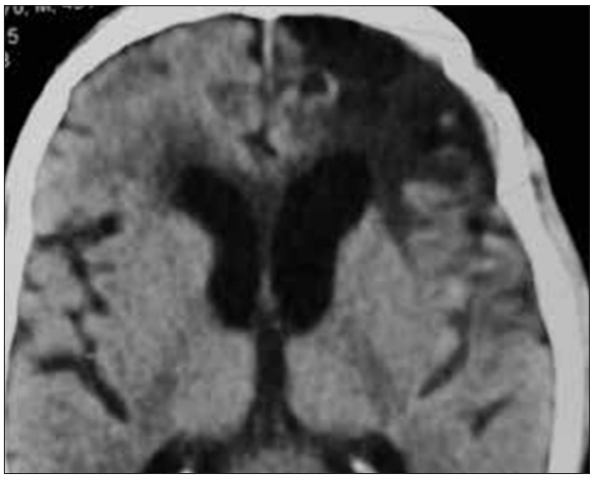

Figure 3: Histopathological examination showing melanoma infiltration in the form of nests with pigment deposition (melanin) and surrounding brain tissue (H and E, ×40)

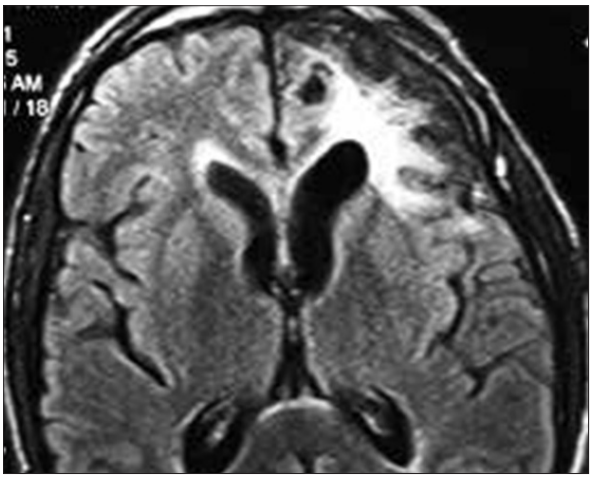

Figure 4: Histopathological examination showing infiltration of melanoma cells with high nucleo-cytoplasmic ratio, prominent nucleoli, and melanin pigment (H and E, ×400)

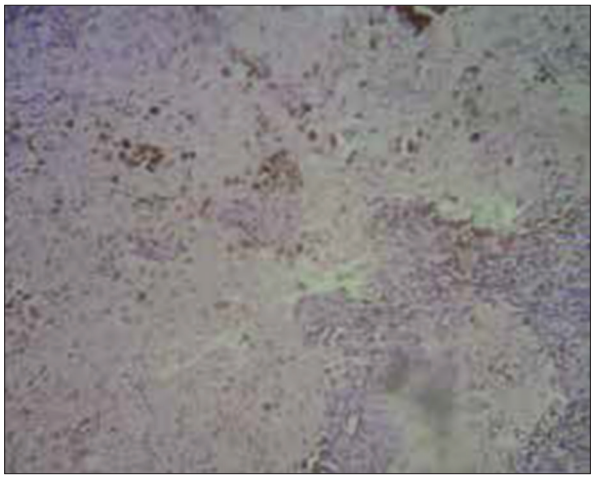

Figure 5: Postoperative noncontrast computed tomography head of patient showing complete excision of mass lesion

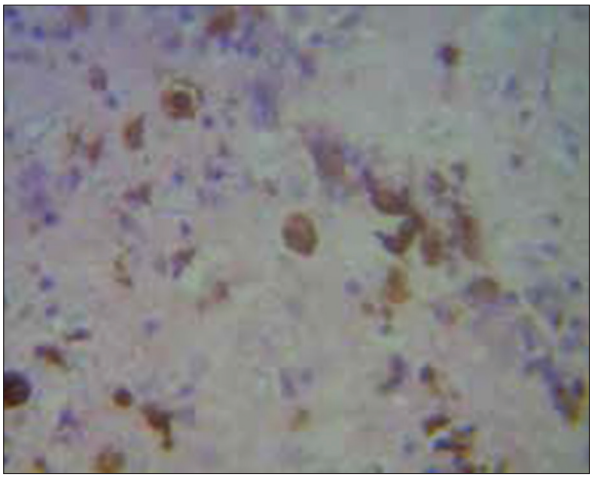

Figure 6: Fluid attenuation inversion recovery sequence of postoperative magnetic resonance imaging showing mild edema with no mass lesion

neural crest and migrate actively to peripheral sites. In humans, melanocytes can be found in skin, mucous membranes, parenchyma, and uvea.[3-5] In most instances, melanomas involving the CNS represent metastatic disease.

Melanoma is the most common tumor, after lung and breast cancers, which produces brain metastases.[6,7]

According to the literature regarding primary melanoma of the nervous system, the origin of melanin cells is not fully understood, although several histogenetic theories have been proposed: (1) The mesodermal theory: The mesoderm gives rise to pigment cells which reach the brain or spinal cord via the pial blood vessels. (2) The ectodermal theory: Some epithelial cells produce pigment, and therefore CNS melanomas are derived from the aberrant embryonic ectodermal cells. (3) The neurogenic theory: The pigment cells originate from the neural crest, which develop into mesodermal and neural elements.[8] Primary intracranial melanomas are either solitary or of diffuse variety.[9] Diffuse leptomeningeal melanomas preferentially affect children and may be part of neurocutaneous melanosis complex or phakomas.[10] Such melanomas usually present with features of raised intracranial pressure, cranial nerve palsies, and meningism.[11] Focal melanomas present as leptomeningeal or dural based neoplasms and are more common in adults. The melanocytic tumors range between the relatively indolent melanocytomas to the aggressive malignant melanomas.[11] 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.[11] The differential diagnoses of leptomeningeal tumors include meningioma, meningeal melanocytoma, leptomeningeal melanosis, metastases, and sarcoma.[4,5] It is difficult to distinguish primary CNS melanoma from metastatic melanoma on neuroimages alone.[12] Hayward[13] 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. In this case, the preoperative diagnostic consideration was a meningioma. Meningioma has similar neuroradiologic findings, such as enhanced hyperdense lesion on CT scan, and contact with the skull and dura mater. 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.[3,14] Tumor bleeding can be detected by positive Prussian blue stains for iron. Radiotherapy, chemotherapy, and immunotherapy may be considered nonsurgical therapy options for primary melanoma of the CNS, but their effects have not been clearly established. Various authors suggested that higher total doses of radiation (>40 Gy) should be irradiated to the tumor area for patients with good general conditions and absence of or controlled extracranial disease.[13-15] As for chemotherapy, dimethyl-triazeno-imidazole-carboxamide is the most commonly employed a chemotherapeutic agent.[16,17] Unfortunately, a standardized therapy concept is still lacking. Literature search showed 95% mortality in case of metastasis of melanoma to the brain. Despite treatment, the median survival is less than a year.[18] The clinical outcome of patients with primary CNS melanoma is reported to be better than that of patients with metastatic disease.[19]

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Conflicts of interest

There are no conflicts of interest.

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