Case Report

A rare case of primary malignant melanoma in CNS

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ABSTRACT

A case of primary Malignant Melanoma at unusual location is being presented. A 50 year old male patient was admitted to Neurosurgery Department of Gandhi Hospital with history of weakness of right upper and lower limbs and difficulty of speech since 20 days. Routine investigations including CT scan were done which revealed a parietal space occupying lesion in the brain. Patient underwent surgery and specimen was sent for histopathological examination. Bleaching and IHC confirmed it as Malignant Melanoma.

Keywords: Primary melanoma, Unusual, CNS, IHC

INTRODUCTION

Intra-cranial melanomas are commonly metastatic from melanoma elsewhere in the body. The occurrence of a melanoma primarily in the brain parenchyma is rare. Primary melanocytic tumours of CNS are very rare tumours with incidence of 0.9/10 million population and account for 1% of all melanoma cases in CNS. Primary melanoma of CNS was first reported by Virchow in 1859.

CASE REPORT

A 50 year old male presented with history of weakness of right upper and lower limbs since 20 days and difficulty in speech since 20 days as informed by patient’s attendant. The complaints were acute in onset and progressive in nature. There was no significant history of trauma, fever, seizures or loss of consciousness. On clinical examination patient was conscious and coherent with right hemiplegia. Bilateral pupil was normal in size and reacting to light. Plain computed tomography gave evidence of a heterogeneous area noted in left parietosagittal location causing compression of left lateral ventricle with grade II perilesional oedema (Figure 1). MRI showed a 5x5x3 cm lobulated haemorrhagic mass lesion in left high frontal parasagittal region with significant surrounding oedema (Figure 2). Imaging studies so suggested presence of parietal space occupying lesion in the brain. Patient underwent surgery and tissue excised was sent for histopathological examination.

Figure 1: CT Brain: heterogeneous area in left parieto parasagittal location causing compression of left lateral ventricle with perilesional oedema.
Cytological smears studied showed abundant necrotic tissue with only little viable tissue showing features of anaplastic oligodendroglioma. Sections from the tissue studied showed tumour tissue composed of round to polygonal cells arranged in sheets and in peritheliotomatous pattern. At places pseudorossette arrangement of cells was seen. The individual tumour cells were having pleomorphic nuclei with scant clear cytoplasm and most of them had intra-cytoplasmic golden brown pigment (Figure 3). There were few areas of necrosis and entrapped islands of native glial tissue. Features were suggesting of pigmented malignant neoplastic lesion with the following possibilities of

1. Pigmented oligodendroglioma,
2. Primitive Neuro Ectodermal Tumour (PNET),
3. Malignant melanoma.

Sections were subjected to bleaching and immunohistochemical (IHC) studies. Bleaching was positive suggesting the intracytoplasmic pigment to be melanin (Figure 4). IHC with HMB 45 and S 100 was positive (Figure 5) but with GFAP, NSE and CD 99 was negative (Figure 6) confirming the diagnosis as malignant melanoma.

DISCUSSION

Primary CNS melanomas are dural based and have been reported in spine, supracellular, pineal, CP angle and cerebral regions. \(^1\) \(^5\)

Primary melanocytic tumors of central nervous system (CNS) are rare tumors and the spectrum ranges from diffuse leptomeningeal melanocytosis, melanocytoma to its overtly malignant counterpart, melanoma. \(^3\)
Metastasis from the extracranial primary melanoma is more frequent to appear in the brain and is the third most common site of intracranial metastasis after carcinoma of breast and lung. However, primary intracranial melanomas are exceptionally rare presenting either solitary or diffusely. Intracranial melanomas are often complicated to diagnose with differential diagnosis of other pigmented lesions like pigmented meningioma, schwannoma, medulloblastoma, choroid plexus papilloma, astrocytoma and pituitary tumors. The melanotic cells originate either from embryonic neuroectodermal cell rests or from the pial sheaths of the vasculature in the CNS.

Hayward in 1976 had defined primary CNS malignant melanomas based on clinical findings. According to him, solitary cerebral lesion, intramedullary or leptomeningeal involvement, hydrocephalus and pineal/pituitary tumors could be primary melanomas if no melanomatous lesion was found outside CNS. A meticulous search for a cutaneous, mucosal or ocular primary melanoma should prove unrevealing before a malignant melanoma can be accepted as indigenous to the CNS. HMB-45 is an antimelanosomal antibody, which is specific for melanocytic tumors. S-100 is a sensitive marker for cells of Neural crest origin and lacks specificity.

Primary melanoma should be suspected after excluding other sites like skin, retina, anal canal, oesophagus & penis or vagina. There should be lack of history of removal of melanocytic lesions previously, lack of history of presence of benign pigmented nevi elsewhere in the body, lack of history of distant metastasis.

CONCLUSION

Primary CNS melanomas are exceedingly rare and are difficult to diagnose preoperatively. They have very poor prognosis. MRI picture, gross findings and IHC play a major role in the diagnosis. Radical excision is the mode of treatment.

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