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PRIMARY INTRACRANIAL MALIGNANT MELANOMA A RARE PIGMENTED C.N.S LESION : A CASE REPORT AND REVIEW OF LITERATURE

Neurosurgery

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ABSTRACT

Primary Pigmented lesions of the Central nervous System are rare and diverse group of entities that run the gamut from Benign to malignant[1]. Among Pigmented CNS Tumor, Primary intracranial melanomas possesses a great interest among Neurosurgeons with its varied Clinical and Radiological patterns as they are most often confused and misdiagnosed with Meningioma, Schwannoma and thrombosed aneurysms . Primary cerebral melanomas, derived from the melanocytic cells that are precursor Cells of Melanoblasts considered to be originated from neural crest cells that are normally present in the leptomeninges. They are rare and occur in around 1% of all melanoma cases[2,4]. Primary intracranial malignant melanoma accounts 0.1% of intracranial neoplasms[3]. This report highlights a case of Primary Intracranial malignant melanoma of clivus in 40 year male, who has undergone left Pterional Craniotomy Trans Sylvian approach and excision of Tumor after proper dermatological, ophthalmological and systemic evaluations to exclude presence of any other lesions in the body. Which was proved malignant melanoma via Histopathology. Herein we have reported this case for its rarity and consideration as differential for prompt treatment.

KEYWORDS

Melanoma, Pigmented Intracranial lesions, Primary intracranial Malignant Melanoma.

INTRODUCTION

Melanocytes have developed from Precursor Cells Melanoblasts considered to be originated from neural crest cells, which occur normally within the skin, mucous membrane, Brain parenchyma, uvea and leptomeninges. Melanocytic lesions consists of rare form neurocutaneous melenosis to benign well differentiated tumor (melanocytoma) to malignant Melanoma. Primary malignant melanomas developed from occult melanocytic lesions present in leptomeninges without any lesions on skin, mucous membranes and uvea. Primary CNS malignant melanoma are rare and difficult to diagnose clinically and radiologically and are generally not considered as one of the differentials. CNS Melanomas are rare and aggressive lesions. They are mostly due to metastasis from lesion outside (skin, mucous membrane and Uvea) after Lung and Breast. It is important to differentiate Primary melanocytic melanoma from Metastatic melanoma, because these lesions require a different Patients Pre-operative workup and alternate therapeutic Managements. Primary CNS malignant melanoma only accounts for ~1% of all cases of melanomas [4] and accounts for 0.07-0.17% of all intracranial neoplasms. They generally have Male Predominance and occur mostly in their 5th decade of life. Clinically they present mostly as raised Intracranial Pressure (43%) neurological deficit (35%) and Convulsions or Subarachnoid Hemorrhage (16%).

Clinical Presentations with misleading Radiological evaluations via CT and MR imaging further necessitate to Histopathological analysis of the Specimen taken via Surgical interventions , So that Further in the Course of treatment can be done in the form of Whole Brain Radiotherapy or systemic Chemotherapy. CNS melanoma is reported to be better than that of patients with metastatic disease owning to the Possibility of long term tumor control [3]. Although the life Span of Patients with focal Solitary lesions may be prolonged with Surgery following early diagnosis and further Managements, Generally, Malignant melanomas have Poor Prognosis.

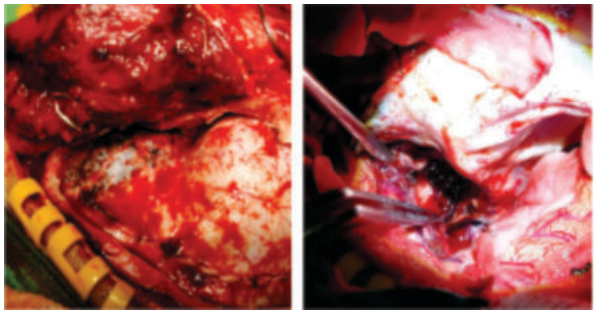

Fig 1. T1 contrast images of the patient brain mri showing Hyperintense lession located in left middle cranial fossa extending upto left sphenoid wing .

Rapid deterioration of vision starting in left eye and gradually involving the Right eye in 2 months. His rest neurological status was intact.

He has no special family History. The patients were thoroughly examined to rule out any lesion in Skin and mucosa with other investigations such as routine Blood Chemical analysis and a complete Blood Count, ECG, CECT Abdomen and chest x-ray were found to be within normal limits.

Ophthalmic evaluations :-

There is Sudden onset rapid deterioration of vision , Started with Blurring of vision first involving the left eye then gradually involved the right eye .Fundus examination shows B/L Papilloedema . Slit Lamp examination is within normal limits.Visual acuity in left eye was 6/60 and right eye was 6/18.

On Imaging:-

NCCT SCAN showing a well-defined extra axial , dural based lesion upon left sphenoid wing .

CEMR: -

A 3.5×3.5× 3.8 cm altered signal intensity extra axial lesion is seen in left middle cranial fossa lying upon left sphenoid wing. Isointense on T1W images with Peripheral Hyperintensity, heterogeneously hyperintense on T2W. No restriction is seen on diffusion weighted \ images mild Heterogenous ewhenmoment is seen on Post content study with enhancing dural tail. Lesion extending to supraseller Cistern and displacing left optic nerve with mild Perilesional edema, meningioma , thrombosed aneurysm and Schwanoma.

A left-

fronto temporal Craniotomy is performed and at dural Opening there was BLACK Patches were seen at Dura further via Trans-sylvian approach a large black vascular firm tumor is seen at left Clinoidal Segment Compressing Both optic Nerves and encasing the ICA of left side. Despite of Tumor being adherent to arachnoid, complete tumor excession was done in Bits and pieces.

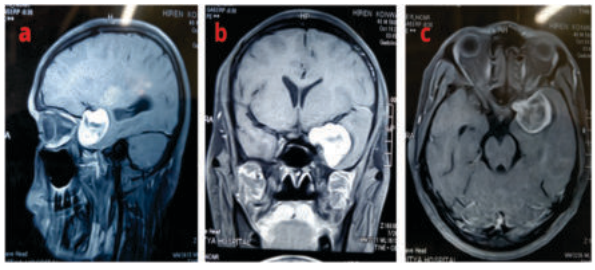

Fig 2. Intraop images of the patient underwent left Pterional

Craniotomy Figure on left showing black pigmentation over dura and on right showing Black clouded tumour while retracting the brain .

CSF analysis :-
Lumber Puncture was done which Showed Raised level of CSF Protein, Rest all values were within Normal limits . There were no metastatic or melanocytic Cells found in the Specimen.

Pathology:-
On Histopathology it showed Malignant Melanoma as shown in the image.

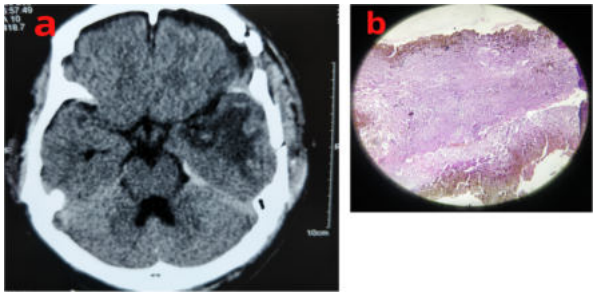

Fig 3 . a) Showing post op CT scan of the patient having GTR of the tuomour. b) Showing Histopathology image of the Specimen Showing melanocytes.

Treatment outcome:-
Patient discharged on Post-op day 4 , with no New Neurological deficit and he was advised for further Radiation therapy as Treatment protocol.

DISCUSSION:-
Pigmented lesions of CNS are rare and it includes Primary melanocytic Lesions of CNS and metastatic melanoma, as well as other CNS neoplasms that may undergo melanization like Schwannana, meningioma medulloblastoma and Some gliomas. CNS melonooytic tumors includes meningeal melanoCytosis, melanomatosis , melonocytoma and melanoma. These lesions originate from melanocytes, Which are Present in the Human Skin, mucous membrane,leptomeninges, Cerebral Parenchyma and uvea. leptomeningeal melanocytes are abundant in Skull base of the cervical Cord [5,6] LANG [7] reported that 90% of the Cases of intracranial metastatic Melanoma investigated were induced by skin melanoma metastases, whereas 2% arose from melanoma in the Ocular region,and in 8%,the original lesion was not identified.

To differentiate whether it's a Primary or metastatic melanoma there are Criteria set forth by SOMERS etal [8] CNS melanoma is likely to be Primary if there is no evidence of melanoma outside the CNS.

In our Patient after thorough dermatological check- up along with CT Chest, CECT-Abdomen no another lesion detected .

Terao et al [9] reported Clinical differences between metastatic CNS melanomas and Primary CNS melanomas Metastatic melanomas are Characterised by:-1).multiple Intracerebral tumors, 2).a rapid and poor Clinical Course due to systemic metastasis and 3). older age group There are few theories regarding melanin deposition in CNS leading to origin of Primary Cerebral melanoma.

Mesodermal theory:-
the mesoderm gives rise to Pigment cells that reach the brain or the Spinal land via the Pial blood vessels.

Ectodermal theory:-
Some epithelial cells produce pigments, and therefore,CNS melanomas are derived from the aberrant embryonic ectodermal cells.

Neurogenic theory:-
The pigment Cells originate from the neural crest, which develop into mesodermal and neural elements.

PIMMs are extremely rare and have a very poor prognosis; these tumors were divided into 2 categories by Gibson et al. in 1957 [10]: diffuse tumors of the leptomeninges and discrete (solitary) tumors. Diffuse meningeal tumors, particularly those involving the skull base or spinal cord, are usually complicated by communicating hydrocephalus.[11]

The clinical characteristics of such melanomas are quite similar to those of intracranial hypertension, cranial nerve deficits, and/or meningism. Otherwise, solitary melanomas occur more frequently in adults and usually mimic leptomeningeal or dura-based lesions [12,13] Some studies suggested that the prognoses of patients with secondary malignant melanomas are mildly worse than those of patients with PIMMs.[14] However, the outcomes of patients with PIMMs with the current treatment strategy remain unsatisfactory. In most studies, total resection, as far as possible, plus postoperative RT seems to be the preferred treatment for eliminating mass effect, improving preoperative symptoms, and achieving a histological diagnosis.[15]

Yamane et al [16] reported 27 cases of primary intracranial melanoma, and the average survival in patients with solitary masses and diffuse-type lesions was 20.7 and 6.7 months, respectively.

Rodriguez et al [17] reported 81 cases of solitary intracranial leptomeningeal melanomas and found that OS was related to lesion site and extent of resection. The mean survival in patients who underwent total removal of their tumor (19.6 months) was significantly longer than that in patients who underwent partial resection or biopsy (9.3 months).

CONCLUSIONS
Gross Total Resection, radiotherapy, and chemotherapy are primary modality of treatment and are used as favorable predictors for improved Overall Survival(OS). PIMMs of the diffuse and solitary types have different clinical characteristics and OS rates. Patients who received GTR plus RT and/or chemotherapy had better outcomes than patients who received other treatment regimens. Immunotherapy and targeted therapy, as developing treatments, have bright prospects for treating PIMM. The present study will provide the beneficial information that will help to optimize clinical evaluations and assist in selecting suitable treatments for patients with PIMM.

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