Choroidal melanoma with isolated brain metastasis— A rare occurance

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Abstract
Choroidal melanoma is the second most common primary malignant melanoma of the body. The most common site for metastasis of choroidal melanoma is liver. Isolated brain metastases in choroidal melanomas is extremely rare. Enucleation is the classic treatment approach to choroidal melanomas and has been the preferred modality of treatment for large and complicated tumors, which compromise visual function and for which other therapies tend to fail. Prognosis of such cases are extremely poor. We report a case of a 47 year old male who presented to us with headache and right sided hemiparesis, and later was diagnosed as a case of choroidal melanoma with isolated brain metastasis.

Keywords: Choroidal melanoma, Enucleation, Tumor, Metastasis.

Introduction
Choroid is present between the retina and sclera. It is a part of the uveal tract. Choroidal melanoma is a most common primary Intra-ocular malignant tumor and is the second common site of malignant melanomas of the body.[1] It is rare in the Indian population. Worldwide its incidence is 0.02–0.06%.[2] In Asian population its incidence is very less as compared to western population. Mostly affected age group are in between 40 and 60 years.

Earlier, diagnosis of choroidal melanoma was challenging and was mostly done on clinical experience. Now a days, different modern diagnostic tools are available like indirect ophthalmoscopy, fundus fluorescein angiography (FFA), A- and B-ultrasonography scans, and trans illumination test for diagnosis. In case of diagnostic dilemma, fine needle aspiration cytology (FNAC) can be done and it should only be considered if therapeutic intervention is required. Till 1970's the rate of diagnosis of choroidal melanomas were only 30%, but after the advent of modern diagnostic techniques in the last three decades diagnosis accuracy has improved dramatically.[3]

Case Report
A 47 year old male presented to us with the complaint of headache since 15 days, which was bifrontal, moderate intensity, more severe in the morning hours. He was having non projectile vomitings since 7 days. He developed progressive weakness of right half of the body over 3 days. There was no history of convulsions, blurring of vision, diplopia, trauma, and weight loss.

On examination vitals were stable. General examination was normal. CNS examination revealed; higher function- patient was drowsy, responding to verbal commands. Pupils were equal in size and bilaterally reacting to light. On motor system examination patient was having right hemiparesis with all deep tendon reflexes exaggerated and right planter extensor. Other system examination were normal. Findings of CT and MRI of brain have been described. HRCT of thorax and CECT abdomen and pelvis were normal.

Considering left orbital mass on neuroimaging, a detailed fundal examination was done which revealed pigmented nodular, dome-shaped, and well-circumscribed mass seen which was confirmed on biopsy as melanoma of choroid. Final diagnosis was made as choroidal melanoma with isolated brain metastasis. Patient was referred to oncologist who advised enucleation with whole brain radiotherapy (WBRT). But patient did not comply for the same and took discharge.

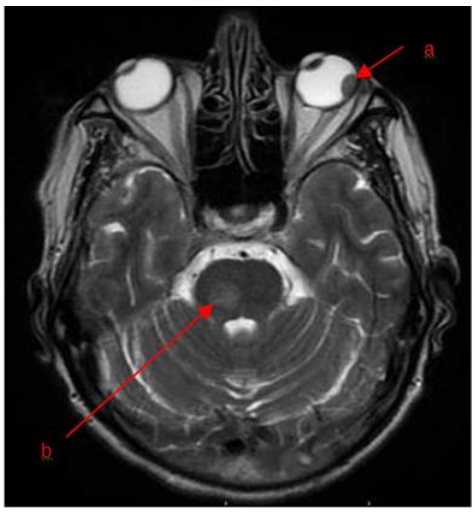

Fig.1a. T_2W MRIBrain showing well defined hypo intense lesion in left orbit; b. T_2W MRI Brain showing well defined hyper intense lesion in the right pons

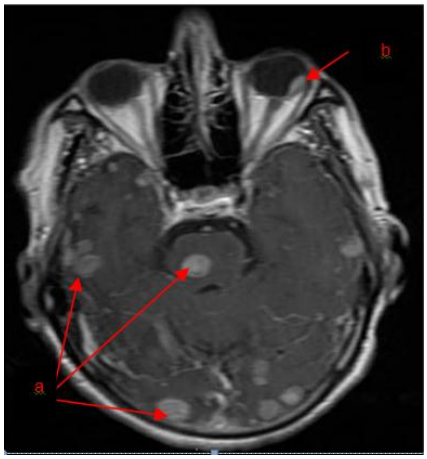

Fig. 2a: Post Contrast T_1W Image showing multiple homogenous enhancing lesion in pons, temporal cortex, and occipital cortex with some dural based lesion.; b. Sameenhasnsing lesion in left orbit

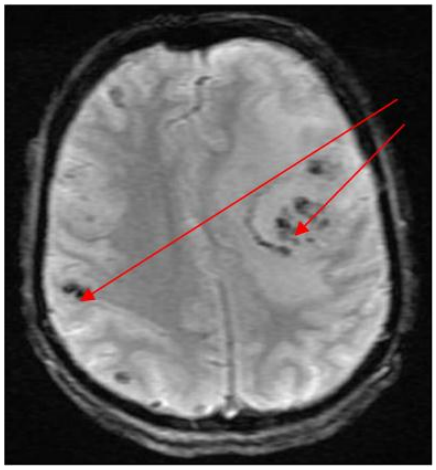

Fig. 3: Gradient echo image showing multiple area of blooming in the lesions as describe above

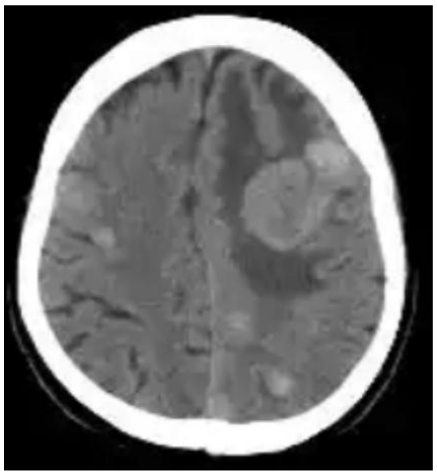

Fig. 4: CT image showing well circumscribed hyper dense lesion with perilesionaledema

Discussion

Choroidal melanoma with metasiasis is a life-threatening disease with limited life expectancy.[4] Usually within 5 years after enucleation, which is the treatment of choice, 40% to 70% of patients develop clinically detectable metastases and die within 6 months. Median survival rate is 2 months for patients not receiving treatment compared to 5.2 months for those receiving treatment for metastatic choroid melanoma.[5] The liver, followed by lungs, bone, and skin, are the most common sites of metastases.

Isolated brain metastases are very rare. Lorigan et al, in their clinical and radiologic review of 110 cases of metastatic choroidal melanoma, found that 5 cases (4%) had brain metastases and all these cases also had concomitant hepatic metastases.[6] Another study in a series of 41 patients with metastatic choroidal melanoma reported only one case with brain metastasis.[7] Enucleation is the classic treatment of choice for choroidal melanoms and it should only be preferred treatment for large and complicated tumors, which compromise visual function and for which other therapies tend to fail.

Survival of choroidal melanoma patients with brain metastases is generally measured in months. It depends on various other prognostic factors including patient age, performance status, and the presence of extracranial metastatic disease.[8,9] Very little progress till date has been done in developing effective treatment for patients with multiple brain metastases because of melanoma. The most commonly administered treatment is whole brain radiotherapy (WBRT). However, the median survival of patients with brain metastases because of melanoma treated with whole brain radiotherapy alone is approximately 3 months.[10] Three phase III trials showed some breakthroughs in the management of brain metastasis. Two out of them showed that resection followed by WBRT yielded better survival than WBRT alone for patients with solitary brain metastases. Additionally, radiosurgery plus WBRT resulted in better survival than WBRT for those with solitary lesions.[11-14] For the majority of patients who present with multiple brain metastases from melanoma, WBRT remains the most commonly administered therapy. WBRT alone results in median survival ranging from 3 to 6 months.[15] Our case falls in the extreme rare category of isolated brain metastasis because no other site was involved in our case.

In conclusion, patients with choroidal melanoma should be followed up for whole of their lives to detect any isolated sign of metastatic disease, and should be treated immediately according to the best clinical protocols available. Isolated brain metastasis is rare. Solitary metasis is treated with resection/ radiosurgery followed by WBRT and WBRT alone remains the mainstay for multiple metastasis to brain from choroidal melanoma.

References

1. Albert DM. Principles and Practice of Ophthalmology. In: Albert, Jakobiec, editors. Philadelphia, PA: W.B. Saunders Co;1994.pp. 3197–8. Ch. 258.
2. Duke‑Elder S, Perkins ES. Diseases of uveal tract in System of Ophthalmology. Vol.9. London: Henry Kimpton;1966. p. 843.
3. Singh P, Singh A. Choroidal melanoma. Oman J Ophthalmol. 2012 Jan-Apr;5(1):3–9. doi:10.4103/0974-620X.94718.
4. Gragoudas ESEgan KMSeddon JM Survival of patients with metastases from uveal melanoma. *Ophthalmology.* 1991;98383-390.
5. Albert DMNiffenegger ASWillson JKV Treatment of metastatic uveal melanoma: review and recommendation. *Surv Ophthalmol.*1992;36429-438.
6. Lorigan JG, Wallace S, Maulight GM. The prevalence and location of metastases from ocular melanoma: imaging study in 110 patients. *Am J Radiol.* 1991;1571279-1281.
7. Char DH. Metastatic choroidal melanoma. *Am J Ophthalmol.*1978;8676-80.
8. Rades D, Bohlen G, Dunst J, et al. Comparison of short-course versus long-course whole-brain radiotherapy in the treatment of brain metastases. Strahlenther Onkol. 2008;184:30–35.
9. Rades D, Bohlen G, Pluemer A, et al. Stereotactic radiosurgery alone versus resection plus whole-brain radiotherapy for 1 or 2 brain metastases in recursive partitioning analysis class 1 and 2 patients. Cancer. 2007;109:2515–2521.
10. Carella RJ, Gelber R, Hendrickson F, et al. Value of radiation therapy in the management of patients with cerebral metastases from malignant melanoma: Radiation Therapy Oncology Group Brain Metastases Study I and II. Cancer. 1980;45:679–683.
11. Patchell RA, Tibbs PA, Regine WF, et al. Postoperative radiotherapy in the treatment of single metastases to the brain: a randomized trial. *JAMA.* 1998;280:1485–1489.
12. Vecht CJ, Haaxma-Reiche H, Noordijk EM, et al. Treatment of single brain metastasis: radiotherapy alone or combined with neurosurgery? *Ann Neurol.* 1993;33:583–590.
13. Mintz AH, Kestle J, Rathbone MP, et al. A randomized trial to assess the efficacy of surgery in addition to radiotherapy in patients with a single cerebral metastasis. Cancer.1996;78:1470–1476.
14. Andrews DW, Scott CB, Sperduto PW, et al. Whole brain radiation therapy with or without stereotactic radiosurgery boost for patients with one to three brain metastases: phase III results of the RTOG 9508 randomised trial. Lancet. 2004;363:1665–1672.
15. Margolin K, Atkins B, Thompson A, et al. Temozolomide and whole brain irradiation in melanoma metastatic to the brain: a phase II trial of the Cytokine Working Group. *J Cancer Res Clin Oncol.* 2002;128:214–218.