LIBYAN INTERNATIONAL JOURNAL OF ONCOLOGY

Case report

Ocular Ultrasonography and Magnetic Resonance Imaging of Orbit in the diagnosis of Uveal Melanoma: A Case Report

Manohar Kachare, Zaid Inamdar*, Manthan Aher, Asmita Raje, Aashish Sachdev

Citation: Kachare M, Inamdar Z, Aher M, Raje A, Sachdev A. Ocular Ultrasonography and Magnetic Resonance Imaging of Orbit in the diagnosis of Uveal Melanoma: A Case Report. Libyan Int J Oncol. 2024;3(1):31-36.

Received: 11-02-2024
Accepted: 12-05-2024
Published: 04-07-2024

Funding*: This research received no external funding.*

Conflicts *of Interest: The authors declare no conflict of interest.*

Department of Radiodiagnosis, Government Medical College, Miraj City- Miraj, India.
***Correspondence**: docinamdarz96@gmail.com

Abstract

Uveal melanoma is a primary intraocular malignancy affecting adults. It originates from the pigmented melanocytes of the uvea present in the choroid, ciliary body and iris. The diagnosis of this tumour was conventionally based on Fundoscopy and B scan Ultrasonography. However, Magnetic Resonance Imaging (MRI) of orbit has become an important radiological imaging modality in recent times. With the advent of newer conservative eye and vision saving surgeries, MRI has assumed a central role in diagnosis as it provides useful information about the local extent of disease which has implications in planning of treatment modality. It also aids in assessment of tumour response to radiotherapy as well as follow up imaging. MRI plays a pivotal role in differentiating uveal melanoma from other benign ocular conditions and intraocular metastasis from other malignancies. Functional MRI sequences like Diffusion weighted imaging and Perfusion weighted imaging are being explored as alternatives to conventional histopathology in patients undergoing conservative therapies. Metastasis occurs in almost 50% cases of uveal melanoma and is associated with poor outcomes. Hence, prompt diagnosis and treatment of disease in early stage is essential to improve prognosis. Thus, a combination of ocular ultrasonography and Magnetic resonance imaging of the orbit has high sensitivity for the diagnosis of uveal melanoma. We report a case of Uveal melanoma diagnosed radiologically by combining the imaging features of ocular B scan Ultrasonography and MRI of Orbit.

Keywords*: Uveal Melanoma; Ocular Ultrasonography; Magnetic Resonance Imaging.*

Introduction

Although uveal melanoma is a rare tumour, it is described as the most common primary intraocular malignancy in adults. [1] It has an estimated incidence of 6 cases per million per year. [2] The incidence in Asian population is reported at 0.2-0.4 cases per million per year. Majority of these tumours originate from the choroid (around 85 %), whereas the remainder arise from the ciliary body or iris.[3] The conventional imaging modality used for radiological assessment of uveal melanoma is ocular B Scan ultrasonography. However, Magnetic resonance imaging (MRI) of the orbit has emerged as a promising imaging modality in recent years. Conventionally, enucleation was described as the mainstay of treatment in cases of uveal melanoma. Newer treatment modalities have been introduced recently which focus on vision conserving therapy. MRI orbit, due to its excellent spatial resolution and soft tissue contrast has thus become an important investigation in determining tumour extent and extraocular spread of tumour which in turn are important factors in choice of treatment. Metastasis is a part of the natural history of the disease in more than half of the cases of uveal melanoma and is associated with poor outcomes due to lack of systemic therapy. Most common site for extra ocular spread is the liver.
This case report aims at combining the imaging features of ocular B scan Ultrasonography and MRI of Orbit for comprehensive radiological diagnosis of Uveal melanoma.

Case Report

A 53-year-old male patient presented to the ophthalmology out-patient department with a 6-month history of gradual onset of decreased vision in left eye. The patient also experienced

pain along medial canthus of left eye since past 1 month. The patient had no h/o trauma or ocular infection and gave no history of associated comorbidities like hypertension or Diabetes mellitus. Patient had undergone small incision cataract surgery (SICS) in left eye 1 year ago. Ophthalmic examination revealed loss of perception of light in left eye. Fundoscopy examination was not possible due to opaque media.

The patient was referred to the Radiology department for further evaluation. Ocular B Scan Ultrasonography was performed using 7.5 MHz high frequency linear transducer. The ultrasound revealed an isoechoic solid mass lesion of approximate size 20 x 15 mm arising from the uveal tissue along medial aspect of left eyeball (Fig 1). V-shaped flap of retina was noted with apex at optic disc, associated with mild hypoechoic subretinal fluid collection suggestive of exudative retinal detachment. (Fig 2). The lesion showed mild vascularity on Colour Doppler imaging. Additionally, vitreous chamber showed presence of dense internal echoes consistent with vitreous haemorrhage.

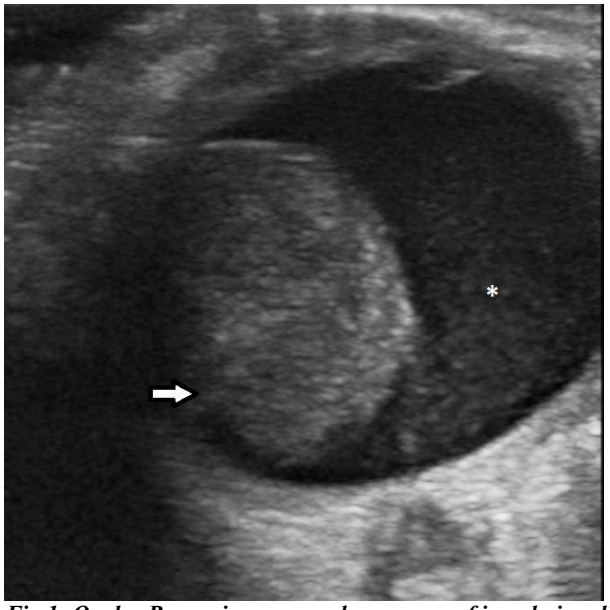

Fig 1: Ocular B scan image reveals presence of isoechoic solid mass lesion along medial aspect of left eyeball in subretinal space (white arrow). Also note the presence of dense echoes in vitreous chamber, suggestive of vitreous haemorrhage (*).

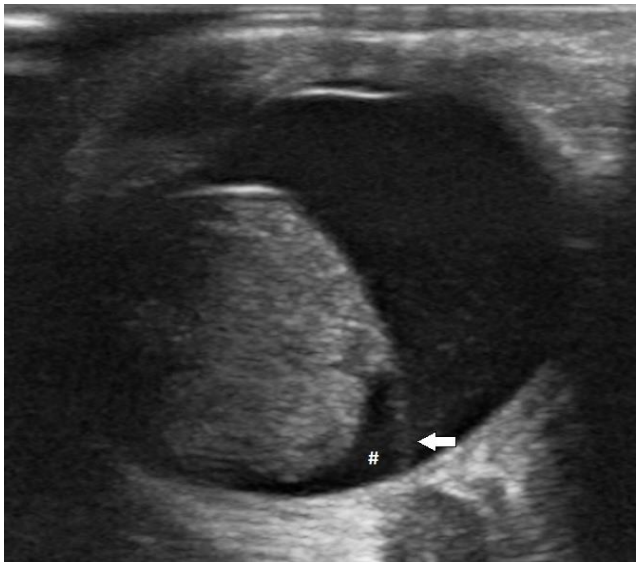

Fig 2: The lesion is raising V-shaped retinal flap with apex at optic disc (white arrow). Mild subretinal fluid is noted around the lesion (#). Findings are consistent with exudative retinal detachment.

The patient was then subjected to an MRI Orbit (Plain + Contrast) scan. The non-contrast sequences showed following imaging findings: A well-defined intraocular, intraconal solid mass lesion of approximate size 18 x 15 mm was noted along medial aspect of left eyeball in subretinal location. The lesion appeared hypointense on T2 weighted images (Fig 3) and iso to hyperintense on T1 weighted images. The lesion showed typical mushroom configuration with thin stalk projecting medially on coronal T2 weighted images (Fig 4).

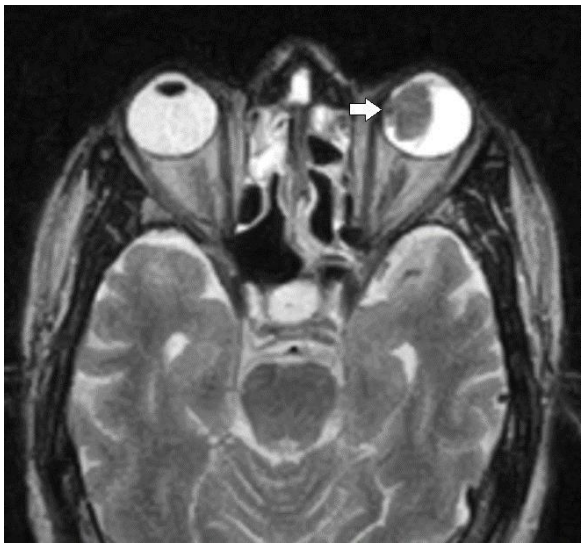

Fig 3: Axial T2 weighted image reveals a hypointense intraocular solid lesion along medial aspect of left eyeball in subretinal space (white arrow). Note the retinal flap attached at the optic disc with T2 hyperintense subretinal fluid around the lesion.

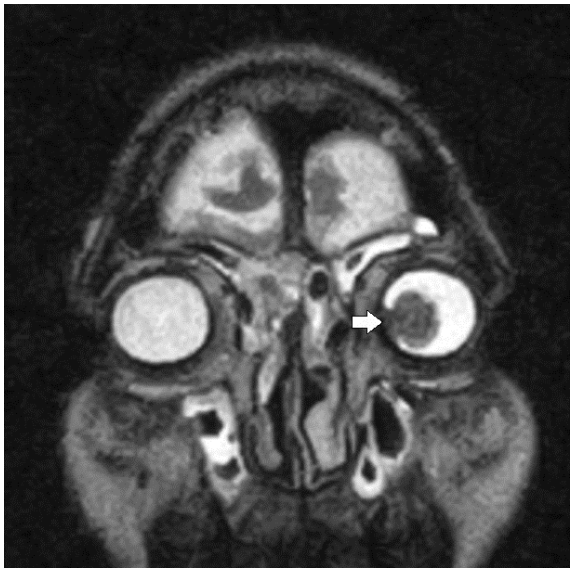

Fig 4: Coronal T2 weighted image shows the typical 'mushroom' configuration of tumour with stalk-like projection medially (white arrow)

Associated findings were the presence of exudative retinal detachment along with T1 hyperintense collection noted within vitreous chamber suggestive of vitreous haemorrhage (Fig 5). Post contrast images were obtained after injection of I.V gadolinium contrast media. The lesion revealed avid homogenous enhancement on post contrast T1 weighted images (Fig 6). There was no evidence of extrascleral spread of the lesion or infiltration of the extraocular muscles.

These imaging features point towards a radiological diagnosis of Uveal Melanoma without extrascleral/extraocular spread of disease.

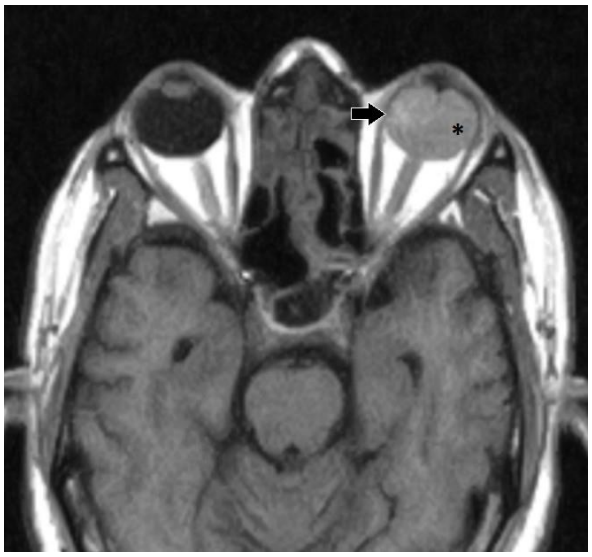

Fig 5: Axial T1 weighted image shows iso to hyperintense lesion (black arrow). Also note the presence of T1 hyperintense collection s/o vitreous haemorrhage (*)

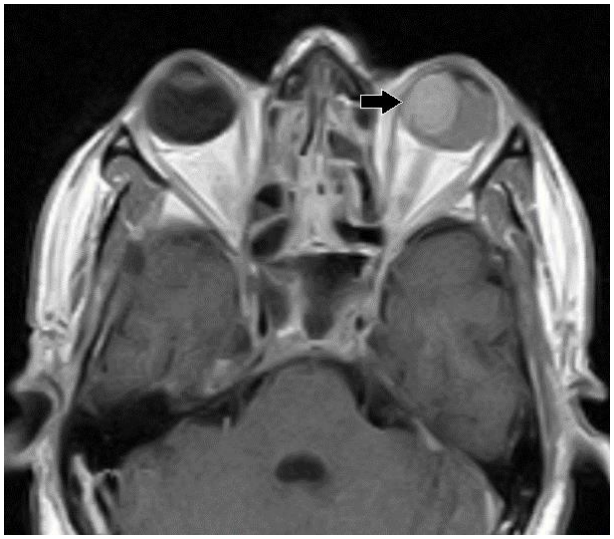

Fig 6: Post i.v. gadolinium contrast axial T1 weighted image shows avid homogenous enhancement of tumour (black arrow)

Discussion

Uveal melanoma is a malignant intraocular tumour occurring in adults with a median age at presentation being 62 years.[4] The average age of incidence is almost a decade earlier among Asian populations. It predominantly affects white males.[5] The tumour arises from the pigmented melanocytes of the uvea and involves the choroid, ciliary body and iris, in decreasing order of incidence.[6]

The pathogenesis involves genetic and molecular alterations. The earliest event in the disease is said to be the Inactivation of the Rb (retinoblastoma) tumour suppressor pathway.[7] Overexpression of Bcl-2 and inhibition of p53 tumour suppressor oncogene are also implicated in the molecular pathogenesis of uveal melanoma.[8]

Clinical features depend on size and location of the tumour. The most common clinical feature of uveal melanoma includes decrease in visual acuity. Occasionally it may be associated with dark spots in visual field, photopsia, metamorphopsia, floaters, pain etc. [9] Smaller lesions may be asymptomatic and are detected incidentally at fundoscopy. Fundoscopy features of choroidal melanoma include a mass deep to the retina without retinal feeder vessels. Occasionally vitreous haemorrhage may be present which obscures tumour visualization on fundoscopy. In such cases the tumor is only detected on ocular B scan USG or MRI.[10]

Choroidal melanomas may be pigmented (55%), non-pigmented (15%) or mixed. The tumour configuration may be dome shaped (predominantly), Mushroom shaped or diffuse /

flat. The mushroom shaped lesion invades through the Bruch's membrane entering into subretinal space.[11] Ciliary body melanomas are less common and present with prominent episcleral/sentinel vessels. Larger lesions may lead to lens displacement with resultant refractive errors/astigmatism. Iris melanomas are rare and occur in background of pre-existing naevi. Complications such as hyphaema may occur.[12]

Three pathological subtypes of uveal melanoma are identified on histopathology namely spindle shaped, epitheloid type and mixed melanoma.[13] The spindle shaped melanoma subtype is associated with the best prognosis. However, histopathological examination of biopsy specimen may not always be representative as the Uveal melanoma is a heterogenous tumour in terms of chromosomal aberrations.

The tumour prognosis is related to tumour size and location, extraocular extension, histopathology features and distant metastasis. Distant metastasis at initial presentation is rare, however around 50 % of patients develop metastasis to distant organs, most commonly involved site for metastasis being the liver. [14] Extrahepatic metastasis is found in lungs, bone, skin and lymph nodes.[15]

Enucleation of the affected eye was considered as the surgical treatment of choice conventionally. However, in the past few years there has been a growing emphasis on globe sparing and vision conserving therapies in uveal melanoma leading to increased quality of life in many patients. The local tumor control strategies include: endoresection and exoresection followed by post operative ruthenium plaque radiotherapy.[16] Locoregional therapy for liver metastasis involves surgical metastasectomy, Trans arterial chemo embolization (TACE) and isolated hepatic perfusion.[17],[18]

Ocular B scan ultrasonography remains the first line radiological investigation in suspected cases. It is also useful in assessment of morphological features like size and shape of tumour and detection of vascular pulsations within the mass which differentiates it from vitreous haemorrhage. It is particularly useful in the event of opaque media where the tumour cannot be visualized on fundoscopy. [19]

The role of MRI in uveal melanoma encompasses various domains. Characterization of tumor origin, evaluating the local extent of disease, assessment of tumour response to radiotherapy as well as follow up imaging. Due to its excellent soft tissue contrast, MR imaging shows high sensitivity (100%) and specificity (89%) in the diagnosis of extrascleral extension. [20] This is an important consideration in patients undergoing conservative tumor management. MR appearance of melanoma depends mainly on the melanin content of the tumour as melanin has paramagnetic effect with T1 and T2 shortening. [21] The typical imaging appearance of uveal melanoma is described as high signal intensity (hyperintense) on T1Wi and low signal intensity (hypointense) on T2Wi with avid enhancement post gadolinium contrast injection.[22] Poorly pigmented or amelanotic tumours may show intermediate signal intensity on both T1 and T2 weighted images.[23] Being a malignant lesion, uveal melanoma exhibits high cellularity and shows restricted diffusion which is indicated by high signal intensity on Diffusion weighted images and low signal intensity on corresponding ADC maps. [24] Perfusion weighted imaging (PWI) is being explored as a potential alternative to histopathology in patients who undergo conservative therapies.

MRI also plays a crucial role in excluding the common differential diagnosis of uveal melanoma The most common differentials include: choroidal nevus, choroidal haemangioma, melanocytoma, uveal metastases and choroidal detachment.[25] Intraocular metastases occur most commonly from breast and bronchial carcinomas.

Conclusion

This case report emphasizes the importance of multimodality approach in the diagnosis of uveal melanoma. Ocular B scan ultrasonography remains the first line imaging modality during initial assessment. Owing to its wide range of utility, MRI orbit has assumed a pivotal role in uveal melanoma recently. With the availability of newer conservative therapies, MRI serves as an important tool not only in diagnosis but also in planning of management approach, due to its multiplanar and multiparametric capabilities. Additionally, it helps in assessing tumour response to radiotherapy as well as follow up imaging.

Conflict of interest

The authors do not report any financial or personal connections with other persons or organizations which may negatively affect the contents of this publication and/or claim authorship rights to this publication.

Acknowledgements
None.

Funding
The research received no external funding.

References

1. Jovanovic P, Mihajlovic M, Djordjevic-Jocic J, Vlajkovic S, Cekic S, Stefanovic V (2013). Ocular melanoma: an overiew of the current status. Int J Clin Exp Pathol 6(7):1230–1244
2. Chattopadhyay C, Kim DW, Gombos DS et al (2016). Uveal melanoma: from diagnosis to treatment and the science in between. Cancer 122(15):2299–2312.
3. Grech Fonk L, Ferreira TA, Webb AG, Luyten GPM, Beenakker JM (2020). The economic value of MR-imaging for uveal melanoma. Clin Ophthalmol 14:1135–1143.
4. Andreoli MT, Mieler WF, Leiderman YI (2015). Epidemiological trends in uveal melanoma. Br J Ophthalmol 99(11):1550–1553.
5. Virgili G, Gatta G, Ciccolallo L, et al. Incidence of uveal melanoma in Europe. Ophthalmology. 2007; 114:2309–15.
6. Damato B. Treatment of primary intraocular melanoma. Expert Rev Anticancer Ther. 2006; 6:493–506.
7. Landerville S, Agapova OA, Harbour JW. Emerging insights into the molecular pathogenesis of uveal melanoma. Future Oncol. 2008; 4:629–36.
8. Triozzi PL, Eng C, Singh AD. Targeted therapy for uveal melanoma. Cancer Treat Rev. 2008; 34:247–58.
9. Kaliki S, Shields CL (2017). Uveal melanoma: relatively rare but deadly cancer. Eye 31(2):241–257.
10. Shields CL, Manalac J, Das C, Ferguson K, Shields JA. Choroidal melanoma: clinical features, classification, and top 10 pseudomelanomas. Curr Opin Ophthalmol. 2014; 25:177–85.
11. Tailor TD, Gupta D, Dalley RW, Keene CD, Anzai Y (2013) Orbital neoplasms in adults: clinical, radiologic, and pathologic review. Radiographics 33(6):1739–1758.
12. Shields CL, Shields JA, Materin M, Gershenbaum E, Singh AD, Smith A. Iris melanoma: risk factors for metastasis in 169 consecutive patients. Ophthalmology. 2001; 108:172–8.
13. Kivela T, Simpson E.R, Grossniklaus H.E, Jager M.J, Sough, A.D, Caminal J.M. Uveal melanoma. In AJCC Cancer Staging Manual, 8th ed.; Amin, E.B., Edge, S., Greene, F., Byrd, D.R., Brookland, K.R., Washington, M.K., Eds.; Springer: New York, NY, USA; pp. 805–817.
14. Gragoudas ES, Egan KM, Seddon JM, et al. Survival of patients with metastases from uveal melanoma. Ophthalmology. 1991; 98:383–9
15. Bedikian AY. Metastatic uveal melanoma therapy: current options. Int Ophthalmol Clin. 2006; 46:151–66.
16. Konstantinidis L, Groenewald C, Coupland SE, Damato B. Long-term outcome of primary endoresection of choroidal melanoma. Br J Ophthalmol. 2014; 98:82–5.
17. Rivoire M, Kodjikian L, Baldo S, Kaemmerlen P, Negrier S, Grange JD. Treatment of liver metastases from uveal melanoma. Ann Surg Oncol. 2005; 12:422–8.
18. Agarwala SS, Eggermont AMM, O'Day S, Zager JS. Metastatic melanoma to the liver. Cancer. 2014; 120:781–9.
19. Tarlan B, Kiratli H (2016) Uveal melanoma: current trends in diagnosis and management. Turk J Ophthalmol 46(3):123–137.
20. Mahajan A, Crum A, Johnson MH, Materin MA (2011) Ocular neoplastic disease. Semin Ultrasound CT MR 32(1):28–37
21. Lemke AJ, Hosten N, Wiegel T et al (2001) Intraocular metastases: differential diagnosis from uveal melanomas with high-resolution MRI using a surface coil. Eur Radiol 11(12):2593–2601
22. Tailor TD, Gupta D, Dalley RW, Keene CD, Anzai Y (2013) Orbital neoplasms in adults: clinical, radiologic, and pathologic review. Radiographics 33(6):1739–1758.
23. Purohit BS, Vargas MI, Ailianou A et al (2016) Orbital tumours and tumour like lesions: exploring the armamentarium of multiparametric imaging. Insights Imaging 7(1):43–68.
24. Sepahdari AR, Kapur R, Aakalu VK, Villablanca JP, Mafee MF (2012) Diffusion-weighted imaging of malignant ocular masses: initial results and directions for further study. AJNR Am J Neuroradiol 33(2):314–319.
25. Keraliya AR, Krajewski KM, Braschi-Amirfarzan M et al (2015) Extracutaneous melanomas: a primer for the radiologist. Insights Imaging 6(6):707717.