

Orbital cavernous venous malformation: Clinical features and role of 3T magnetic resonance imaging

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Abstract

Background: Orbital cavernous malformation is venous type of malformation. They are more common in middle-aged women. Cavernous malformation having slow progressive growth, painless proptosis is most common clinical presenting. Less common symptoms include pain, lid swelling, diplopia, lump, and recurrent obscured vision. MRI is a non-invasive, nonoperator dependent effective imaging modality and plays a crucial role in early detection, localisation, characterization, intracranial and extra orbital extension, morphologic changes in bony orbit and to rule out complications of cavernous malformations. MRI is the imaging modality of choice for the evaluation of the cavernous haemangioma. The purpose of our study was to analyse imaging features of cavernous haemangioma in detail and correlate it with clinical features and histopathology. **Materials and Methods:** This study was performed in the department of Radio diagnosis of a tertiary health care institute. 16 histopathology proven patients of orbital cavernous malformation referred from ophthalmology department from January 2017 to November 2019 were included in this study. There were 9 women and 7 men with mean age of 38.75 yrs. All of them were evaluated with detailed clinical history and subsequently with dedicated imaging of orbit using 3 T MRI 32 channel Siemens MRI magnetom verio machine. MR Sequences used were T1 axial and coronal without and with fat Sat, T2 and STIR axial and coronal, T1 post contrast in 3 standard planes and dynamic 3D post contrast scans **Results:** In our study, cavernous haemangioma is more common in middle aged women (56%), most common symptom being glove protrusion (81%), most common sign is painless proptosis (87 %), most common location was intraconal (58%) followed by extraconal (37%) , 94 % lesions were unicompartmental, extra orbital extension is seen in 18 % and no intracranial extension seen. Almost all the lesions (34 %) were isointense on T1WI, hyperintense on T2WI, showed sequential enhancement on T1 post contrast and dynamic contrast studies. **Conclusion:** MRI is an excellent non-invasive modality with high level of accuracy in diagnosis, localisation and characterisation of cavernous haemangiomas. It is an accurate clinical screening tool to guide surgeon for operative management.

Key Words: Diagnostic accuracy, Magnetic resonance imaging, cavernous haemangiomas.

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Received Date: 09/10/2019 Revised Date: 11/11/2019 Accepted Date: 21/12/2019

DOI: <https://doi.org/10.26611/10131326>

Access this article online

Quick Response Code:



Website:

www.medpulse.in

Accessed Date:
05 February 2020

INTRODUCTION

Orbital cavernous venous malformation is also known as cavernous haemangioma. Cavernous haemangioma is the most common vascular lesion of the orbit in adults³. Cavernous haemangiomas are well circumscribed vascular masses which commonly occur in middle-aged females. The majority occurs within the intraconal compartment of the orbit most commonly lateral to the optic nerve and presents with slowly progressive and painless proptosis^{1,2}. Magnetic resonance imaging plays a crucial role in the localisation, characterization and

How to cite this article: Rathi Varsha, Mate Shriram B, Desai Vidya V. Orbital cavernous venous malformation: Clinical features and role of 3T magnetic resonance imaging. *MedPulse – International Journal of Radiology*. February 2020; 13(2): 52-56.

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differentiation of cavernous haemangiomas from other tumours, especially with dynamic MR imaging. The contrast enhancement pattern on dynamic contrast-enhanced MR imaging is the most sensitive imaging criteria⁴.

AIMS AND OBJECTIVES

Based on the superior soft tissue characteristics provided by MRI compared to CT, we aimed to analyse the MRI findings in cavernous haemangiomas. We aimed to analyse the efficacy of MRI in characterization, localization and extent of orbital cavernous haemangioma.

MATERIALS AND METHODS

This study was performed in the department of Radiodiagnosis in a tertiary health care institute including 16 histopathology proven patients referred from ophthalmology department with history of proptosis and vision loss over a period of twenty-four months. Both male and female patients of age group 1 to 80 years were included in this study. Patients with any absolute contraindication for MRI like metal implants and pacemakers were excluded from the study. The study group patients were evaluated with detailed clinical examinations and subjected subsequently with MRI of orbit using dedicated head coil on 3T Siemens Verio MRI scanner in a 32 channel head coil. Routine dedicated orbit protocol was done using T1W, T2W with and without fat suppressed axial, coronal and sagittal images, STIR coronal, SWI, DWI, ADC, T1W post contrast in axial, coronal and sagittal planes, dynamic 3D angiography following gadolinium-based contrast media.

RESULTS

Age and sex distribution of patients: Out of 16 patients, there were 9 females and 7 males in this study. Female patients exceeded the number of male patients in all the age groups. The cavernous haemangioma most commonly found in age group of 31-60 years.

Side of involvement: in our study right eye (56 % - 9 patients) is affected more commonly than left eye (46% - 7 patients).

Symptoms: the most common symptom in our study is globe protrusion (81 % - 13 patients), followed by diminution of vision (31.25 %- 5 patients), lid swelling (25 % -4 patients), restriction of extraocular muscles motion (12 % - 2 patients), diplopia (18%- 3 patients), pain (12 %).

SYMPTOMS	No	%
Globe Protrusion	13	81.25%
Diminution of Vision	5	31.25%
lid swelling	4	25.00%
Restriction Of extraocular muscle motion	2	12.50%
Diplopia	3	18.75%
Pain	2	12.50%

Signs: In our study we have incorporated two major signs of cavernous haemangioma i.e. painless and painful proptosis. Painless proptosis was most common sign and it is present in the 87 % patients (14 cases). Painful proptosis was seen in 13 % patients (2 cases).

Signs	No.	Percentage
Painless Proptosis	14	87%
Painful Proptosis	2	13%

Duration of symptoms: the average duration of symptoms of patients of cavernous haemangioma who participated in our study was ranging from 2 months to 4 years. But most commonly duration of symptom was 1-10 months.

Location of the lesion

Conventionally the orbit is divided into intraconal, conal and extraconal compartments⁵ by myofascial cone. In our study the intraconal compartment was most commonly involved (69%- 11 cases), followed by extraconal compartment (16% - 7 cases) and conal compartment (6%- 1 case).

LOCATION	NO.	PERCENTAGE
INTRACONAL	11	69%
CONAL	1	6%
EXTRACONAL	7	16%

The distribution of lesion in vertical and horizontal axis in orbit is divided into superior, inferior, nasal and temporal side⁵. In our study nasal location was most common (38%), followed by superior (31 %), temporal (25%), and inferior (25%).

LOCATION	NO.	PERCENTAGE
Nasal	6	38%
Temporal	4	25%

In our study cavernous hemangioma was more common on nasal side (38%) than temporal side (25 %). In our study the orbital cavernous haemangioma is most commonly uni-compartmental (94 %), two compartment involvement is seen in the 6 % cases.

Extensions of the lesions outside the orbit: In our study the 19 % cavernous haemangiomas (3 cases) showed extra orbital extension and no intracranial or intracanalicular extension seen.

Magnetic resonance imaging characteristics:

Sr no	Imaging features		Number of cases	Percentage
1.	Homogeneity	Homogenous	12	75%
		Heterogenous	4	25%
2.	T1 and T2 signal characteristics			
	T1WI	Isointense	15	94%
		Hyperintense	1	6%
	T2WI	Hyperintense	15	94%
		Hypointense	1	6%
3.	Post contrast studies			
	T1 post contrast sequential scans	Present	14	88%
		Absent	2	12%
	Dynamic 3D post contrast scan	Present	14	88%
		Absent	2	12%
4.	Thrombosis / areas of bleed (areas of T1 hyperintensities)		7	44%
5.	Internal septations		5	31%
6.	Pseudocapsule		10	63%
7.	Deformation of globe by lesion		3	19%
8.	Deformation of lesion by globe		10	81%
9.	Changes in bony orbit		7	44%
10.	Borders	Regular	14	88%
		Irregular	2	12%

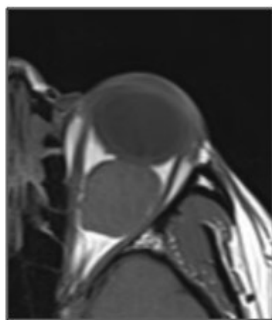


Figure 1

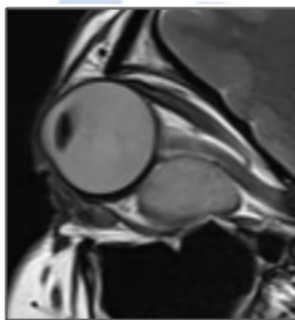


Figure 2

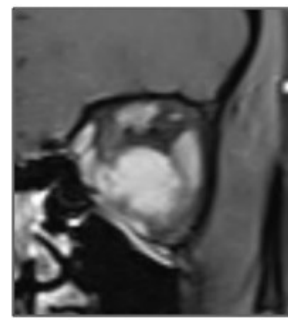


Figure 3



Figure 4

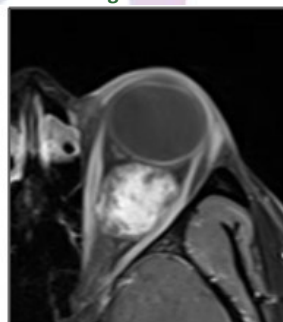


Figure 5

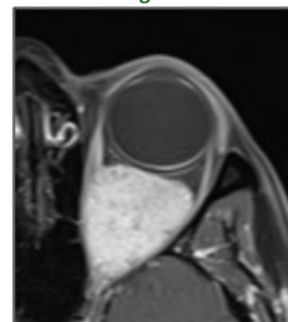


Figure 6

Figure 1: Axial T1W non-fat suppressed image showing isointense intraconal retrobulbar lesion; **Figure 2:** Oblique sagittal T2W image showing intraconal retrobulbar oval lesion with distinct hypointense pseudocapsule; **Figure 3:** Coronal T1W fat suppressed image showing isointense lesion in temporal aspect of intraconal space with medial displacement of the optic nerve; **Figure 4, 5, 6:** T1W post contrast fat suppressed images showing progressive filling in of contrast within the lesion

DISCUSSION

Advent of MR imaging and its increasing use in orbital imaging has revolutionized the diagnosis of pathologies of orbital tumours. Many studies have been published on sensitivity and specificity of MRI in characterization of

orbital pathologies and imaging features compared with clinical and USG b scan findings. These have suggested MR to be an effective tool for evaluation of orbital tumours. This study included 16 patients with histopathologically proven cases of cavernous

haemangioma. The study group constituted of 9 women and 7 men with mean age 38.5 yrs. We analysed the orbital MR imaging findings of 16 cases of cavernous haemangiomas with respect to following parameters: anatomic extent (compartments of orbit), margins, internal features, presence or absence internal haemorrhage or thrombosis, presence or absence pseudocapsule, internal septations, mass effect caused by lesion on globe and bony orbit and contrast enhancement pattern discussed below. We also analysed clinical features, age and gender distribution of the cavernous haemangioma.

Age and sex distribution:

In our study orbital cavernous haemangioma affected middle aged females most commonly (56%) with a mean age of 38-40 years and range of 3 to 80 years. In study conducted by *Ruchman et al*⁶, nine were women, four were men. The average age was 43, range 21-67.

Side of involvement:

In our study cavernous haemangiomas are found 56% on right side and 44 % on left side. In study conducted by *Ruchman et al*⁶ Seven had right-sided involvement, six had left.

Signs and symptoms:

In our study most common symptom was globe protrusion (81 % - 13 patients), followed by diminution of vision (31.25 %- 5 patients), lid swelling (25 % -4 patients), restriction of extraocular muscles motion (12 % - 2 patients), diplopia (18%- 3 patients), pain (12 %). The most common sign was painless proptosis (87%). According to *Sameer A. Ansari et al*² the most common presenting sign and symptom is painless proptosis (mean: 5 – 6 mm; range: 0 – 15 mm). According to *Xian J et al*⁷ et al the most common sign was painless proptosis.

Anatomic extent:

In our study the most common location of cavernous haemangioma is intraconal compartment 69% patients (11 cases), followed by extraconal compartment (16% - 7 cases) and intraconal compartment (6%- 1 case). In our study cavernous hemangioma was more common in nasal side (38 %) than temporal side (25 %). In study conducted by *Xian J et al*⁷, Most orbital cavernous haemangiomas are identified in the intraconal space, predominantly in the lateral aspect of the intraconal space. In their series, only 8 of the 43 orbital cavernous haemangiomas were in the extraconal space. Valuation of Four-Quadrant Location Method in Diagnosis and Differential Diagnosis study of *Akter GS et al*⁸, most of the cavernous haemangioma was found between the optic nerve and extraocular muscles with temporal aspect of the intraconal space. In our study among the vertical axis the cavernous haemangioma was more commonly found in superior location (31%) than inferior one (25 %). In our

study 16 % cases showed extra orbital extension. No intracranial extension was found.

Changes produced by cavernous haemangioma in globe and bony orbit:

In our study expansion of the bony orbit is seen in 44 % patients (7 cases). Indentation of globe by cavernous haemangioma is seen in 16 % patients (in 3 cases). In our study globe has indented the cavernous haemangioma in 81 % (in 13 cases). Cavernous haemangiomas usually round or oval in cross-section and although frequently about the globe, they do not deform it, but rather are deformed by the globe, on account of their soft consistency^{9,10}.

MR imaging characteristics:

Homogeneity:

In our study 75 % lesions were homogenous and 25 % were heterogenous. Study conducted by *Xian J et al*⁷ showed that 78-85 % lesions were homogenous and 9-13 % lesions were heterogenous. According to *Sameer A. Ansari et al*², On MR imaging, orbital cavernous haemangiomas are homogeneous and well-defined intraconal lesions.

T1 and T2 signal intensities:

In our study 94 % lesions were T1 isointense and T2 hyperintense. 6 % lesions were T1 hyperintense and T2 intermediate to hypointense. Cavernous haemangioma study conducted by *Xian J et al*⁷ showed that 88 % lesions were isointense on T1 and 11 % lesions were hypointense on T1. 91 % patients were hyperintense on and 8.5 % lesions were hypointense on T2 WI.

Contrast enhancement characteristic:

In our study T1W post contrast fat suppressed sequential images showed gradual enhancement in 94 % patients (15 cases). Similar enhancement is seen in dynamic post contrast 3D sequence. In study conducted by *Xian J et al*⁷, 100 % lesions showed gradual enhancement on dynamic scans.

Pseudocapsule and septae:

In our study 63% (10 cases) patients showed T2W hypointense pseudocapsule and internal septations in 31% (in 5 cases). According to *Smoker et al*¹, Thin, regular circumferential, hypointense line at the periphery of the tumour may also be observed, corresponding to the fibrous pseudocapsule surrounding the lesion and internal septa may be visible within larger lesions.

CONCLUSION

MRI is an excellent, non-invasive, radiation free precise imaging modality with multiplanar capabilities and excellent soft tissue delineation. Appropriate sequences and analysis of images in all three planes helps to accurately detect, localise, characterise and differentiate the orbital cavernous haemangioma from other tumours,

thereby guiding further management of patient. Hence MRI should be the initial investigation of choice in the evaluation of all cases of cavernous haemangiomas.

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Source of Support: None Declared
Conflict of Interest: None Declared

