AMERICAN JOURNAL OF DIAGNOSTIC IMAGING, 2022
VOL 8, NO. 2, PAGE 34–40
10.5455/ajdi.20210208052003

ORIGINAL ARTICLE

Open Access

Lymphoplasmacyte-rich meningioma of orbit mimicking lymphoma—An unusual variant in atypical site

Babu Peter Sathyanathan[1], Aishwarya Ravichandran[2], Thangameena Subramaniam[3]

ABSTRACT

Optic nerve sheath meningiomas are benign tumors from the arachnoid cap cells of the optic nerve sheath. They represent 20% of all orbital meningiomas, the majority of which are direct extensions from intracranial meningiomas. Clinical presentation is with a combination of visual loss and proptosis. A number of meningioma subtypes have been proposed through the years, and the current (2021) World Health Organization (WHO) classification includes fif15 named entities. We report an extremely rare case of Lymphoplasmacyte rich meningioma of orbit, belonging to Grade I WHO classification, hitherto unreported in literature and mimicking lymphoma.

ARTICLE HISTORY

Received February 08, 2021
Accepted January 27, 2022
Published February 06, 2022

KEYWORDS

Lymphoplasmacyte rich meningioma; optic nerve sheath meningioma; Magnetic Resonance Imaging (MRI); optic nerve; orbital lymphoma

Case Report

A sixty65-year-old male patient presented with complaints of proptosis of the left eye for 3 months, a headache of insidious onset and painless progressive loss of vision in the left. On inspection, proptosis of the left eye was seen with eyelid retraction causing facial asymmetry. Conjunctiva appeared dry and mildly inflamed. No conjunctival chemosis was seen. There was no bruit or tenderness on palpation.

Plain computed tomography (CT) of orbit revealed a hyperdense lesion in the left orbit, which filled the intraconal space, showing loss of fat plane with extraocular muscles, encasing the optic nerve and causing proptosis (Figs. 1 and 2).

Magnetic Resonance Imaging (MRI) study of the orbit was done in a 3 Tesla MRI (SKYRA, SIEMENS HEALTHINEERS, ERLANGEN GERMANY). The study showed a T2/ Fluid Attenuation Inversion Recovery (FLAIR) hypointense lesion in the left intraconal compartment with loss of fat plane with all extraocular muscles. The lesion surrounded the optic nerve with minimal thickening of the nerve. There was widening of the optic canal and extension of the lesion posteriorly into the pre-chiasmatic region (Figs. 3–5). In T1 weighted images (T1WI), the lesion appeared isointense to grey matter (Fig. 6). The lesion showed restricted diffusion with low Apparent Diffusion Coefficient (ADC) values (Figs. 7 and 8). In contrast images, the lesion showed homogenous enhancement (Figs. 9 and 10). Based on the above imaging findings, a diagnosis of intra-orbital meningioma was made.

Surgical excision was done, and the grayish-brown soft tissue fragments were sent for histopathological examination (Fig. 11). The section studied showed fragments of neoplasm arranged in cords and whorls. The cells were round to oval with dark staining nuclei and scant to moderate eosinophilic cytoplasm. Tumor cells were seen infiltrating around muscle fibers. Some foci showed diffuse infiltration by lympho-plasmacytoid cells with intervening adipocytes and small blood vessels. Few nerve bundles and many hyalinized, congested blood vessels were also seen. The histopathological picture was in favor of Lympho-plasmacyte rich meningioma (Figs. 12 and 13).

Discussion

INTRODUCTION

Nath meningioma (ONSM) represents the second most common tumor to arise from the optic nerve sheath complex after optic glioma [1]. Orbital meningioma falls into two broad categories—primary & secondary. Primary lesions derive commonly from cells lining the intra-ocular segments

Contact Aishwarya Ravichandran aishuravi1292@gmail.com

© 2022 The Authors. This is an open access article under the terms of the Creative Commons Attribution NonCommercial ShareAlike 4.0 (https://creativecommons.org/licenses/by-nc-sa/4.0/).

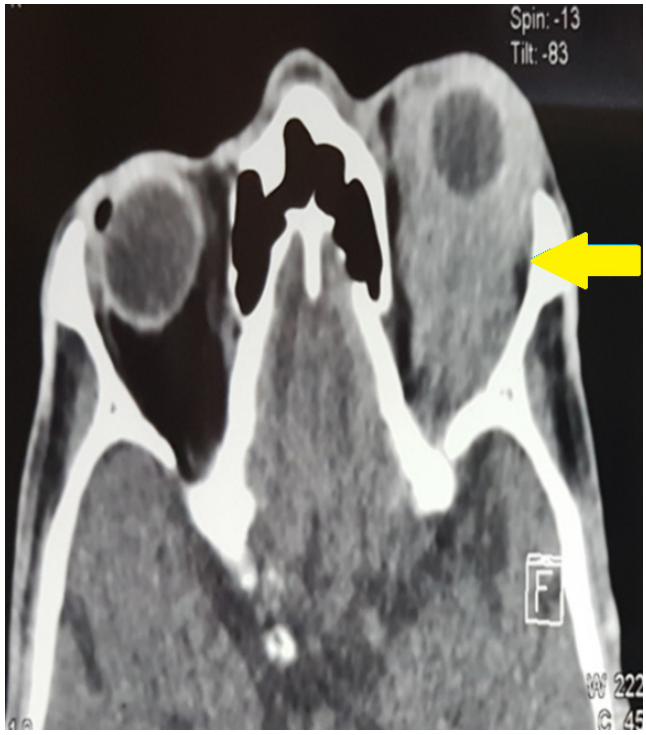


Figure 1. CT Orbit (plain) axial view—hyperdense left intraconal lesion encasing the optic nerve with loss of fat plane. No bony erosion noted.

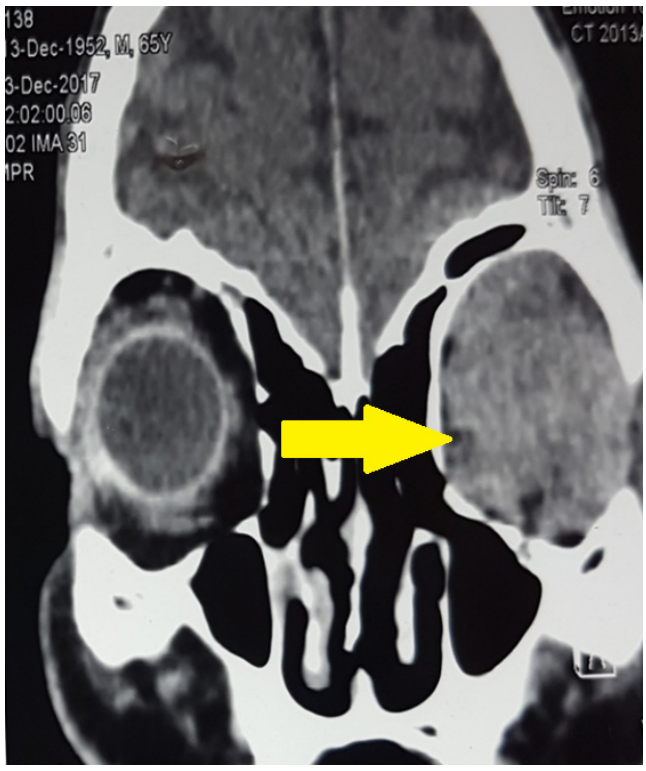


Figure 2. CT Orbit (plain) coronal image—hyperdense left intraconal lesion showing loss of fat plane with optic nerve and extraocular muscles. No bony erosion noted.

of the optic nerve. Secondary intra-orbital meningiomas arise intracranially and subsequently extend into the orbit [2]. ONSM is usually benign but may cause vision loss if without treatment. The vision loss is typically progressive, and patients may come to medical attention with varying degrees of optic nerve atrophy. Several meningioma subtypes have been proposed through the years, and the current (2021) World Health Organization (WHO) classification includes fif15 named entities [3]. Lymphoplasmacytic-rich meningioma (LPM) is an exceedingly rare type belonging to the Grade I WHO classification [4]. It affects young and middle-aged patients without sex predilection. It is a rare type of intra-orbital meningioma with no cases recorded in literature till now and mimics lymphoma.

Clinical presentation

The most common signs and symptoms of intra-orbital meningioma include slow, usually painless, progressive unilateral vision loss, and proptosis [5]. Central vision is preserved until late in the disease course. Other signs and symptoms are diplopia, transient visual obscuration, and headache. The classic triad is visual loss, optic atrophy, and opto-ciliary venous shunting. It has a predilection for young and middle-aged patients. It may sometimes be seen with secondary hyper-gammaglobulinemia and anemia, probably due to associated immunological reaction. No gender predilection is seen. ONSM in juvenile patients has a distinct natural history with the average age at presentation being 10 years and is more likely to be associated with neurofibromatosis 2.

Pathophysiology

Meningiomas are divided into 3 grades and 15 subtypes according to the 2021 WHO classification of tumors of the central nervous system [4]. LPM, which was first reported by Jain D et al [2], is a Grade I subtype of intracranial meningiomas. This grading system was developed primarily through clinic-pathologic correlations using intracranial meningiomas, and the current WHO classification has not been applied to a large series of intra-orbital tumors. Meningothelial meningioma is the most common subtype, followed by fibrous and transitional subtypes [6]. These three subtypes account for approximately 80 % of all meningiomas. Morphologically ONSM are solid, well-defined enlargement of optic nerve complex [5]. It encases

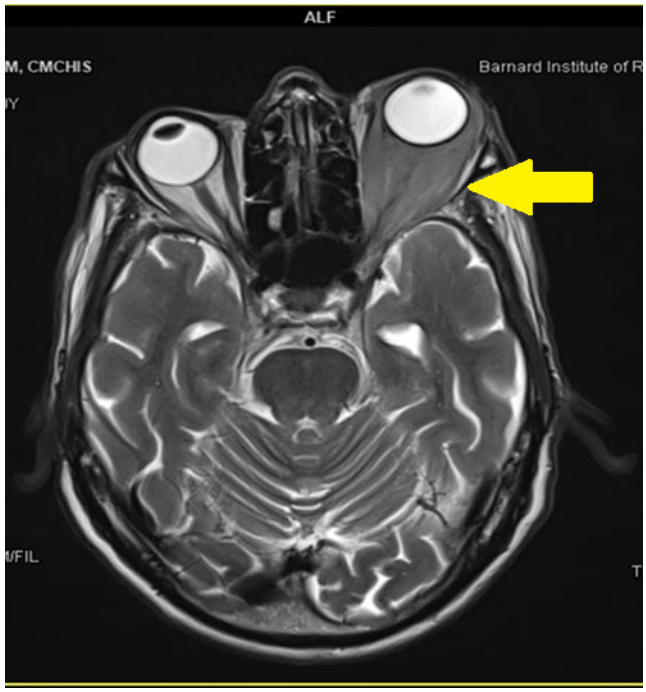


Figure 3. T2W axial image of brain & orbit—hypointense left intraconal lesion encasing the optic nerve with proptosis of left eye. There is also mild widening of the left optic canal.

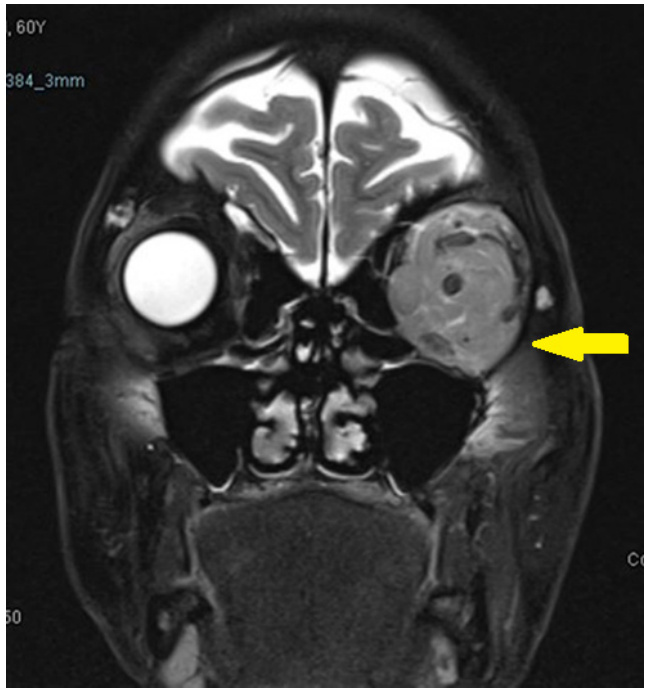


Figure 4. T2W coronal image of orbit—hypointense left intraconal lesion with optic nerve encasement and proptosis of left eye.

the optic nerve in the circumferential pattern but maybe eccentric or pedunculated as well. Diffuse thickening is more common than the segmental pattern, and en-plaque variants may also occur.

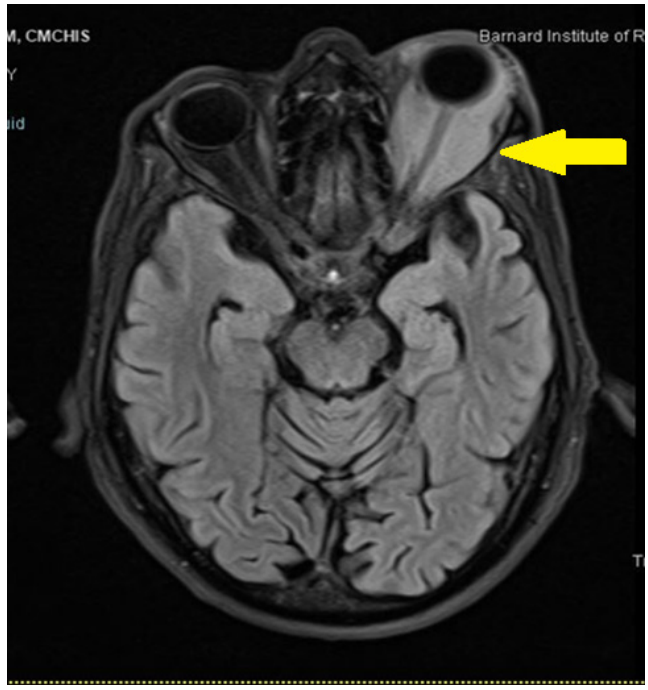


Figure 5. FLAIR axial image of brain & orbit—intraconal lesion causing proptosis with loss of fat plane with extraocular muscles of left orbit. There is also mild widening of the left optic canal.

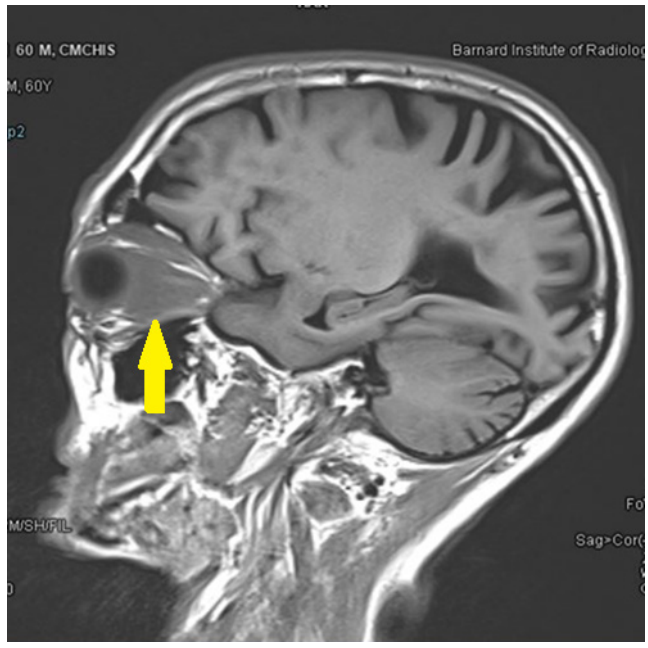


Figure 6. T1W weighted sagittal image of brain & orbit—isointense intraconal lesion of left orbit.

The lymphoplasmacytic rich variant of meningioma contains extensive chronic inflammatory infiltrates [4]. The lymphoplasmacytic-rich subtype characteristically arises as an en-plaque meningioma on the dura, with irregular forms and unclear

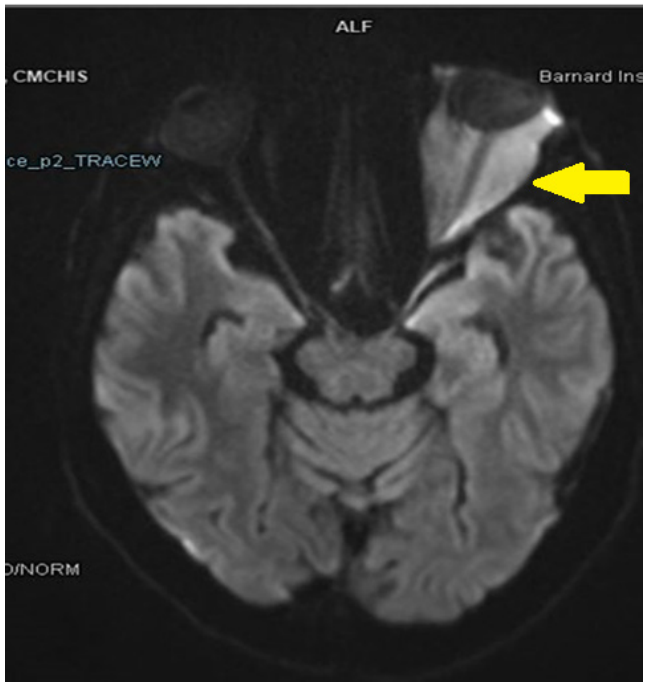


Figure 7. DWI image of brain & orbit—restricted diffusion of the intraconal lesion of left orbit.

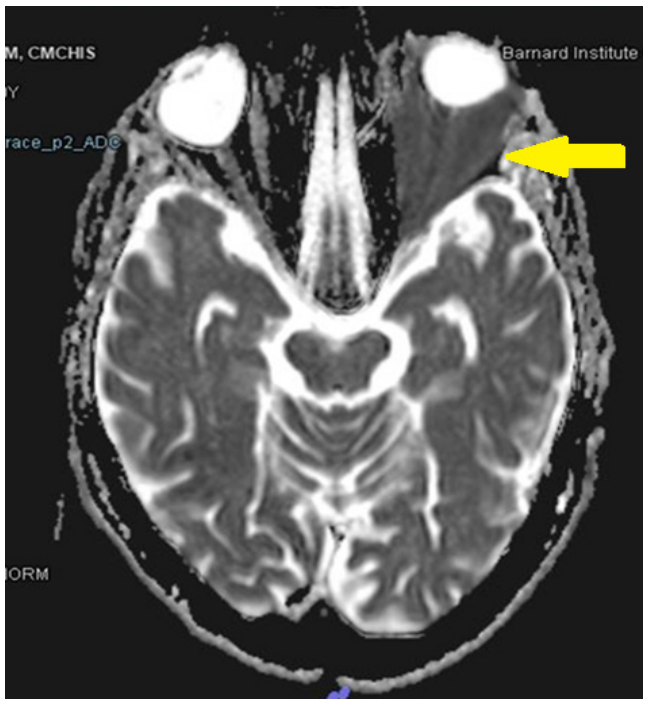


Figure 8. ADC image of brain & orbit—restricted diffusion with low ADC values of the intraconal lesion of left orbit.

tumor boundaries, invading into the adjacent brain tissue when the location is intracranial. Also, cyst formation is seen in 30% and peritumoral brain edema in nearly 100% of the cases. Intra-orbital

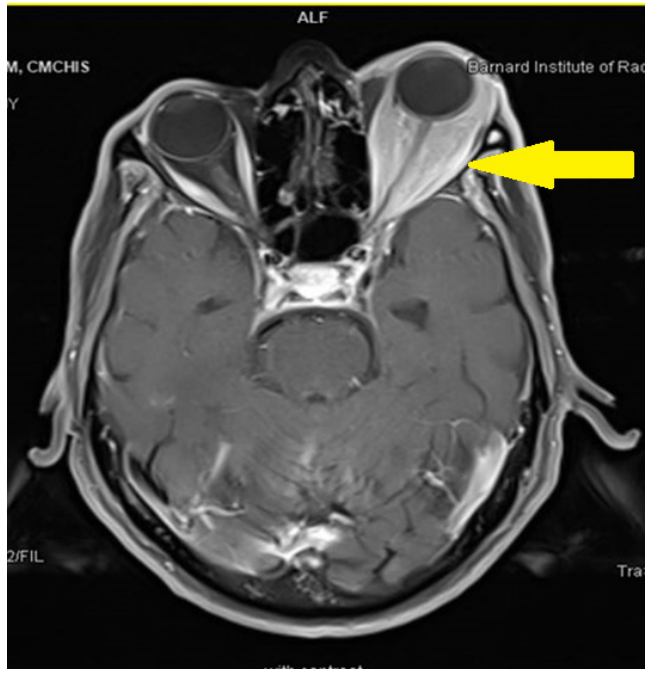


Figure 9. T1 FS axial image of brain & orbit—intense homogenous contrast enhancement of the intraconal lesion of left orbit.

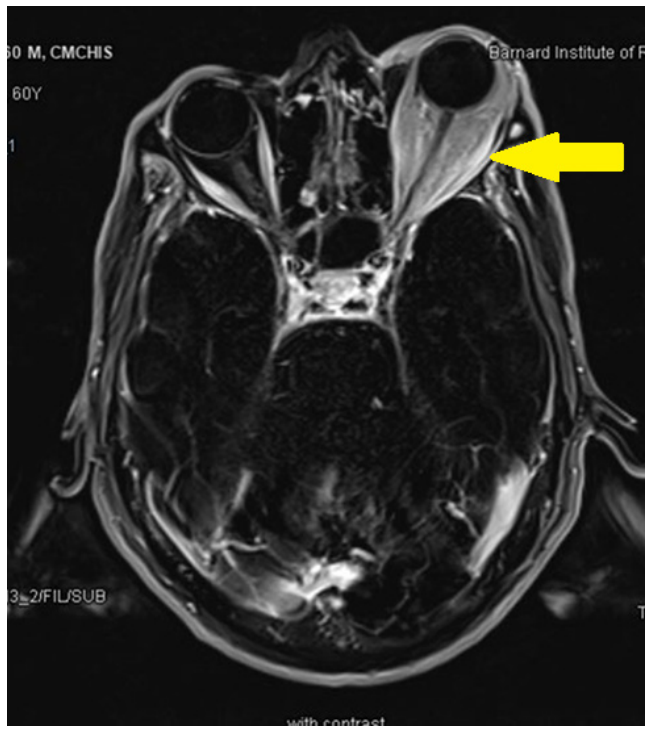


Figure 10. T1 subtracted image of brain & orbit—intense homogenous contrast enhancement of the intraconal lesion of left orbit.

lymphoplasmacytic rich meningioma has not been recorded in the literature as far as we have searched.

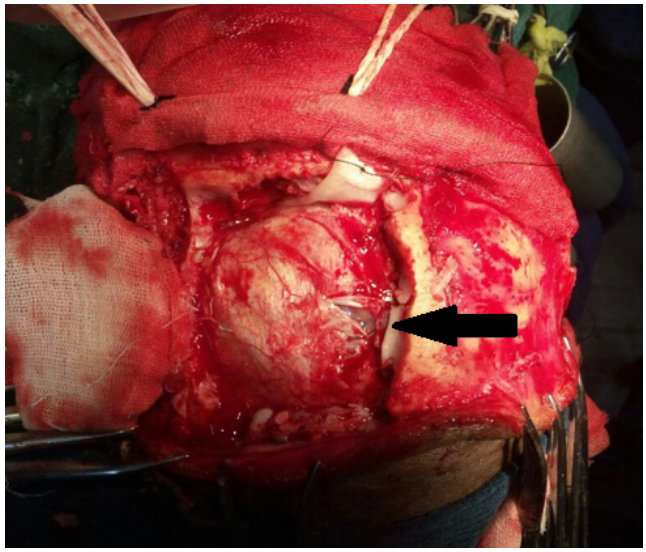


Figure 11. Intra operative image of left orbit.

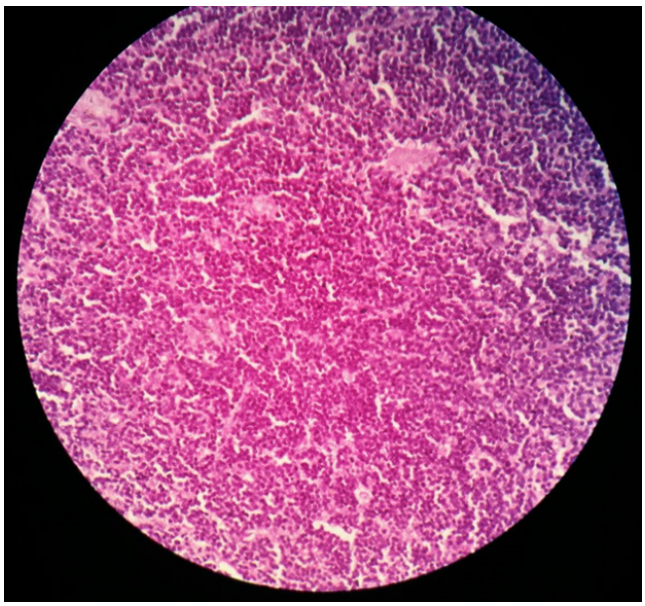


Figure 12. High power view of tumor composed of lymphocytes showing showed fragments of neoplasm arranged in cords and whorls. Some cells have eccentrically placed nucleus with pink cytoplasm which are plasma cells.

Imaging findings

Computed Tomography (CT)

Typically, the tumor is hyperdense in non-contrast CT. CT may be useful for demonstrating calcification within the lesion, a relatively specific finding for meningioma, which occurs in 20%–50% of cases and is not observed in optic nerve sheath glioma [7]. Another feature of optic nerve meningioma is the so-called tram-track sign, which is defined as

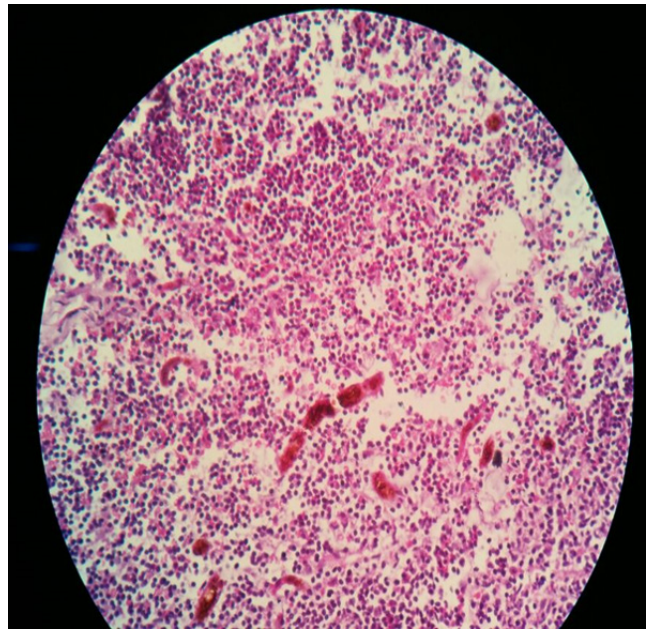


Figure 13. High power view of tumor composed of lymphocytes. Tumor cells are seen infiltrating around muscle fibers with intervening adipocytes and small blood vessels.

relative hypoattenuation of the optic nerve on CT. However, this is not a specific appearance and can be seen in other pathologies like pseudo-tumor, optic neuritis, sarcoid, lymphoma and metastases.

Magnetic Resonance Imaging (MRI)

The tumor is iso- to hypo-intense in T1WI [5]. T2WI show mixed hyper- to iso-intensity more often than hypo-intensity. Post-contrast MR images show strong and homogeneous enhancement with indistinct margins, likely reflecting tumor extension and inflammatory cell infiltration [6]. Tram track sign may also be seen due to peripheral enhancement of the tumor around the nerve. Restricted diffusion may be seen.

Differential diagnosis

In our case, with the imaging characteristics of the lesion, namely, T2 hypointensity, restricted diffusion with low ADC values, intense homogenous contrast enhancement, encasement of the optic nerve with loss of fat plane with all extraocular muscles and molding to the intraconal compartment in a plastic fashion, the possibilities considered were lymphoma, inflammatory pseudotumor, optic nerve glioma, orbital sarcoidosis, and metastasis.

Orbital lymphoma is a solid, pliable, homogeneously enhancing tumor, and it can involve any part of orbit with lacrimal predilection [5]. On imaging, it

Table 1. Maging Findings in various differntial diagnosis of Orbital lesions.

Differential diagnosis	CT	MRI
Lymphoma	On non-contrast CT, the mass is usually homogeneous in density, either iso-dense or slightly hyperdense when compared to the extraocular muscles. Following administration of contrast, only mild to moderate enhancement is seen, similar again to the extraocular muscles and lacrimal gland.	T1—iso to hypointense. T2—mildly T2 hypointense. Diffusion Weighted Imaging (DWI)—restricted diffusion Contrast—shows moderate to marked homogeneous enhancement.
Optic nerve glioma	The optic nerve is variably enlarged. The nerve may be elongated with kinking or buckling.	Optic nerve is enlarged. T1—iso to hypointense. T2—central T2 hyperintensity with thin low signal at the periphery representing the dura. Contrast—variable contrast enhancement.
Inflammatory pseudotumor	Enlargement of the muscle belly of one (or more) extraocular muscles with the involvement of their tendinous insertions. It may extend outside of the orbit via superior or inferior orbital fissures.	T1: enlarged extraocular muscles appear iso to hypointense. T2: typically hypointense due to fibrosis. Contrast: moderate to a marked diffuse enhancement
Orbital sarcoidosis	Asymmetrically enlarged lacrimal glands which show enhancement post contrast.	T1—asymmetrically enlarged lacrimal glands which are hypointense. T2—hypointense Contrast—enhancement is seen.
Orbital metastasis	Extra-conal soft tissue deposits, well defined or infiltrating. Show enhancement post contrast. Bone destruction may be seen.	T1 isointense to extra-ocular muscles. T2—hyperintense to extraocular muscles. Contrast—variable enhancement.
Cavernous hemangiomaCAVERNOUS HEMANGIOMA	Well circumscribed intraconal soft tissue density lesion which shows gradual filling of contrast. Calcification may be seen in sclerosing subtype. No bony destruction.	T1 isointense to extra-ocular muscles. T2—h –Hyperintense to extraocular muscles. Hypointense pseudocapsule may be seen. Contrast— – gradual filling of contrast

shows lobulated margins & molds to adjacent structures in a "plastic" fashion. On non-contrast CT, the mass is usually homogeneous in density, either iso-dense or slightly hyperdense when compared to the extraocular muscles [8]. Following administration of contrast, only mild to moderate enhancement is seen, similar again to the extraocular muscles and lacrimal gland. On MRI, it is mildly hypointense on T2WI with decreased ADC values, particularly in true lymphoma, and shows moderate to marked homogeneous enhancement.

Optic nerve glioma is an aggressive subtype in adults with unilateral enlargement, moderate irregular enhancement, and posterior extension [9].

Inflammatory pseudo-tumors are usually ill-defined, often associated with fat tissue infiltration or edema, and present with painful exophthalmos [10]. Imaging demonstrates an enlargement of the muscle belly of extraocular muscles with the involvement of their tendinous insertions. Imaging may also show an infiltrative mass extending into the superior and inferior orbital fissures. <AQ> The enlarged extraocular muscles are iso- to hypointense on T1 & hypointense on T2WI due to the presence of fibrosis. Diffuse enhancement is seen post-contrast. Sarcoidosis is a non-caseating granulomatous inflammation with diffuse infiltration of orbital structures and dural thickening [11]. Imaging features include asymmetrically enlarged lacrimal glands, which shows enhancement on contrast images. Metastasis within the orbit appears as T1 iso- & T2 hyperintense extra-conal deposits, with heterogeneous enhancement and bone destruction [12]. Another common intra-conal lesion in adults, presenting with gradual proptosis & visual disturbances, is the cavernous hemangioma which is a slow flow venous malformation. Imaging features include a well-defined soft tissue density lesion (CT) which shows gradual filling of contrast. In MRI, the lesion appears hyperintense to muscles in T2WI and isointense to the muscles in T1WI. A T2 hypointense pseudocapsule may also be demonstrated [5]. These lesions usually spare the orbital apex but may on occasion have intracranial extension [13].

Conclusion

ONSMs are benign tumors from the arachnoid cap cells of the optic nerve sheath. They represent 20%

of all orbital meningiomas, the majority of which are direct extensions from intracranial meningiomas. Patients usually present with loss of vision and proptosis. Lymphoplasmacyte-rich meningioma is an exceedingly rare type belonging to the Grade I WHO classification. It affects young- and middle-aged patients without sex predilection. Typically, the tumor is hyperdense on non-contrast CT and iso- to hypo-intense on T1WI. T2WI show mixed hyper- to iso-intensity more often than hypo-intensity. Restricted diffusion may be seen. Post-contrast MR images show strong and homogeneous enhancement with indistinct margins, likely reflecting tumor extension and inflammatory cell infiltration. Lymphoplasmacyte rich meningioma is a rare type of intra-orbital meningioma with no cases recorded in literature till now and mimics lymphoma.

Acknowledgment

Nil.

References

1. Daniel E. Meltzer MD. Orbital imaging-a pattern-based approach. Radiol Clin North Am 2015; 53(2015):37–80.
2. Jain D, Ebrahimi KB, Miller NR, Eberhart CG. Intraorbital meningiomas-a pathologic review using current World Health Organization criteria. Arch Pathol Lab Med 2010; 134:766–70.
3. Louis DN, Perry A, Wesseling P, Brat DJ, Cree IA, Figarella-Branger D, Louis D, Perry A, Wesseling P et al. The 2021 who classification of tumors of the central nervous system: a summary.The 2021 WHO Classification of Tumors of the Central Nervous System: A Summary. Neuro Oncol Neuro-Oncology.2021; 23(8):1231–-51;. doi:10.1093/neuonc/noab106 -— Pubmed
4. Zhu HD, Xie Q, Gong Y, Mao Y, Zhong P, Hang FP, et al, Lymphoplasmacyte-rich meningioma: our experience with 19 cases and a systematic literature review. Int J Clin Exp Med 2013; 6(7):504–15.
5. Diagnostic imaging: head and neck, 3rd edition, Elsevier, 2017.
6. Kunimatsu A, Kunimatsu N, Kamiya K, Katsura M, Mori H, Ohtomo K. Variants of meningiomas: a review of imaging findings and clinical features. Jpn J Radiol 2016; 34:459–69.
7. Grainger & Allison's diagnostic radiology-a textbook of medical imaging. 6th edition,ALLISON'S 2014.
8. Maksimovic O, Bethge WA, Pintoffl JP, Vogel M, Claussen CD, Bares R, et al. Marginal zone B-cell non-Hodgkin's lymphoma of mucosa-associated lymphoid tissue type: imaging findings. AJR Am J Roentgenol 2008; 191(3):921–30.
9. Müller-Forell WS, Boltshauser E. Imaging of orbital and visual pathway pathology. Springer Verlag, 2005.
10. Schaffler GJ, Simbrunner J, Lechner H, Langmann G, Stammberger H, Beham A, et al. Idiopathic sclerotic inflammation of the orbit with left optic nerve compression in a patient with multifocal fibrosclerosis. AJNR Am J Neuroradiol, 2000; 21(1):194–7.
11. Koyama T, Ueda H, Togashi K, Umeoka S, Kataoka M, Nagai S. Radiologic manifestations of sarcoidosis in various organs. Radiographics 2004; 24(1):87–104.
12. Moedder U, Telger TC. Head and neck imaging. Thieme, Stuttgart, Germany, 2008.
13. Mawn LA, Jordan DR, Gilberg SM. Cavernous hemangiomas of the orbital apex with intracranial extension. Ophthalmic Surg Lasers. 1998 Aug; 29(8):680–-4. PMID: 9715495.