



Case report ⁶⁸Ga-DOTATATE of optic nerve sheath meningioma

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ARTICLE INFO

Keywords:

Meningioma
Molecular imaging
Somatostatin receptor
PET-CT
⁶⁸Ga-DOTATATE

ABSTRACT

We report a patient with an optic nerve sheath meningioma whose diagnosis and management were guided by using Gallium-68 DOTA-Tyr³-octreotate (⁶⁸Ga-DOTATATE). Positron Emission Tomography-Computed Tomography (PET-CT).

1. Case presentation

A 44-year old woman with no significant medical history was referred to the neuroophthalmology department for evaluation of left-sided optic nerve edema that was incidentally found on an annual eye examination.

Visual acuity was 20/20 on the right and 20/25 on the left. There was a left relative afferent pupillary defect. No proptosis was noted, and the patient had intact color vision with a full range of ocular movements. Slit lamp examination was unremarkable, while fundus examination showed swelling of the left optic disc (Fig. 1). Humphrey 24-2 visual field testing showed non-specific superior and inferior visual field changes in both eyes. (Humphrey Field Analyzer; Zeiss, Germany).

Magnetic resonance (MR) scan of the brain and orbits with Gadolinium-chelate enhancement was interpreted at an outside hospital as optic perineuritis. However, review of the images in Neuro-ophthalmology was more suggestive of an optic nerve sheath meningioma (ONSM) with homogeneous intense enhancement producing the classic “tram track” appearance around the nonenhancing left optic nerve (Fig. 3). There was no intracranial extension of the lesion, or any evidence of surrounding-structures invasion.

Orbit computed tomography (CT) scan with contrast enhancement was performed that revealed asymmetric enlargement/enhancement of the left optic nerve/sheath with mild retrobulbar stranding, but no calcifications were identified (Fig. 4).

Given the conflicting interpretations, a ⁶⁸Ga-DOTATATE PET-CT was obtained (Fig. 5) and showed an asymmetric fusiform enlargement of the left optic nerve with high curvilinear radiotracer uptake and a maximum standardized uptake value (SUVmax) of 2.4, consistent with the enhancement observed on MRI. The combination of anatomic location and uptake was suggestive of an optic sheath meningioma. The patient was referred for radiation therapy and underwent fractionated radiotherapy to the left optic nerve to dose of 50.4 Gy in 28 fractions. On follow-up exam after finishing the treatment course, optic disc edema (as shown in Fig. 2) showed significant improvement. There was also improvement in the visual field defects in the left eye.

2. Discussion

Optic nerve sheath meningiomas (ONSM) are benign neoplasms of the meninges surrounding the optic nerve. Clinical presentation includes visual changes, color vision loss, visual field defects, optic disc edema and less frequently motility disturbance and exophthalmos ^{1,2}

The diagnosis of ONSM relies heavily on imaging findings.

MRI is the modality of choice for diagnosis of ONSM. The tumors are typically isointense or slightly hypointense to brain and optic nerve tissue on T1-weighted images and isointense or slightly hyperintense on T2-weighted images. They present a homogeneous enhancement often suggesting a “tram track” appearance around the hypointense optic nerve in axial sequences. CT may show calcification within the

Abbreviations: CT, Computed Tomography; ONSM, Optic Nerve Sheath Meningioma; MRI, Magnetic Resonance Imaging; PET, Positron Emission Tomography; SUV, Standardized Uptake Value.

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<https://doi.org/10.1016/j.ajoc.2021.101048>

Received 10 September 2020; Received in revised form 16 November 2020; Accepted 21 February 2021

Available online 5 March 2021

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Fig. 1. Fundus photographs show left-sided optic disc edema; the right disc is normal.

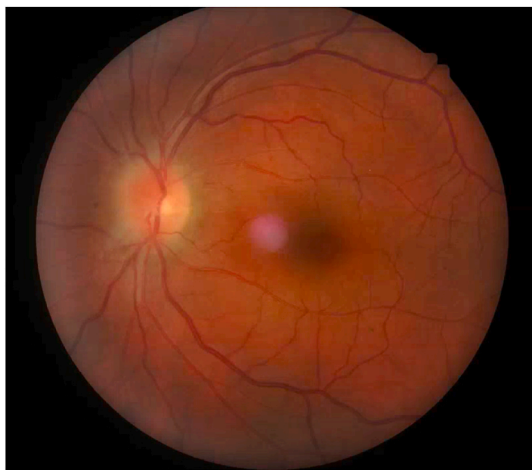


Fig. 2. Fundus photograph after treatment shows significant improvement in optic disc edema in the left eye.

meningioma. Contrast-enhanced structural imaging techniques such as MRI and CT are routinely used for defining the extent of the tumor, treatment planning, and monitoring, as well as for follow-up after treatment. However, these imaging techniques are not specific for diagnosing meningioma, and optic perineuritis, or other perineural inflammation or tumor may have a similar appearance on MRI and CT.^{3, 4}

PET-CT has gained considerable importance for diagnostic purposes in general oncology.⁵⁻⁸ Meningioma cells strongly express somatostatin receptor subtype 2 (SSTR2). Somatostatin analogs include Indium-111 pentetreotide for SPECT imaging and ⁶⁸Ga-DOTA peptides in PET imaging including ⁶⁸Ga-DOTATOC, ⁶⁸Ga-DOTATATE, and ⁶⁸Ga-DOTANOC. These PET somatostatin analogs show uptake in meningiomas, but not in other orbital lesions such as perineural inflammation, leukemic infiltration, or optic nerve glioma. In a ⁶⁸Ga-DOTATE PET-CT study quantifying SSTR2 expression of meningiomas and SUVmax values, receiver operator curve analysis revealed the threshold for best discrimination of meningioma from nonmeningioma pathology as an SUVmax of 2.3.⁹

Prior reports of orbital optic nerve meningiomas imaged with Gallium-68 DOTA-Tyr3-octreotate (⁶⁸Ga-DOTATATE Positron Emission Tomography-Computed Tomography (PET-CT)) showed focal masses within the orbits, and the PET/CT helped establish the diagnosis

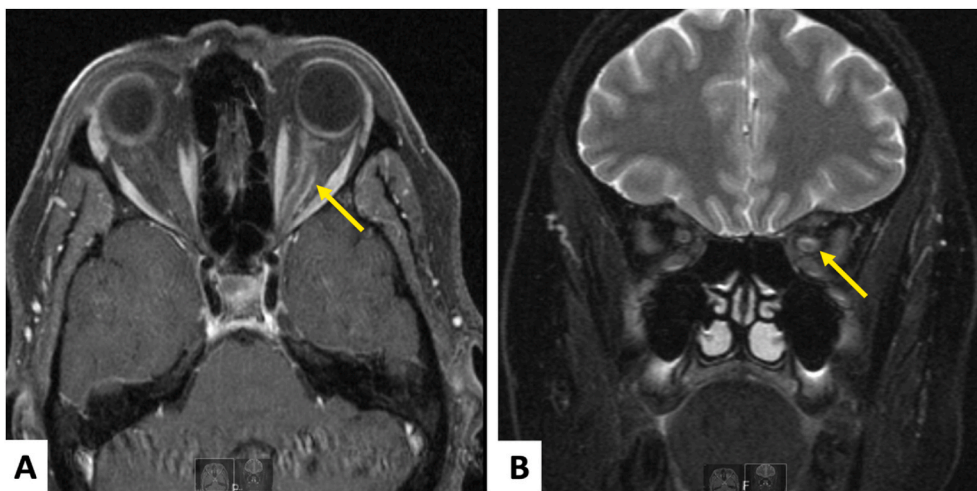


Fig. 3. (A) Axial fat-saturated postcontrast T1-weighted MRI shows homogeneous enhancement producing a “tram track” appearance around the nonenhancing left optic nerve (yellow arrow), with minimal intraconal fat-stranding. (B) Coronal fat-saturated T2-weighted MRI shows circumferential T2-prolongation (yellow arrow) surrounding the normal-appearing left optic nerve. . (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

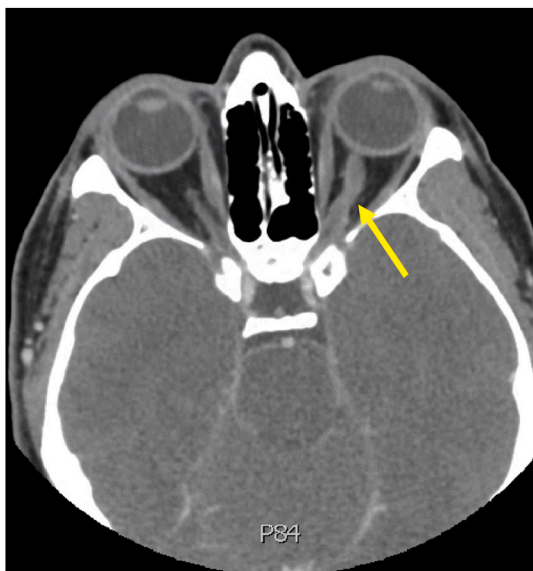


Fig. 4. Axial postcontrast CT scan shows enhancement around the non-enhancing left optic nerve (yellow arrow), with minimal retrobulbar stranding, and no associated calcification. (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

of meningioma vs other tumor histologies.^{10,11} This case shows diffuse thin enhancement around the optic nerve which was initially interpreted on MRI and CT as *perineuritis*, not tumor. In this case, the ⁶⁸Ga-DOTA-TATE PET/CT was *essential* to establish the diagnosis of optic nerve sheath *meningioma*, rather than inflammation/*perineuritis* which can have identical imaging findings as meningioma on MRI/CT.

⁶⁸Ga-DOTATATE PET-CT is a valuable diagnostic tool for the classification of equivocal tumors of the anterior optic pathway with both a sensitivity and specificity of 100%.¹²

3. Conclusion

MRI remains the procedure of choice for the diagnosis of ONSM. The MR imaging findings in optic nerve sheath meningioma are quite typical, but optic perineuritis has a similar MRI appearance. The somatostatin receptor ligand Ga⁶⁸-DOTATATE PET-CT shows uptake by meningiomas, but not by other lesions such as optic perineuritis or leukemic infiltrates, so it can distinguish ONSM from other intraorbital/optic nerve lesions. In this case, the benign noninvading growth pattern, the clinical manifestation and the lack of response to steroid therapy also strongly suggest and corroborate the imaging diagnosis of meningioma. Biopsy is not recommended because stripping the meningioma from the optic nerve will strip the optic nerve vasculature, blinding the patient. Ga⁶⁸-DOTATATE PET-CT is a valuable tool in the evaluation and diagnosis of ONSM.

Patient consent

Written consent to publish this case has not been obtained. This report does not contain any personal identifying information.

Funding

No funding or grant support

Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

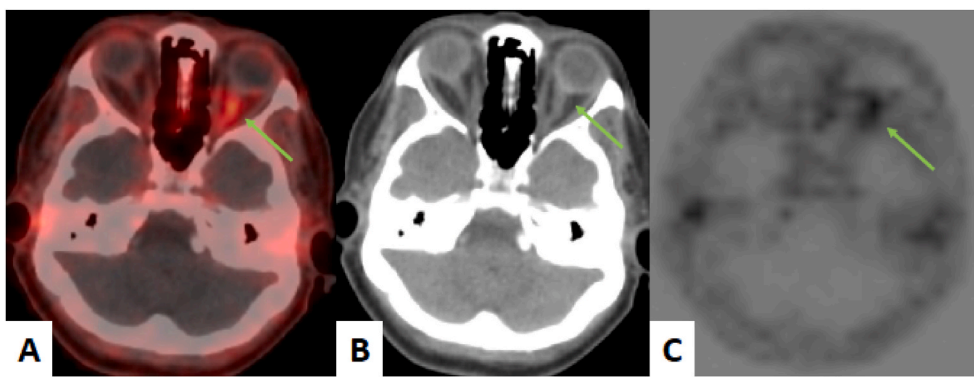


Fig. 5. ⁶⁸Ga-DOTATATE PET-CT demonstrates asymmetric fusiform enlargement of the left optic nerve-sheath complex with high curvilinear uptake corresponding to the contrast enhancement on MRI (green arrows). (A) fused axial PET-CT, (B) axial CT, (C) axial PET. (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

Acknowledgement

We thank the patient for granting permission to publish this information.

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