Ophthalmoplegia: A Rare Initial and Unusual Presentation of Pituitary Adenoma

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Abstract

Introduction: Visual manifestations in pituitary adenoma range from no presenting symptom to visual acuity deterioration to blindness. The presentation of pituitary adenoma is varied. It classically presents with visual field defects, bitemporal hemianopia being the most common Ocular motility disorders in pituitary adenomas are rare presentations. Pituitary adenoma presenting with oculomotor palsies as an initial symptom is very rare, total ophthalmoplegia being the rarest initial presentation. Aim: To report a rare case of pituitary adenoma with cavernous sinus invasion presenting initially with total ophthalmoplegia.

Keywords: Pituitary macroadenoma, total ophthalmoplegia, initial presentation, cavernous sinus invasion.

INTRODUCTION

Pituitary adenomas are benign tumors of intracranial neoplasms representing approximately 15–20% [1]. These are further classified as micro or macro based on size, with a 1 cm cutoff. Macroadenomas are in close proximity to crucial neural and vascular structures including the pituitary gland, hypothalamus, optic nerve and chiasma and carotid arteries. Left untreated, approximately 50% of macroadenomas will enlarge with more than two-thirds of the patients experiencing worsening of their vision [2]. The majority of patients who have pituitary adenoma will seek medical attention because of a mass effect (i.e. the growing tumour applying pressure to tissues) from the macroadenoma.1 Headsaches are a common presenting symptom for expanding adenomas of the pituitary and result from erosion of the bony sella turcica or stretching of the diaphragm sellae.

Pituitary adenomas classically present with headache and visual field defects, bitemporal hemianopia being the most common.

Visual symptoms in pituitary adenomas range from no presenting symptoms to marked visual impairment to blindness. Impaired vision is the most commonly reported symptom associated with an expanding pituitary adenoma [4]. Visual field defects exist in 60–70% of patients with pituitary adenoma at the time of suspicion of the diagnosis, which results from pressure on the optic chiasm by the tumour [3, 5]. Compression of the chiasma can result in a symmetrical bi-temporal hemianopia but more commonly there is an asymmetrical bi-temporal hemianopia or even a unilateral temporal field defect [5]. Macroadenoma growing superiorly out of the pituitary fossa (or for that matter other pituitary region masses) will contact, elevate and compress the central part of the chiasma in most individuals.

Some macroadenomas demonstrate invasive growth and in such cases, extension into the cavernous sinuses is characteristic. It may compress cranial nerves resulting in deficits, although this is uncommon [6].

Ocular motor palsy due to pituitary adenoma is reported to occur in only 5–17% of cases [7]. Such palsies include either indirect compression on the ocular cranial nerves by compressing the cavernous sinus or direct compression through cavernous sinus invasion [7, 8]. Ocular motility disorders in pituitary adenoma are very rare presentations. Only few reports address oculomotor palsies as an initial presenting symptom, ranging from isolated single-nerve palsy to total ophthalmoplegia [7, 9]. Adenomas that usually invade the cavernous sinus grow through fragile medial sinus wall with mediolateral expansion resulting in lateral displacement of the internal carotid artery, with third nerve compression in the oculomotor trigone (roof of the cavernous sinus). Thus the most commonly involved
cranial nerve by the compression is the third nerve. At the oculomotor trigone there is a whole cistern of cerebrospinal fluid which represents a possible route for tumor invasion [10,11]. In cases of 3rd nerve palsy occurring due to pituitary tumors, of all muscles supplied by the third nerve, the levatorpalpebraesuperioris was the most commonly affected, as shown by partial or complete ptosis[12]. The sixth nerve passes within the cavernous sinus adjacent to the internal carotid artery being less amenable to compression. Lateral rectus palsy occurs when the sixth nerve is compressed by posterior tumor expansion toward the petrous bone where the abducens nerve is tethered to a firm structure at Dorello’s canal. This is commonly observed with aggressive expansion of pituitary neoplasms as reported by many studies [13-15]. The fourth nerve is protected at its entry into the cavernous sinus by a doubling of the dura mater; so isolated fourth nerve palsy is very rare except in case of massive compression of the cavernous sinus, involving all the ocular motor nerves [7, 13, 16].

Here we report a case of pituitary macroadenoma in a middle aged adult male who presented with an initial symptom of total ophthalmoplegia, being the rarest initial presentation. Early tumor excision with cranial nerve decompression has a favourable outcome in these patients.

**CASE REPORT**

A 48 year old male presented to eye OPD with chief complaint of decreased vision and drooping of left upper eyelid for 6-7 days with history of mild headache on and off for 6-7 days. There was no significant past medical history of DM, hypertension, TB, hypo and hyperthyroidism. On examination, best corrected visual acuity in right eye was 6/9 and in left eye was 6/12. There was complete ptosis with marked restriction of ocular movements in all gazes in left eye (figure-1). No associated proptosis, nor signs of any inflammation was seen in left eye. The pupils were sluggishly reacting to light in both eyes. The fundus examination showed mild temporal disc pallor in both the eyes. The rest of the ocular examination in right eye was within normal limits. A diagnosis of total ophthalmoplegia, involving 3rd, 4th and 6th cranial nerves, in left eye was made and patient was advised a contrast enhanced MRI brain with orbits. MRI revealed a well-defined lobulated peripherally enhancing lesion showing cystic area, suggestive of necrosis measuring approx. 18.5(AP)×30.2(TR)×26.8(CC)mm in sellar and suprasellar location with ballooning of sella, infiltrating into left cavernous sinus, encasing slightly less than 50% of left ICA with maintained flow void causing mild superior displacement and stretching of optic chiasm with effacement of chiasmatic and infundibular recesses of 3rd ventricle -suggestive of pituitary macroadenoma with central necrosis (figure-2).

Full hormonal profile was done and revealed S prolactin- 2.36ng/ml (normal- 2.1-17.7ng/ml), Lutenizing hormone -1.06 mIU/ml (normal- 1.5-9.3mIU/ml), follicle stimulating hormone-3.17mIU/ml (normal-1.4-18.1mIU/ml), cortisol <1.0 microgram/dl (normal- 3.7-19.4).

Patient was referred to neurosurgeon and was taken up for endoscopic Trans-nasal Trans- sphenoidal approach tumour excision with cranial nerve decompression. On follow up after 2 weeks, patient showed near complete improvement in ophthalmoplegia.

![Fig-1: Showing restriction of ocular movement left eye](image-url)
CONCLUSION

A diagnosis of pituitary adenoma with cavernous sinus invasion should be kept as a possibility in acquired ocular motility disorders. These patients can present rarely with ocular palsies ranging from isolated single nerve palsy to total ophthalmoplegia. An early tumour excision has a favourable outcome in the improvement of ocular motility.

REFERENCE

1. Melmed, S. (2011). Pathogenesis of pituitary tumors. Nature Reviews Endocrinology 7 257–266. Wass, J. A., & Karavitaki, N. (2009). Nonfunctioning pituitary adenomas: the Oxford experience. Nature Reviews Endocrinology, 5(9), 519.
2. Jaffe, C. A. (2006). Clinically non-functioning pituitary adenoma. Pituitary, 9(4), 317-321.
3. Hennessey, J. V., & Jackson, I. M. (1995). Clinical features and differential diagnosis of pituitary tumours with emphasis on acromegaly. Bailliere's clinical endocrinology and metabolism, 9(2), 271-314.
4. Dekkers, O. M., Pereira, A. M., & Romijn, J. A. (2008). Treatment and follow-up of clinically nonfunctioning pituitary macroadenomas. The Journal of Clinical Endocrinology & Metabolism, 93(10), 3717-3726.
5. Pisaneschi, M., & Kapoor, G. (2005). Imaging the sella and parasellar region. Neuroimaging Clinics, 15(1), 203-219.
6. Lau, K. K. W., Joshi, S. M., Ellamushi, H., & Afshar, F. (2007). Isolated bilateral oculomotor nerve palsy in pituitary apoplexy: case report and review. British journal of neurosurgery, 21(4), 399-402.
7. Zada, G., Lin, N., & Laws, E. R. (2010). Patterns of extrasellar extension in growth hormone–secreting and nonfunctional pituitary macroadenomas. Neurosurgical focus, 29(4), E4.
8. Saffra, N., Kaplow, E., Mikolaenko, I., Kim, A., Rubin, B., & Jafar, J. (2011). Isolated sixth cranial nerve palsy as the presenting symptom of a rapidly expanding ACTH positive pituitary adenoma: a case report. BMC ophthalmology, 11(1), 4.
9. Yasuda, A., Campero, A., Martins, C., Rhoton Jr, A. L., & Ribas, G. C. (2004). The medial wall of the cavernous sinus: microsurgical anatomy. Neurosurgery, 55(1), 179-190.
10. Jankowski, P. P., Crawford, J. R., Khanna, P., Malicki, D. M., Ciacci, J. D., & Levy, M. L. (2015). Pituitary tumor apoplexy in adolescents. World neurosurgery, 83(4), 644-651.
11. Yen, M. Y., Liu, J. H., & Jaw, S. J. (1990). Ptosis as the early manifestation of pituitary tumour. British Journal of Ophthalmology, 74(3), 188-191.
12. Müslüman, A. M., Cansever, T., Yılmaz, A., Kanat, A., Oba, E., Çavuşoğlu, H., ... & Aydınl, Y. (2011). Surgical results of large and giant pituitary adenomas with special consideration of ophthalmologic outcomes. World neurosurgery, 76(1-2), 141-148.
13. Yokoyama, S., Hirano, H., Moroki, K., Goto, M., Imamura, S., & Kuratsu, J. I. (2001). Are nonfunctioning pituitary adenomas extending into the cavernous sinus aggressive and/or invasive?. Neurosurgery, 49(4), 857-863.
14. Zoli, M., Mazzatenta, D., Pasquini, E., Ambrosetto, P., & Frank, G. (2012). Cavernous sinus apoplexy presenting isolated sixth cranial nerve palsy: case report. Pituitary, 15(1), 37-40.
15. Möller-Goede, D. L., Brändle, M., Landau, K., Bernays, R. L., & Schmid, C. (2011). Pituitary apoplexy: re-evaluation of risk factors for bleeding into pituitary adenomas and impact on outcome. European Journal of Endocrinology, 164(1), 37-43.