CASE REPORT

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Pituitary Macroadenoma with Superior Orbital Fissure Syndrome

Tapan Nagpal, Ankit Singhania

ABSTRACT

Introduction: Pituitary adenomas are benign tumours which arise within the anterior lobe (adenohypophysis) of the gland in the sella turcica. Lateral extension into the cavernous sinuses can result in involvement of intracavernous part of 3rd, 4th and 6th cranial nerves resulting in superior orbital fissure syndrome. **Case Report:** We report a case of pituitary macroadenomas in which the involvement of the 3rd, 4th and 6th cranial nerves was more extracavernous than intracavernous. **Conclusion:** We report this rarity here with an endoscopic transnasal transsphenoidal treatment protocol that is minimally invasive and has less morbidity than open neurosurgical techniques.

Keywords: Pituitary adenoma; Superior orbital fissure syndrome

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INTRODUCTION

Pituitary adenomas account for 10-15% of all intracranial neoplasms [1]. Tumors more than 1 cm are called macroadenomas. The prevalence of macroadenomas with a diameter of more than 1 cm is estimated to be 0.2% [1]. Clinical symptoms depend on whether the tumor is secreting or non-secreting. Eighty percent of macroadenomas are of the non-functioning or non-secreting type [1]. Secreting adenomas manifest with specific endocrine syndromes. Hence they present earlier and are often smaller and confined to the gland. Non-secreting adenomas grow to a larger size and compress adjacent structures, most commonly causing a bitemporal hemianopsia due to chiasmatic compression. Lateral extension into the cavernous sinuses can result in involvement of intracavernous part of 3rd, 4th and 6th cranial nerves resulting in superior orbital fissure syndrome [2, 3]. With the advent of the endoscope these pituitary tumours can now be treated with an endoscopic transnasal transsphenoidal approach that is minimally invasive and has less morbidity than conventional open neurosurgical techniques [4].

CASE REPORT

A 27 year old female patient presented to our hospital with complaints of pain above the right eye for 1 month with dropping of the right eyelid and diplopia for 15 days. Physical examination revealed a young non-obese female. Ophthalmologic examination revealed complete ptosis of the right eye (Figure 1), anisocoria and right pupil dilated and slightly less reactive to light than the left. She had almost complete ophthalmoplegia of the right eye

Figure 1: Pre-operative photograph of patient showing complete ptosis of right eye.

Figure 2: MRI T1W Sagittal cuts with contrast showing 20x13 millimeters hypointense mass in the sellar area.

Figure 3: MRI T1W Coronal cuts with contrast showing 15x13 millimeters heterogeneously enhancing well encapsulated mass in the sellar area with right paracavernous extension.

(complete absence of lateral, medial and superior gaze and restricted inferior gaze), consistent with complete third and sixth cranial nerve palsy and partial fourth cranial nerve paresis. Slit lamp study indicated a normal anterior chamber. Dilated funduscopy did not reveal any pathology in the posterior chamber. Perimetry revealed mild peripheral restriction of both visual fields suggestive of early bitemporal hemianopsia. Her thyroid profile and prolactin levels were normal. She had no clinical signs of Cushing's syndrome, acromegaly or any menstrual disturbances.

Magnetic Resonance Imaging (MRI) revealed a 20 x 13 x 15 millimeters heterogeneously enhancing sellar mass with lateral extension to the right superior orbital fissure area. The lesion was well-encapsulated and limited by the internal carotid arteries on both sides. There was no significant suprasellar extension. The optic chiasma was unaffected (Figure 2, 3, 4). Considering her normal endocrinology, a diagnosis of non-functioning null cell pituitary adenoma was made.

The patient underwent an uneventful transnasal transphenoidal extracapsular removal of pituitary tumor. She responded well post-operatively without any sign of hypopituitarism and normal serum electrolyte levels. She was discharged on 5th post-operative day. An ophthalmologic exam on the 15th post-operative day

Figure 4: MRI T2W Axial cuts with contrast showing 15x 13 millimeters heterogeneously enhancing mass in the sellar area with right para extracavernous extension in area of superior orbital fissure with enhancement of right orbit due to probably blockage of superior ophthalmic vein in the medial most part of superior orbital fissure.

showed substantial improvement in third, fourth and sixth cranial nerve function, but her right pupil was still partly dilated and less reactive to light (Figures 5, 6, 7). The post-operative histopathology report was confirmatory for the diagnosis of pituitary adenoma. Immunocytochemical analysis to confirm the type of adenoma whether acidophilic, basophilic or null cell type could not be done due to limited resources. Close follow-up with neuroophthalmology and endocrinology and a repeat MRI after 3-4 months were recommended.

DISCUSSION

Non-functioning type of pituitary macroadenomas are clinically silent early on and present late with symptoms due to mass effect as they grow to a larger size with suprasellar extension and optic chiasma involvement [1]. Our patient had mass effect symptoms for about 1 month before she presented to us. Ophthalmoplegia is a rare complication of pituitary adenoma and occurred in 59 out of 1000 patients in one large series. It occurs due to lateral extension in the cavernous sinus [2]. In our case the lateral extension leading to ptosis, ophthalmoplegia and mydriasis in the right eye was more extracavernous in the

Figure 5: Post operative photopgraph showing substantial improvement in ptosis of right eye at 15th post operative day.

area of superior orbital fissure than intracavernous. The order of injury to cranial nerves by lateral extension of pituitary adenomas was to the third, sixth, fourth and lastly, the ophthalmic division of the fifth cranial nerve [2]. The fourth cranial nerve along with the frontal and lacrimal branches of ophthalmic division of 5th cranial nerve (V1) pass through the lateral-most part of the superior orbital fissure. The two divisions of the third nerve with the sixth nerve between them pass through the middle part of the superior fissure, hence these are involved early. Observational studies have revealed that third nerve palsy from a pituitary adenoma presents initially with mydriasis followed by ophthalmoplegia and lastly, ptosis. After surgical resection patients tend to improve in the reverse order in which symptoms developed [2]. In our case there was only restricted movement and incomplete palsy of the superior oblique muscle supplied by 4th cranial nerve. V1 was unaffected with no complaint of retro-orbital pain. The pattern of recovery in our case also showed substantial improvement in ptosis and ophthalmoplegia with mydriasis and weak light reaction persisting even at the 15th post-operative day. Most pituitary adenomas with cranial nerve palsies are

Figure 6: Latest clinical photograph of patient taken on 1-9-2010 showing complete clinical improvement.

Figure 7: Latest CT brain with contrast coronal cuts showing non-enhancing fat in the sellar area with no residual tumour.

associated with apoplexy and can be initially treated with high dose steroids with significant improvement [2]. In our case pre-operative steroids did not result in much improvement and early surgical intervention was contemplated. The recovery of cranial nerve function has been directly correlated with the earlier timing of surgery. The average time for third nerve recovery has been reported from 36 days to as long a year [3]. Surgery for pituitary adenomas is nowadays done with endoscopes, and the preferred approach is the transnasal transsphenoidal [4]. Extracapsular excision of tumor, though difficult is better than simple curettage as it helps

to identify a clear plane between the tumor and extradural nerves and more importantly the usually normal pituitary stalk and posterior pituitary are preserved which prevents post-operative hypopituitarism [5]. In our case extracapsular dissection was attempted with partial success after removal of the floor of the sella right up to the optic chiasma (where the double fold of dura is identified) and medial opticocarotid recess in the paraclinoid area. This enabled us to remove the entire tumour (though piecemeal) with decompression of the superior orbital fissure and preservation of the posterior pituitary.

CONCLUSION

Most of the pituitary macroadenomas are of non-functioning type tending to present late with mass effect. Lateral extension into the cavernous sinus leads to superior orbital fissure syndrome due to involvement of cranial nerves. Extracavernous involvement of the superior orbital fissure as in our case is a rarity. An endoscopic transnasal transsphenoidal extracapsular dissection of the tumor is a much better technique for removal of the tumor while preserving normal pituitary function than the conventional open neurosurgical techniques.

Author Contributions

Tapan Nagpal – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published

Ankit Singhanian – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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