An Unusual Presentation of Pituitary Macroadenoma

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Abstract

It is rare for a pituitary adenoma to present with cranial nerve palsy. Prolactinomas and non-functioning tumors are the common pituitary tumours which attain large sizes (macroadenomas). We are presenting two such cases where third cranial nerve palsy was the presenting symptom. A thorough ocular evaluation was done in both cases. One patient presented with unilateral ptosis, a dilated pupil and restriction of elevation in right eye. Another patient presented with unilateral ptosis and ill sustained pupillary reaction in the left eye. MRI evaluation in both cases revealed pituitary macro adenomas. One was an ACTH producing clinically nonfunctioning tumor, and other was a prolactinoma. It is rare for both the tumors to present as cranial nerve palsy. The proximity of the anterior pituitary to the third cranial nerve in cavernous sinus makes it more vulnerable to ocular motor nerve impairment.

Key words – pituitary adenoma, ptosis, third nerve palsy.

Introduction

Pituitary adenomas account for 10-15% of intracranial neoplasms. They can cause anterior pituitary hormonal imbalance or symptoms and signs related to invasion of surrounding structures. Pituitary tumors are classified by Hardy according to size and invasive characteristics. Stage I are microadenomas (less than 10mm in diameter). They may cause hormonal oversecretion but are not associated with structural problems. Stage II tumors are macroadenomas (greater than 10mm) with or without suprasellar extension. Stage III tumors are macroadenomas that locally invade the floor of the sella and may cause sellar enlargement and suprasellar extension with multiple cranial nerve palsy. We report this case because it is rare for a pituitary adenoma to present as isolated third nerve palsy.

Case 1

45-year-old male presented to us with complaints of drooping of right eye of 2 weeks duration (Figure: 1). He was not a known diabetic or hypertensive. There was no history of visual disturbances, headache or vomiting as well as hormonal imbalance. The best-corrected vision in both eyes was 6/6. The right eye showed restriction of elevation. There was mild ptosis of 4mm with good LPS function. The pupil was 5mm, sluggishly reacting to direct light and not reacting to consensual. Results of examination of all other cranial nerves were within normal limits. Fields, color vision and fundus examination in both eyes were normal. MRI revealed features suggestive of large pituitary macroadenoma filling sphenoid sinus and extending into nasopharynx, 3rd ventricle, right hypothalamus, right posterior cerebral artery and right crus cerebri. Both cavernous sinuses and internal carotid artery were also involved.
On histopathological analysis, the subepithelium showed tumor cells arranged in sheets and trabeculae with intervening fibrovascular core. Cells were round to oval exhibiting moderate amount of eosinophilic cytoplasm with vesicular nucleus and prominent nucleoli showing minimal pleomorphism that are PAS stain positive (Figure: 2). Immunohistochemical studies showed that ACTH was strongly positive (> 85% cells) and both GH and prolactin were negative.

He underwent endoscopic assisted trans-sphenoidal decompression. Postoperatively his ptosis improved remarkably.

**Case 2**

A 37 years old lady presented with progressive dimness of vision since last two months involving left eye along with occasional headache and inability to open the eye completely over a period of two months (Figure: 3). She also had amenorrhea for the last six months and lactation for two days. No history of vomiting, headache, seizure and loss of consciousness. She was a known diabetic for four years and hypertensive for two months. Unaided vision in both eyes was 6/12 improving to 6/6. There was moderate ptosis with mild fullness in LE. Extra ocular movements were normal in both eyes. The pupil was dilated with ill sustained pupillary reaction. Fundus was within normal limits. She had bitemporal field loss. MRI brain showed sellar lesion suggestive of pituitary adenoma with parasellar extension. Serum prolactin levels were elevated.

She was put on medical management with oral Cabergoline (Dopamine agonist) 0.5mg twice a week with periodic hormonal assay and follow up.

**Discussion**

It is rare for a pituitary adenoma to present with third cranial nerve palsy. There are only a few reports in literature. It could be due to tumor invasion or apoplexy. The onset is acute in apoplexy and in tumor invasion it is gradual. When correctly diagnosed and treated, the third nerve dysfunction appears to be reversible.

In both our cases, third nerve palsy had pupillary involvement unlike diabetic third nerve neuropathy in which pupillary reactivity is usually preserved. Lateral extension of pituitary tumor is usually associated with involvement of fourth and sixth cranial nerves, pain or numbness in the distribution of fifth cranial nerve and symptoms of compression of internal carotid artery. It is the third nerve being singularly involved that makes these cases different. Third nerve is liable to be easily compromised because of its close proximity to the anterior pituitary, which is the commonest site of pituitary adenoma (Figure: 4).
Prolactinomas and nonfunctioning tumours are the commonest pituitary macroadenomas. Though this is a case of ACTH producing tumour, it is clinically nonfunctional. ACTH producing adenomas account for about 10-15% of all pituitary tumours. Most ACTH producing pituitary tumours are relatively small microadenomas less than 5mm in diameter but macroadenomas are also seen. Some ACTH secreting adenomas are clinically silent but may present unusually with a third nerve palsy as seen in our case\textsuperscript{5,10,11}. Only less than 10% of pituitary tumors present with visual loss. Most of these are nonfunctioning tumors. Prolactinomas usually present with visual complaints, the most frequent objective finding being bitemporal hemianopia\textsuperscript{5,11}. However cranial nerve palsies are rarely seen in prolactinomas\textsuperscript{4}.

Conclusion

Invasive pituitary adenomas with extension to the cavernous sinus are rare and comprise 6-10% of all pituitary tumors\textsuperscript{4}. Among these, third nerve palsy is very rare. Nevertheless, any patient presenting with an isolated third nerve palsy should make one suspect a pituitary adenoma.

References