Case Report

A rare case of acquired ptosis in a child secondary to Pituitary macroadenoma

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Abstract

Pituitary adenomas are rare neoplasms with a propensity for unusual presentations and an aggressive clinical course. An 8 years old female child presented to Department of Ophthalmology at SVS Medical College and Hospital Mahabubnagar with the chief complaints of mild ptosis of RE since 1month, associated with fever, headache, vomiting and diplopia since 2 days. On ocular examination, BCVA in BE - 6/9 and anterior segment examination suggested RE mild ptosis with hypotropia and adduction restriction. Fundus examination was normal. There was no palpable mass/ bruit in the orbit. MRI revealed lobulated enhancing soft tissue lesion in sella and parasellar region extending into basifrontal region causing pressure over optic chiasm, 3rd ventricle and cavernous sinuses suggestive of pituitary macroadenoma. Hormonal essays revealed elevated prolactin, beta HCG levels and abnormal thyroid profile. On the basis of clinical examination and investigations a clinical diagnosis of pituitary macroadenoma was made. Then the case was referred to department of neurosurgery where transphenoidal resection under GA was planned and after 2days of admission she had a respiratory arrest and could not be resuscitated.

Key words

Pituitary macroadenoma, Ptosis, Diplopia, Ocular motility defect.

Introduction

Pituitary adenomas are rare neoplasms with a propensity for unusual presentations and an

aggressive clinical course [1]. Pituitary adenomas are neoplasms arising in the adenohypophysis [2]. They represent 10–25% of

all intracranial neoplasms and the estimated prevalence rate in the general population is nearly 17% [3, 4]. Most pituitary adenomas are benign. However, there are some aggressive adenomas that grow rapidly, tend to recur or progress and are resistant to currently available treatment options including surgery and radiation. Pituitary adenomas are, however, rare in children and adolescents constituting only about 3% or less of all diagnosed intracranial tumours and 5% of all pituitary adenomas [5]. When compared with the adult tumours, pituitary children adenomas in are predominantly comprised of secreting tumours, with prolactin, adrenocorticotropic hormone (ACTH), and growth hormone (GH) secreting tumours being the most frequent and in turn related to substantial morbidity [6]. Due to its rarity and complexity, paediatric pituitary adenoma has been relatively infrequently studied, and recommendations regarding its optimal management are debated.

Case report

An 8 years old female child presented to Department of Ophthalmology at SVS Hospital with chief complaints mild ptosis of RE since 1 month. She was apparently alright 2 days back. Then she suddenly developed diplopia which was more in right gaze and was stationary without any diurnal variation. She complained of severe headache, which was present in frontal area, throbbing in nature and bilateral and was associated with 2 episodes of vomiting. Vomiting was non-projectile, non-foul smelling and contained ingested food material. There was history of throbbing headache on and off, for which she was taking over the counter pain medications. Non-projectile vomiting were present on and off too, which were usually followed by episodes of headache.

On ocular examination, BCVA (best corrected visual acuity) both eyes was 6/9, colour vision was normal and RE mild ptosis, hypotropia and adduction restriction were noticed (**Figure - 1**).

Figure – 1: Photographs showing extraocular movement examination

a) RE Adduction restriction, b) RE Abduction and LE adduction- full range noted, c) RE Mild ptosis,
d) RE Mild hypotropia, e) BE infraduction –full range noted.







RAPD was absent and the other anterior segment findings were normal.

Fundus examination with 90D lens revealed no abnormality (**Figure - 2**).

Visual field examination couldn't be performed considering the patients clinical condition as she was drowsy and uncooperative.

Along with the routine hematological tests, hormonal panel was done including serum prolactin, beta hCG, and thyroid profile which revealed elevated prolactin, beta hCG levels and abnormal thyroid profile (**Table – 1**).

Serum TSH	3.4 U/ml (0.3-5 u/ml)	
Serum FT3	7.2 mcg/dl (1.8-5.4 mcg/dl)	
Serum FT4	15 mcg/dl (4.5-11.7 mcg/dl)	
Serum Prolactin	55.4 ng/ml(2.8-20.3 ng/ml)	
Serum Beta HCG	27.6 mIU/ml (0-5 mIU/ ml)	

Table – 1: Serum profile.

Contrast enhanced MRI brain and orbits showed large lobulated soft tissue/isointense lesion in sella, suprasellar and parasellar region with extension into basi-frontal region causing pressure over optic chiasm, anterior part of 3^{rd} ventricle and both cavernous sinuses suggestive of pituitary macroadenoma (**Figure – 3, 4, 5**).

Patient was referred to Department of Neurosurgery for further management and follow up where she was advised for transphenoidal resection under GA, but unfortunately after 2 days of admission she had respiratory arrest and coudn't be resuscitated.

Figure – 3: MRI T1 - post contrast images showing: (a) Sagital view showing well lobulated lesion arising from pituitary gland compressing optic chiasm and corpus callosum (arrow).



(b) Axial view showing area of calcification in mid brain and lesion compressing cavernous sinus superiorly, (arrow)



(c) Coronal view showing lesion compressing temporal lobe medially and 3rd ventricle superiorly (arrow)



Discussion

Pituitary adenoma is a benign tumor of pituitary gland, though it accounts for 10-15% of all primary intracranial tumors. Commonly presenting symptoms to Ophthalmology OPD are visual loss, diplopia, colour vision defect, headache and field defect.

Pituitary adenoma can be classified as follows [7]:

Size:

a) Macroadenoma (>10 mm)

b) Microadenoma(< 10 mm)

Hormonal activity:a) Secreting /functional tumorb) Non -secreting /non -functional tumor

26% of adenomas are non-secreting and cause symptoms due to pressure effect over optic chiasma region. Clinically it is presented as chiasmal syndrome with following symptoms: blurred vision, headache, nausea, vomiting, diplopia, desaturation, visual field defect, optic atrophy, poor fixation blindness and visual hallucination.

Pituitary adenomas are slow growing, benign neoplasms of epithelial origin [8].They may be manifested with diplopia initially, and rarely with full ophthalmoplegia when their size exceeds 1cm. About 5% of pituitary adenomas present with a more severe clinical picture leading to erosion of skull base by extending to subtemporal, cavernous sinus, suprasellar and sphenoid sinus involvement. MRI is always a superior diagnostic modality than a CT-scan in detecting sellar and suprasellar lesions. Extra ocular muscle restriction was partially involved in right eye as the mass was arising from pituitary gland and compressing optic chiasm and corpus callosum, cavernous sinus and 3rd ventricle superiorly, which also explains the reason for this patients complaints of diplopia and ptosis. These adenomas are generally associated with a poor prognosis.

Conclusion

Pituitary adenomas are associated with immense diversity in their endocrine manifestation secondary to hypo/hyper functioning of pituitary gland and ophthalmic manifestations [9]. presenting symptoms Commonly to Ophthalmology OPD are visual loss, diplopia, colour vision defect, headache and field defect. Visual acuity testing, colour vision and field testing by confrontation method are simple and yet very important tests that could give a clue to get visual filed testing done in these patients. Extra ocular muscle restriction was partially involved in right eye as the mass was arising from pituitary gland and compressing optic chiasm and corpus callosum, cavernous sinus and 3rd ventricle superiorly, which also explains the reason for the patients complaints of diplopia and ptosis. MRI is always a superior diagnostic modality than a CT-scan in detecting sellar and suprasellar lesions. An opinion of the endocrinologist and a neurologist is valuable and utmost urgency in an evidence of sellar and suprasellar lesions. In a functional pituitary macroadenoma 1st line treatment is surgical resection by Transphenoidal approach [10]. In the follow-up visit, visual acuity, visual field, fundus examination with 90D, extra ocular muscle involvement, hormonal measurement and pituitary imaging has to be done. Adenoma with poor prognosis was considered in this patient

because of similar findings. They offer better surgical outcome if diagnosed and treated early.

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