A Rare Case report of Non-secretory Pituitary Macroadenoma in a 14 years old child

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Abstract: A 14 year old boy presented to Department of Ophthalmology in AGMC with complaints of bilateral sudden visual loss for 7 days. Visual acuity in both eyes was PL positive only not improving with pin hole. Anterior segment examination suggested RAPD in both eyes with medial rectus & inferior oblique involvement in left eye. Due to severely diminished visual acuity, visual field and colour vision could not be elicited. Fundus examination with 90 D revealed temporal pallor with pale optic disc in both eyes. MRI revealed pituitary macroadenoma. Endocrinological reports were within normal limit suggesting non-secretory pituitary macroadenoma. The patient underwent transsphenoidal resection of pituitary and follow up at 6 month showed improved visual acuity by 6/60 in right eye and 5/60 in left eye. Pituitary Macroadenoma is a rare finding in children.

Keywords: Pituitary Macroadenoma, Temporal pallor, Optic Chiasma, Visual field defect, Ocular motility defect.

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I. Introduction

Pituitary adenoma is a benign tumor of pituitary gland, though it accounts for 10-15% of all primary intracranial tumors. [1] Pituitary adenoma can be classified as follows:

- ✓ *Size:* A) Macroadenoma (>10 mm). B) Microadenoma (<10 mm).
- ✓ Hormonal activity: A) Functional (secreting) tumor. B) Non-functional (non secreting) tumor.
- ✓ Staining pattern: A) Acidophilic (Stained with acid fuschin). B) Basophilic (Stained with aniline blue).
- C) Chromophobes (Contain agranular cytoplasm). [2]

75% of adenomas are endocrinologically secreting and produce clinical feature according to hypo/hypersecretion of hormones. [3][4].

Clinical manifestations: Hormonal deficiencies [5]:

Growth hormone deficiency	Gonadotrophin deficiency	Thyrotropin deficiency	ACTH deficiency	ADH deficiency
Adults: Obesity, Reduced exercise capacity, Increased cholesterol. Infants: Hypoglycaemia. Children: Decreased height and growth rate.	Men: Dimished Libido Impotence Women: Diminished libido, Dyspareunia, Breast atrophy. Children: Delayed or frank absence of puberty.	Malaise Weight gain Lack of energy Cold intolerance Constipation	Underactive adrenal gland causing low blood pressure, fatigue, weight loss, vomiting, stress intolerance	Diabetes insipidus

Hormonal overproduction [5] – Clinical effects:

Prolactine	Growth Hormone	ACTH [8]	
Women- Amenorrhoea, infertility	Children- Gigantism	Cushing disease- weight gain, central	
Men- Impotence, decreased Libido.	Adult- Acromegaly	obesity, moon facies, purple striae,	
		buffalo hump.	

25% of adenomas are non-secreting and create symptoms due to pressure effect over optic chiasma region. [6] Clinically it is presented as chiasmal syndrome with following symptoms [7]:

18 | Page

- Blurred vision.
- Headache.
- Diplopia.

- Colour desaturation.
- Visual field defect.
- Optic atrophy.
- Poor fixation blindness.
- Visual hallucination.
- See-saw nystagmus.
- Nausea.
- Vomiting.

Visual field defect in pituitary adenoma presented as:

- Bitemporal hemianopia.
- Incongruous homonymous hemianopia.
- Bitemporal central scotoma.
- Diffuse scotoma.
- Junctional scotoma.

Fundus examination by 90 D lens present with bow-tie atrophy, pale optic disc and temporal pallor [9]. The diagnosis is confirmed by endocrinological evaluation, visual field analysis& radiographic imaging of the pituitary [10]. Treatment options are

- Medical therapy- Cabergoline/ Bromocriptine (Dopamine agonists) in Prolactinoma. Octreotide in Growth Hormone secreting tumor.
- Surgery- Endoscopic craniotomy (Transsphenoidal, Tran frontal).
- Radiotherapy.
- > Gamma knife stereotactic radiotherapy.

	Surgery	Radiotherapy	Medical
Non-secreting adenoma	1 st line	2 nd line	-
Prolactinoma	2 nd line	2 nd line	1 st line
Acromegaly	1 st line	2 nd line	2 nd line
Cushing's disease	1 st line	2 nd line	-

This was the first case report of non-secretory pituitary macroadenoma with bilateral sudden loss of vision in AGMC, which was diagnosed by MRI and treated with transsphenoidal resection of pituitary. Complications might include vision loss & pituitary hormone insufficiency.

II. Case Presentation

A 14 years male muslim boy presented to Department of Ophthalmology in AGMC with bilateral sudden loss of vision for 1 week. He was apparently alright 1 week back. Then he suddenly developed severe headache, which was associated with 2 episodes of vomiting. Headache was present in frontal area, throbbing in nature and bilateral. Vomiting was non-projectile, non foul smelling & contained ingested food material. It was followed by sudden loss of vision, which was painless and bilateral. There was history of gradual painless diminished vision, seeing double objects & difficulty in recognizing colours for last 3-4 years. He was unable to see the writings in school blackboard. There was history of throbbing headache on & off, for which he was taking over the counter pain medications. Non-projectile vomiting was present on & off too, which usually followed by episodes of headache.

Visual acuity was PL positive only in both eyes not improving with pin hole. RAPD was present in both eyes though other anterior segment findings were normal. Fundus examination with 90D lens showed temporal pallor in both eyes suggesting partial optic atrophy (Fig 1).



Fig 1: Temporal pallor with pale optic disc (Bilateral).

Medial rectus and Inferior Oblique was partially affected in left eye. Visual field and colour vision could not be elicited due to severely diminished visual acuity (Fig 2).



Fig 2: Partial involvement of Medial rectus and Inferior oblique (LE).

Along with the routine haematological tests, hormonal panel was done including Serum prolactine, serum growth hormone and thyroid profile, which was within normal limit.

Serum TSH	2.4 U/ml. [0.3-5 U/ml]
Serum FT3	3.1 mcg/dl. [1.8-5.4 mcg/dl]
Serum FT4	6.8 mcg/dl. [4.5-11.7 mcg/dl]
Serum Prolactine	7 ng/ml. [2-18 ng/ml]
Serum Growth Hormone	3.1 ng/ml [<5 ng/ml]

X-ray skull (Fig 3) showed erosion of sella turcica, which pointed toward pituitary mass.



Fig 3: X-ray skull: Erosion of Sella Turcica (Arrow).

3-T high field MRI of brain and orbit suggested a solid cystic seller-supraseller mass with local mass effect compressing and displacing the optic nerve and optic chiasma measuring approx. 3.1 cm (AP) x 3.45 cm (TRANS) x 4.7 cm (CC) (FIG 4).

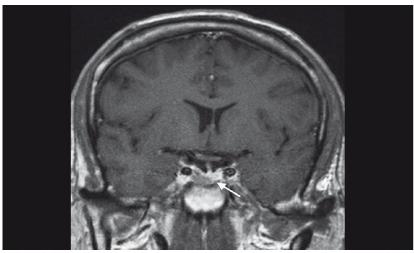


Fig 4: MRI scan with gadolinium illustrates a pituitary macroadenoma within a deformed sella turcica (Arrow).

After consultation with endocrinologist and neurologist, patient was diagnosed as non-secreting pituitary macroadenoma (Fig 5). After proper counselling, patient underwent transsphenoidal resection of pituitary and advised for regular follow-up. In follow-up visits along with ophthalmological examination, MRI brain and orbit was also done.

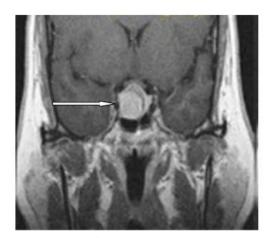


Fig 5: MRI (Before Operation)

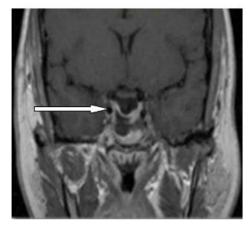


Fig 6: MRI (After operation)

6 month follow-up visit showed improved visual acuity by 6/60 in right eye and 5/60 in left eye. RAPD was absent bilaterally. Medial rectus and Inferior oblique which was involved prior to surgery became normal. Fundus still showed temporal pallor with pale optic disc. Colour vision testing by Ishihara chart was normal in both eyes. MRI revealed clear pituitary zone (Fig 6). Visual field testing by Humphrey field analyser using 24-2 SITA faster strategy was normal bilaterally (Fig 7).

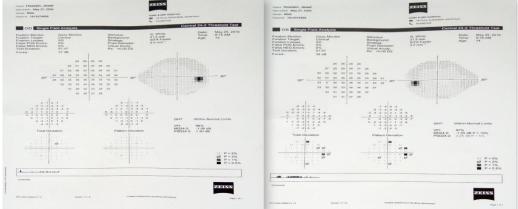


Fig 7: Humphrey 24-2 SITA faster field analyser showing normal visual field (BE).

III. Conclusion

Commonly presenting symptoms 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 such patients.

Presence of RAPD in both eyes suggested optic nerve pathology. Fundus examination with 90D lens at the slit lamp in this patient showed temporal pallor with pale optic disc in both eyes which suggested bilateral partial optic atrophy.

Extra ocular muscle (Medial rectus and Inferior Oblique) was partially involved in left eye as the mass displaced and compressed the bilateral optic nerve, optic chiasma and superiorly effacing the floor of third ventricle.

Routine X-ray Skull can show erosion of sella turcica which can point toward pituitary mass.

MRI scan is always a superior diagnostic modality than a CT-scan in detecting seller and supraseller lesions. An opinion of the endocrinologist and a neurologist is valuable and utmost urgency in an evidence of seller and supraseller lesions.

In non-secreting pituitary macroadenoma 1st line treatment is surgical resection of pituitary. 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.

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