Endogenous Steroid Induced Glaucoma due to Cushing Syndrome

Kirti Singh*, Sonal Dangda, Nitasha Ahir and Mainak Bhattacharyya

Guru Nanak Eye Centre & Maulana Azad Medical College, Maharaja Ranjit Singh Marg, New Delhi, India

Abstract: Exogenous steroid usage is a documented cause of increase in intraocular pressure in susceptible individuals; with systemic use having a lower propensity than topical. Endogenous hypercortisolism is an uncommon cause of steroid induced glaucoma with few reported cases due to adrenal adenoma, ectopic ACTH production in para-neoplastic syndromes, and pituitary adenomas. We report a case of steroid induced glaucoma occurring due to adrenal hyperplasia with confounding feminizing features which caused a diagnostic dilemma.

Keywords: Steroid induced glaucoma, endogenous, adrenal hyperplasia, Cushing's syndrome.

CASE REPORT

A 22 year old male patient was referred for a neurological field examination suspecting pituitary tumour. The patient had a documented weight gain with development of pot belly, easy bruisability and changes in facial contour over the last 6 years. He also complained of enlargement of breast tissue along with impotence for the same duration. There was no history of systemic steroid abuse in the form of anabolic steroids, herbal preparations (which often contain exogenous steroids) or steroid skin creams which can cause the above features.

On systemic examination patient had a plethoric moon face, increased adiposity of abdomen and interscapular regions, abdominal striae, bilateral gynaecomastia, and small non erectile penis with evidence of acute depression (Figure **1**). Neurological exam including cranial nerves was normal. Ocular examination revealed a best corrected visual acuity of 20/30 OU. The intraocular pressure (IOP) noted was 27 mmHg OD and 26 mmHg OS. Gonioscopy revealed completely open angles and optic nerve head examination showed a cupping of 0.8:1(DDLS stage 8) (Figure **2a,b**). Visual fields on Humphrey perimeter documented a double arcuate scotoma OU with a macular split in right eye (Figure **3a,b**).

Haematological studies revealed normal renal and liver functions with abnormal glucose tolerance test. Hormonal assays documented raised serum cortisol levels of 50 µg/dl (normal range 5-15µg/dl) with decreased testosterone levels of 1.7 ng/ml (normal range in males 2.4-12 ng/ml) and elevated Luteinizing hormone (LH) 40 mIU/ml (normal 1.5-8 mIU/ml), Follicle stimulating hormone (FSH) 110 mIU/ml (normal 6-24 mIU/ml), and Prolactin 21 ng/ml (18ng/ml being the higher range in men). A provisional diagnosis of Cushing's syndrome leading to hypergonadotrophic hypogonadism with steroid induced glaucoma was made.

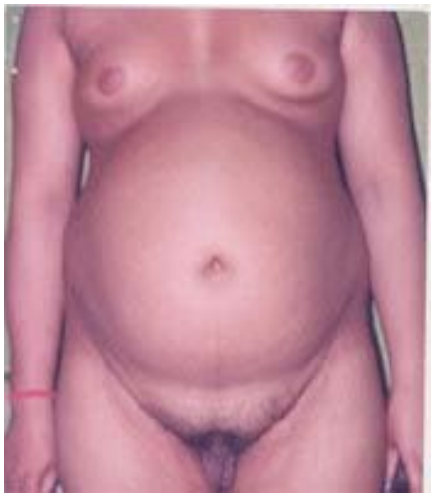
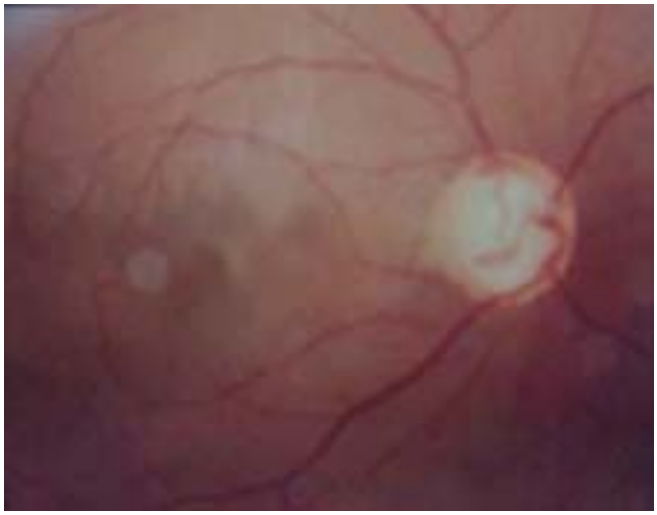


Figure 1: Shows increased adiposity of abdominal region with bilateral gynaecomastia and a small non erectile penis.



*Address correspondence to this author at the Guru Nanak Eye Centre & Maulana Azad Medical College, Maharaja Ranjit Singh Marg, New Delhi, India; Tel: +919968604334; E-mail: mail4kirti@gmail.com

E-ISSN: 2309-6136/14

Figure 2 Continue ……

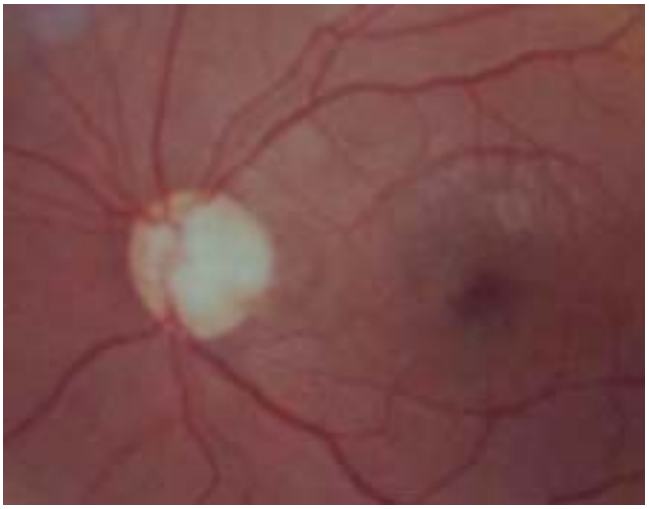


Figure 2a,b: Show an average sized disc with a cupping of 0.8 OU.

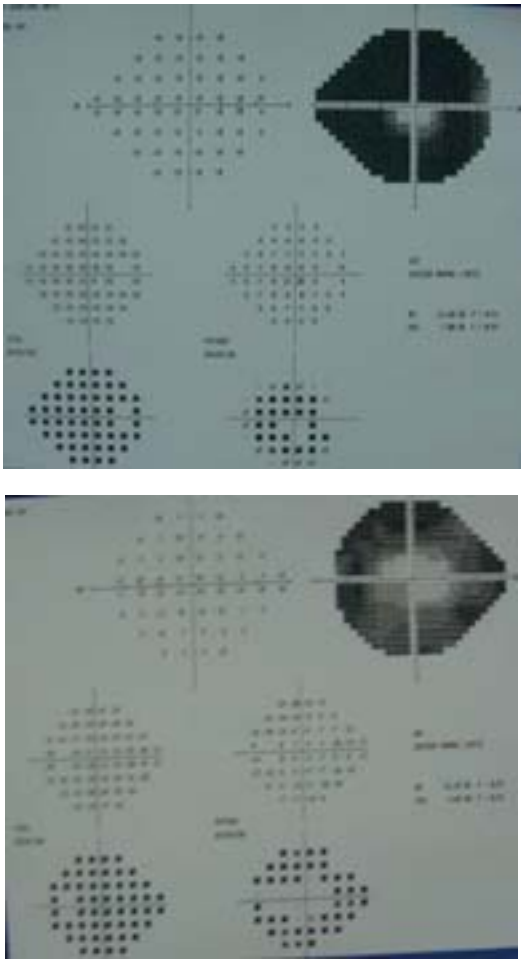


Figure 3a,b: Show a double arcuate scotoma OU.

Contrast imaging studies of brain, chest and abdomen by magnetic resonance imaging & computerized tomography were normal; without any evidence of pituitary, adrenal or para-neoplastic lesion. Over a 10 year irregular follow up, repeat imaging studies at 6 and 12 months and subsequently at 5 and 10 years were normal despite elevated cortisol levels, thus confirming adrenal hyperplasia (Figure **4**).

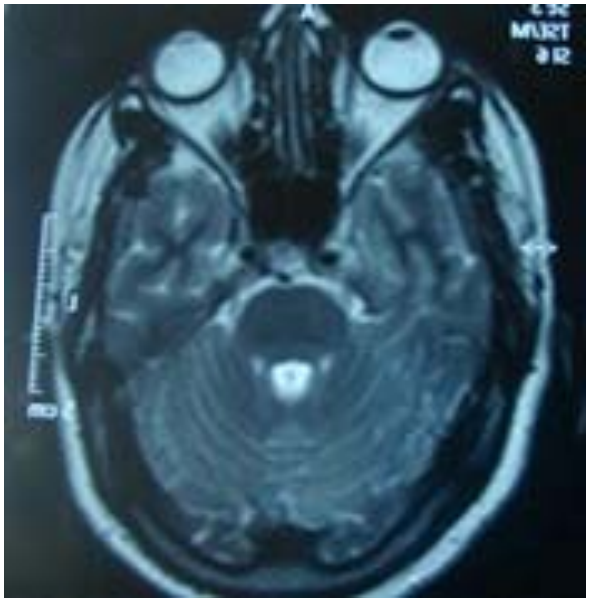


Figure 4: MRI section of pituitary area which revealed a normal scan, 10 year follow up.

Patient was put on anti-glaucoma medications and IOP was controlled within 2 weeks to 18 mmHg OU. To confirm steroid as the trigger for the IOP rise, the patient was subjected to topical glucocorticoid provocative test in left eye after informed consent. The test done with 0.1% dexamethasone was strongly positive within 5 days. Co-efficient of outflow (c-value) recorded by tonography was 0.09 μl/min/mmHg OD and 0.04μl/min/mmHg OS which is significantly less than normal value of 0.28±0.05.

The patient was discharged on topical anti-glaucoma medications, but within 3 months his psychiatric problems coupled with his socioeconomic status led him to discontinue the therapy. Keeping in mind his extremely poor adherence to medications coupled with advanced glaucoma damage, he was advised glaucoma filtering surgery. Trabeculectomy augmented with Mitomycin C was performed for both eyes within a span of 8 weeks. Over a follow up of 10 years the glaucoma is under control with IOP ranging from 12-16 mmHg OU. Both eyes have diffuse, polycystic blebs grade $E_3H_2V_0$ (IBAGS classification). No lenticular changes have been documented till the last follow up (Figure **5**). No deterioration in visual field or optic nerve head status has been documented.

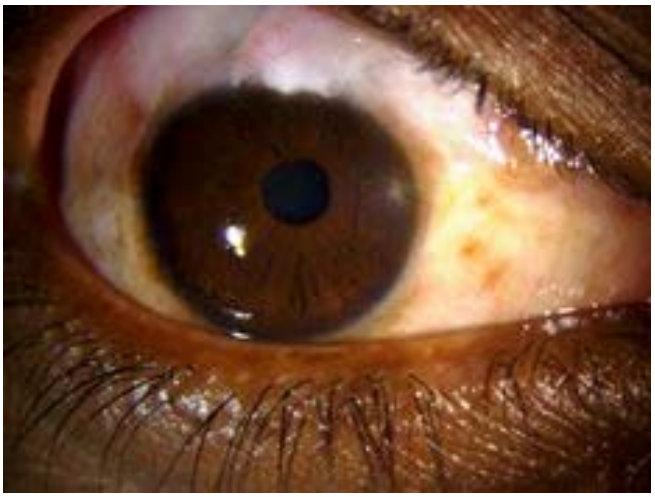


Figure 5: Slit lamp photograph of right eye showing diffuse cystic bleb.

DISCUSSION

It is well established that corticosteroids increase the intraocular pressure by decreasing the outflow facility [1]. This is further substantiated by the diurnal variation of IOP which follows the blood level surge of steroids by a time lag of 3-4 hours emphasizing that endogenous steroid levels play a role in the physiological control of IOP [2]. Also long term usage of systemic steroids has a propensity for IOP elevation [1,3]. However, serum cortisol alone is not responsible for glaucoma as elaborated by Neuner *et al.* who studied a series of patients with Cushing's syndrome and reported glaucoma in only one fourth, thus indicating that development of glaucoma may be dictated by a genetic susceptibility coupled with elevated serum cortisol [4]. The glucocorticoid excess state causing Cushingoid features in this case was coupled with the genetic susceptibility to steroids as confirmed by a positive steroid provocation test, a similar finding previously documented by Haas *et al.* in a patient of benign adrenal adenoma with glaucoma who was subsequently followed up for 16 years [5,6].

In the preliminary work up of a case of Cushing disease imaging studies of brain and body need to be performed to rule out pituitary or adrenal lesions; the common causes of this disease entity, and also ectopic ACTH secreting tumours [7,8]. Multiple imaging studies repeated over a decade in this case failed to document any such lesion thus making a diagnosis of adrenal hyperplasia. The feminizing features with ensuing psychiatric co-morbidities like depression and paranoia are features unique to this patient which led to a delay in diagnosis and made adherence to therapy problematic.

CONCLUSION

A high index of suspicion for endogenous steroid glaucoma must be kept in patients with Cushing's syndrome so that timely intervention can prevent permanent visual impairment.

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Received on 04-10-2014 Accepted on 09-10-2014 Published on 31-12-2014

DOI: http://dx.doi.org/10.12974/2309-6136.2014.02.02.1