Case Report

Systemic steroids in pediatric pseudotumor cerebri – A case report

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Abstract

Study design: Case report.

Purpose: To study the efficacy of systemic steroids in pediatric pseudotumor cerebri.

Introduction: Pseudotumor cerebri is a condition caused by elevated intracranial pressure presenting most commonly with headache. It is a diagnosis of exclusion.

Methods: A 15-year-old girl presented in our outpatient department with a headache for 3 months, more in the posterior aspect, continuous type with severe intensity, non-radiating. On examination, the best corrected visual acuity in her right eye was 6/9, and her left eye was 6/6 parts. She had a normal anterior segment and normal Intraocular Pressure (IOP). Fundoscopy showed both optic discs edematous, pink in color with blury disc margins, tortuous disc vessels, and edematous peripapillary area. The rest of the retina was normal. Lumbar puncture showed elevated opening CSF pressure and normal CSF composition. Hence she was diagnosed with IIH. She was started on oral prednisolone 1mg/kg bw once a day dosage, oral acetazolamide 250 mg once a day, topical nepafenac 0.1% one drop thrice a day and timolol 0.5% eye drops twice a day.

Discussion: The main goals of treatment are alleviation of symptoms, including headache, and preservation of vision. Steroids were commonly used for treating IIH in the past, but cause significant long-term side effects, such as weight gain, that are undesirable in IIH patients. Furthermore, withdrawal of steroids can cause rebound intracranial hypertension. Thus, steroids should not be used routinely for IIH treatment.

Results: By the end of one month, the patient was relieved of symptoms, best corrected visual acuity was 6/6 in both eyes with normal IOP, normal optic disc in both eyes.

Conclusion: Steroids are an effective way of managing idiopathic intracranial hypertension in pediatric patients.

Introduction

Idiopathic intracranial hypertension (IIH, pseudotumor cerebri) is a syndrome of elevated intracranial pressure of unknown cause that occurs predominantly in obese women of childbearing age. It is a diagnosis of exclusion and, therefore, other causes of increased intracranial pressure must be sought with history, imaging, and cerebrospinal fluid examination before the diagnosis can be made. Headache is the most common symptom reported by IIH patients at presentations [1,2].

Case report

A 15-year-old girl presented in our OPD at Manjunatha Eye Hospital, Kundapura, Udupi with a headache for 3 months, more in the posterior aspect, continuous type with severe
intensity, non-radiating. On examination, the best corrected visual acuity in her right eye was 6/9, and her left eye was 6/6 parts. She had a normal anterior segment and normal intraocular pressure in both eyes. Fundus examination showed both optic discs edematous, pink in color with blurry disc margins, tortuous disc vessels, and edematous peripapillary area. Normal foveolar reflex with the normal retinal periphery (Figure 1a,b).

The patient had been evaluated in a tertiary care hospital and her blood reports were normal. Neurological evaluation was also done and a lumbar puncture showed high opening pressure of CSF with normal CSF composition. Hence pseudotumor cerebri was diagnosed. She was using oral acetazolamide 250 mg three times a day for a period of one month which showed no improvement.

She was started on oral prednisolone 1 mg/kgbw once a day dosage, oral acetazolamide 250 mg once a day, topical nepafenac 0.1% one drop thrice a day and timolol 0.5% eye drops twice a day for a period of 2weeks. On follow-up, the patient was relieved of symptoms, and disc edema was also reduced in the next visit. Later her oral prednisolone was tapered on a weekly basis and the rest of the medications were continued.

By the end of one month, the patient was relieved of symptoms, the best corrected visual acuity was 6/6 in both eyes with normal IOP, and the left optic disc was normal with regular well-defined margins and normal vessels. The right optic disc was mildly edematous with ill-defined nasal margins and the rest of the margins were well defined. The patient was advised to continue oral prednisolone in weekly tapering doses with topical anti-inflammatory and anti-glaucoma agents. By the end of 2 months, the patient was completely fine with normal vision and a normal optic disc with well-defined margins (Figure 2a,b). She was followed up for a period of 12 months on a 2-monthly basis and was found to maintain a normal state.

Informed consent has been taken from the patient and the patient’s mother.

Discussion

The main goals of treatment are alleviation of symptoms, including headache, and preservation of vision [3]. Steroids were commonly used for treating IIH in the past, but cause significant long-term side effects, such as weight gain, that are undesirable in IIH patients. Furthermore, withdrawal of steroids can cause rebound intracranial hypertension [4,5]. Thus, steroids should not be used routinely for IIH treatment. High-dose intravenous steroids can, however, be useful for short-term treatment of patients with fulminant disease while a more definitive intervention (e.g., optic nerve sheath fenestration or cerebrospinal fluid shunting) is awaited [6–10]. There are other studies supporting the safe use of acetazolamide up to 4 g daily with weight loss for effective treatment of mild vision loss in IIH, with associated improvements in papilledema, increased intracranial pressure, and quality of life [8–15].

In our study, the patient was already on acetazolamide for a long duration but did not show any signs of improvement in symptoms or disc edema.

Conclusion

Steroids are an effective way of management of idiopathic intracranial hypertension in pediatric patients not responding to acetazolamide single therapy. This also avoids the risk of surgical intervention in pediatric patients.

References


