

Pseudotumor cerebri associated with Pickwickian syndrome and obstructive sleep apnea syndrome: a case report

Ozlem Barut Selver¹, Meltem Soylev Bajin¹, Oya Itil², Ali Osman Saatci¹

¹Department of Ophthalmology, Dokuz Eylul University, Izmir, Turkey

²Department of Pulmonary Medicine, Dokuz Eylul University, Izmir, Turkey

Correspondence to: Ali Osman Saatci. Department of Ophthalmology, Dokuz Eylul University, Izmir, Turkey. osman.saatci@deu.edu.tr

Received:2010-11-22 Accepted:2011-03-14

Abstract

• **AIM:** To report a male patient with Pickwickian syndrome and obstructive sleep apnea (OSA) who presented with pseudotumor cerebri and visual loss.

• **METHODS:** Case report.

• **RESULES:** A 54-year-old obese man with a three-month history of bilateral visual deterioration was evaluated. His visual acuity was 20/200 in OD and 20/400 in OS. Color vision was impaired only in the left eye. Funduscopy revealed bilateral disc edema and peripapillary hemorrhages together with macular exudates in OS. Physical examination and laboratory investigations were consistent with OSA and Pickwickian syndrome. Lumbar puncture demonstrated an opening pressure of 350mm H₂O and computed brain scan was normal. The diagnosis was pseudotumor cerebri in association with OSA and Pickwickian syndrome. Fundus findings and visual acuity improved with phlebotomy, blood pressure regulation, weight reduction and bi-level positive airway pressure therapy.

• **CONCLUSION:** OSA and Pickwickian syndrome should be kept in mind when facing a patient with pseudotumor cerebri.

• **KEYWORDS:** chronic respiratory insufficiency; obstructive sleep apnea syndrome; papilledema; Pickwickian syndrome

DOI:10.3969/j.issn.1672-5123.2011.04.005

Selver OB, Bajin MS, Itil O, Saatci AO. Pseudotumor cerebri associated with Pickwickian syndrome and obstructive sleep apnea syndrome: a case report. *Guji Yanke Zazhi (Int J Ophthalmol)* 2011;11(4):581-583

INTRODUCTION

Pseudotumor cerebri most often occurs in young obese women without any systemic disorder but nearly 10% of patients, pseudotumor cerebri may occur in men and nonobese people in whom there may be associated systemic disorders^[1].

Moreover papilledema occurs in almost 10% of patients with chronic respiratory insufficiency^[2]. In chronic respiratory disorders such as Pickwickian syndrome, hypercapnia and hypoxia may induce vasodilatation and an increase in cerebrospinal fluid (CSF) production. They result in an elevated intracranial pressure and papilledema or in obstructive sleep apnea (OSA), episodic increases in CSF pressure due to intraabdominal and intrathoracic pressure fluctuations with each apneic event may lead to papilledema^[1,3]. Key features of Pickwickian syndrome as currently accepted include obesity (body mass index $\geq 30\text{kg/m}^2$), chronic alveolar hypoventilation leading to daytime hypercapnia and hypoxia ($\text{Pa}_{\text{CO}_2} \geq 45\text{mmHg}$ and $\text{Pa}_{\text{O}_2} < 70\text{mmHg}$) and sleep disordered breathing^[4]. Pickwickian syndrome is frequently associated with obstructive sleep apnea (OSA) which is a condition characterized by repeating episodes of partial or complete obstruction of the airways resulting in the cessation of breathing for 10 seconds or longer during the sleep^[5]. Bi-level positive airway pressure (Bi-PAP), acetazolamide, oxygen therapy and weight reduction are treatment options for these kind of respiratory diseases^[6,7].

We hereby report a male patient with Pickwickian syndrome and OSA who presented with pseudotumor cerebri and visual loss.

CASE REPORT

A 54-year-old male patient with Pickwickian syndrome and OSA was presented with pseudotumor cerebri, visual loss with bilateral optic disk swelling and he referred to us for further evaluation. He was experiencing visual decline for the past three months. Recently computed tomography of the brain was normal. He was a heavy smoker for over 20 years and had poorly regulated blood pressure. He admitted having drowsiness during daytime and labored breathing. Ocular examination revealed reactive pupils without a relative afferent pupillary defect. Visual acuity was 20/200 in OD and 20/400 in OS. Color vision was 19/21 in OD and 2/21 in OS with Ishihara's pseudoisochromatic plates. Slit-lamp examination and intraocular pressure were normal. On funduscopy, there was severe bilateral optic disc edema and peripapillary hemorrhages with hard exudates at the left macula (Figure 1). Automated static perimetry showed generalized depression and blind spot enlargement in OU (Figure 2).

CSF analysis was normal, but the opening pressure was 350mmH₂O. Systemic evaluations and extensive laboratory investigations were carried out. Important findings were summarized.

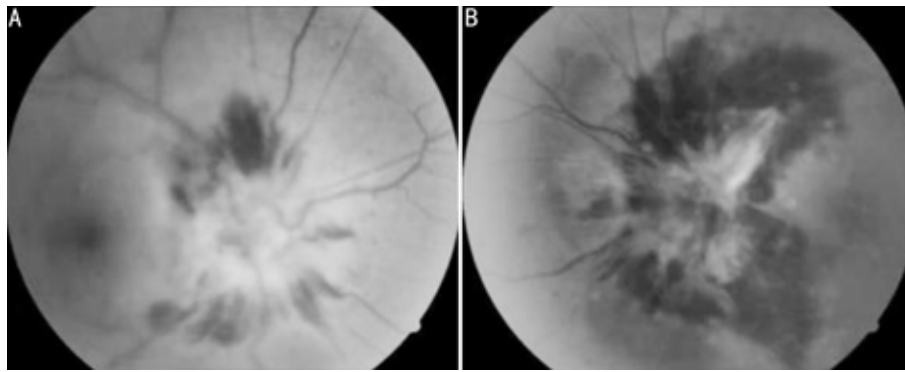


Figure 1 Funduscopy A; Optic disc edema and peripapillary hemorrhages (OD); B; Macular hard exudates accompanying optic disc edema (OS).

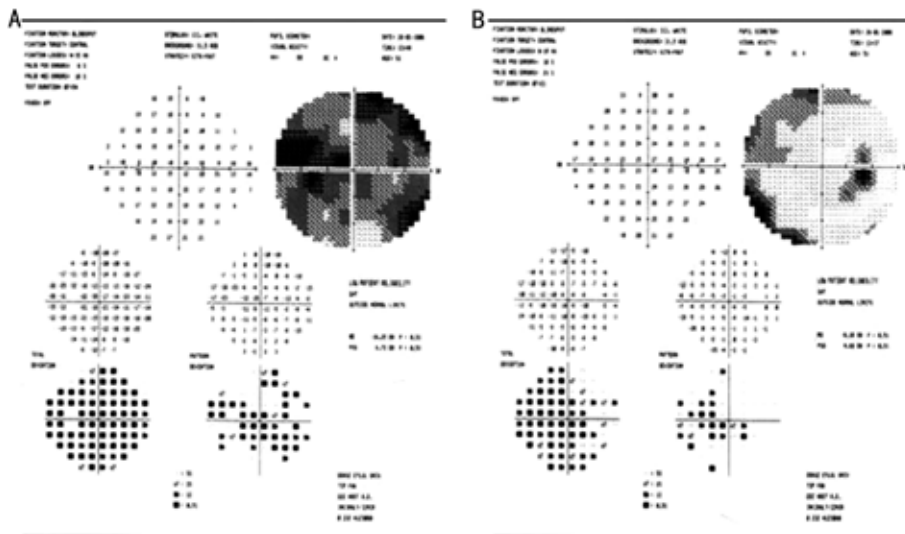


Figure 2 Automated static perimetry examination A, B; Generalized depression and blind spot enlargement in OU.

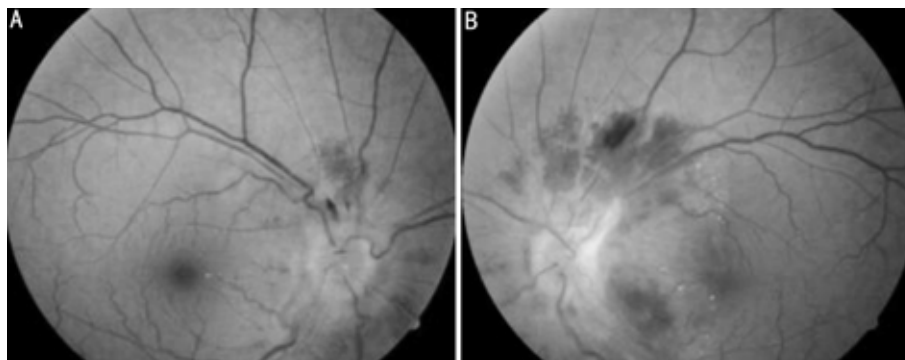


Figure 3 Funduscopy showing marked improvement in disc appearance two months later A; OD; B; OS.

His weight was 132kg and body mass index was 42.71. Hemoglobin concentration was 194g/L, and hematocrit was 59%. Arterial blood gas showed pH 7.41, Pa_{CO2} of 43.7 and Pa_{O2} of 55.1mmHg. Serum HCO₃⁻ was 28.5mmol/L. His chest X-ray was within normal limits. Thyroid function tests were normal. Otolaryngological examination showed total retropalatal and retrolingual obstruction. Our diagnosis was pseudotumor cerebri associated with sleep apnea and Pickwickian syndrome. Nocturnal polysomnography confirmed the diagnosis.

The patient was put on a weight reducing diet, blood pressure was regulated, phlebotomy was performed and Bi-PAP therapy was commenced. Two months later, optic disc appearance was slightly improved (Figure 3) and visual acuity was 20/20 in

OU. Three years later, optic disc was almost normal looking in OU (Figure 4).

DISCUSSION

In Pickwickian syndrome chronic with hypercapnia may precipitate elevated intracranial pressure by inducing vasodilatation and an increase in CSF production. Additionally hypoxemia may increase cerebral blood flow^[1]. On the other hand, episodic increases in CSF pressure are observed in patients with OSA in concordance with intraabdominal and intrathoracic pressure fluctuations with each apneic event^[3]. The incidence of visual loss in patients with idiopathic pseudotumor cerebri is only 23%. Notingly Purvin *et al*^[8] believe that patients with OSA related pseudotumor cerebri seem to be at particular high risk for developing visual loss

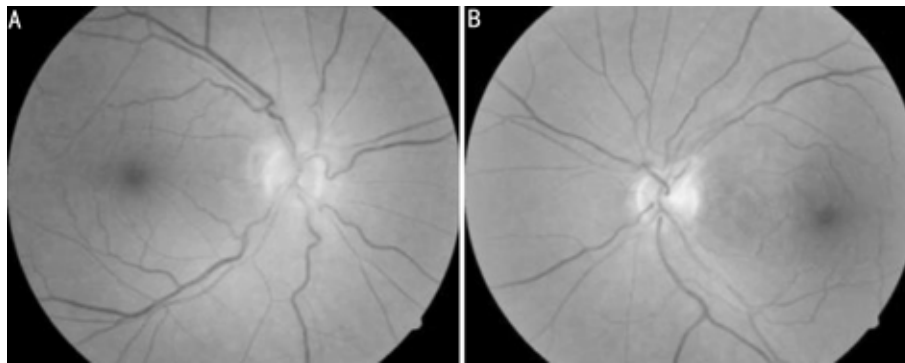


Figure 4 Funduscopy showing almost normal looking discs OU 3 years later A: OD; B: OS.

compared to patients with typical pseudotumor cerebri and they attributed this to nocturnal oxygen desaturation.

Mojon *et al*^[9] postulated several potential mechanisms for optic neuropathy in OSA (1) impaired optic nerve head blood flow autoregulation secondary to apneic episodes; (2) failure of optic nerve autoregulation because of arterial hypertension and arteriosclerosis (3) prolonged hypoxic damage to the optic nerve (4) imbalance between vasodilators and vasoconstrictors. Prevalence of papilledema in patients with OSA syndrome remains unknown. Peter *et al*^[10] prospectively evaluated 95 successive patients with recently diagnosed OSA syndrome and performed funduscopy. None of the patients had papilledema. Authors stated that systematic screening for papilledema does not seem to be warranted in this group of patients.

Lee *et al*^[6] retrospectively reviewed 32 male patients with idiopathic intracranial hypertension and six of the cases had sleep apnea syndrome. All six patients had optic disc edema and enlarged blind spots. Of these six patients, one received acetazolamide alone, four received acetazolamide and continuous positive airway pressure and one was treated with continuous positive airway pressure. Authors commented that treatment of OSA with nocturnal oxygenation may improve the signs and symptoms of pseudotumor cerebri in affected men. Wolin *et al*^[7] described a 46-year-old extremely obese black woman with papilledema who was diagnosed as Pickwickian syndrome and OSA. They treated the patient successfully with Bi-PAP, acetazolamide, oxygen therapy and weight reduction with improvement of papilledema over two months.

The presence of papilledema should be kept in mind in all patients with chronic respiratory insufficiency. Our case clearly demonstrates that pseudotumor cerebri may be a feature of Pickwickian syndrome and OSA. Visual impairment and papilledema could be reversed with weight reduction, Bi-BAP therapy, phlebotomy and blood pressure regulation in our case and we did not administer acetazolamide. Treatment strategies are somewhat different in pseudotumor cerebri cases related to chronic respiratory insufficiency when compared to patients with idiopathic variety.

REFERENCES

- 1 Miller NR, Hoyt WF. Walsh&Hoyt's Clinical Neuro-Ophthalmology. 5th(ed) Baltimore: Williams&Willkins 1997;487-548
- 2 Varelas PN, Spanaki MV, Rathi S, Cohen IS. Papilledema unresponsive to therapy in Pickwickian syndrome: another presentation of pseudotumor cerebri? *Am J Med* 2000;109:80-81

- 3 Sugita Y, Iijima S, Teshima Y, Shimizu T, Nishimura N, Tsutsumi T, Hayashi H, Kaneda H, Hishikawa Y. Marked episodic elevation of cerebrospinal fluid pressure during nocturnal sleep in patients with sleep apnea hypersomnia syndrome. *Electroencephalogr Clin Neurophysiol* 1985;60:214-219

- 4 Mokhlesi B, Kryger MH, Grunstein RR. Assessment and management of patients with obesity hypoventilation syndrome. *Proc Am Thorac Soc* 2008;15:218-225

- 5 Dhillon S, Shapiro CM, Flanagan J, Cameron AJ. Papilledema in pulmonary emphysema. Sleep-disordered breathing and effects on ocular health. *Can J Ophthalmol* 2007;42:238-243

- 6 Lee AG, Golnik K, Kardon R, Wall M, Eggenberger E, Yedavally S. Sleep apnea and intracranial hypertension in men. *Ophthalmology* 2002;9:482-485

- 7 Wolin MJ, Brannon WL. Disk edema in an overweight woman. *Surv Ophthalmol* 1995;39:307-314

- 8 Purvin VA, Kawasaki A, Yee RD. Papilledema and obstructive sleep apnea syndrome. *Arch Ophthalmol* 2000;118:1626-1630

- 9 Mojon DS, Mathis J, Zulauf M, Koerner F, Hess CW. Optic neuropathy associated with sleep apnea syndrome. *Ophthalmology* 1998;105:874-877

- 10 Peter L, Jacob M, Krolak-Salmon P, Petitjean T, Bastuji H, Grange JD, Vighetto A. Prevalence of papilloedema in patients with sleep apnoea syndrome: a prospective study. *J Sleep Res* 2007;16:313-318

匹克威克综合征和阻塞性睡眠呼吸暂停综合征相关性假性脑瘤

Ozlem Barut Selver¹, Meltem Soylev Bajin¹, Oya Itil², Ali Osman Saatci¹

(作者单位:土耳其伊兹密尔, Dokuz Eylul 大学¹ 眼科;² 呼吸科)

通讯作者: Ali Osman Saatci. osman. saatci@deu. edu. tr

摘要

目的: 报告 1 例男性患者合并有匹克威克综合征和阻塞性睡眠呼吸暂停(OSA), 其临床表现为假性脑瘤和视力下降。

方法: 病例报告。

结果: 患者, 男, 54 岁, 过度肥胖, 诉双眼视力渐降 3mo, 检查示: 视力: 右眼 20/200, 左眼 20/400, 左眼合并色觉异常, 眼底镜示双视乳头水肿和视盘旁出血, 左眼合并黄斑部渗出。体格检查和实验室检查符合阻塞性睡眠呼吸暂停与匹克威克综合征诊断。腰椎穿刺: 开放压 350mmHg, 脑部扫描正常。诊断: OSA 和匹克威克综合征相关性假性脑瘤。眼底表现和视力随采血治疗、血压控制、体重控制及双层正压通气治疗而改善。

结论: 对于患有假性脑瘤的患者, 必须对阻塞性睡眠呼吸暂停与匹克威克综合征有所警惕。

关键词: 慢性通气不足; 阻塞性睡眠呼吸暂停综合征; 视神经乳头水肿; 匹克威克综合征