

Case Report

Mona lisa syndrome: a case report

Bulent Yilmaz^{1,2}, Beyazit Yildirim³, Melike Balikoglu-Yilmaz⁴, Ibrahim Egemen Ertas⁵, Ismail Onder Uysal⁶, Sefa Kelekci¹, Mustafa Demir^{1,*}

¹ Izmir Katip Celebi University, Atatürk Education and Research Hospital, Department of Obstetrics and Gynecology, Izmir, Turkey

² Imamoglu State Hospital, Obstetrics and Gynecology Clinic, Adana, Turkey

³ Imamoglu State Hospital, Otorhinolaryngology Clinic, Adana, Turkey

⁴ Dr. Behçet Uz Children Disease and Surgery Education and Research Hospital, Department of Ophthalmology, Izmir, Turkey

⁵ Tepecik Education and Research Hospital, Department of Obstetrics and Gynecology, Izmir, Turkey

⁶ Cumhuriyet University, Department of Otorhinolaryngology, Sivas, Turkey

Abstract

The mysterious smile of the famed portrait ‘The Mona Lisa’ has confused both art historians and researchers for many years. The nature of the model’s enigmatic smile has been explained as the smile of a new, joyous mother, Lisa Gherardini, who had developed Bell’s palsy during her recent pregnancy. Bell palsy which is acute in onset, unilateral, and is considered to be idiopathic is the most common cause of peripheral facial nerve paresis and paralysis. We report an idiopathic facial paralysis (Bell’s palsy) as Mona Lisa Syndrome in a 24 years old primigravid woman at 34 weeks of gestation.

Key words:

Bell’s palsy, Facial paralysis, Mona Lisa Syndrome, Pregnancy.

Introduction

The theory that the enigmatic smile of ‘The Mona Lisa’ was an artistic representation of the facial muscle contraction that develops subsequent to Bell’s palsy, when the facial nerve has undergone partial Wallerian degeneration and has regenerated, was introduced by Adour [1]. This proposal has garnered great attention by the medical community, since it has a high level of scientific credibility [2]. Bell’s palsy is an idiopathic unilateral facial nerve paralysis, often with a rapid onset. Throughout pregnancy the incidence of Bell’s palsy has been risen up to 3.3 times compared to nonpregnant women [3].

The precise etiology and associated factors as well as various treatment options aiming at the prevention of associated complications and improving recovery of facial nerve function have intensively been appraised over the past 3 decades. But, therapeutic approach, particularly in pregnant women, remains undecided. Corticosteroids which are applied after the first trimester might be useful

[4]. Here is the presentation of a pregnant woman affected by Bell’s palsy with the smile of the portrait’s model was due to Leonardo da Vinci’s anatomically precise representation as Mona Lisa Syndrome at third trimester of pregnancy.

Case presentation

A 24 years old primigravid woman at 34 weeks of gestation was attended to Obstetric and Gynecology Clinic of Health Ministry Imamoglu State Hospital complaining of the asymmetria on her face especially while smiling and inability to close right eyelids. After consultation of patients to an otorhinolaryngologist and an ophthalmologist, it was diagnosed as Bell’s palsy. Vitamin B complex containing Vitamin B1, B6 and B12 and artificial tears containing dextran 70 and hydroxypropyl methyl cellulose were given to patient at diagnosis and trimetazidine dihydrochloride was started after delivery. She delivered a male 3950 gr baby with good APGARS and cord gases at 41 weeks gestation via cesarean section. She almost has got recovery after 2 months of delivery.

Article history:

Received 20 April 2014

Accepted 22 April 2014

*Correspondence: Mustafa DEMIR, M.D.

Maliyeciler Mahallesi, 9200/1 Sokak, Çamlıkent Sitesi, No:5/25

Karabağlar, 35600, Izmir, Turkey

Phone: +90 505 759 80 52 | E-mail: musdem27@gmail.com

Discussion

Peripheral neuropathies during pregnancy and the postpartum period are frequent. One of the most common peripheral neuropathies is Bell's palsy [5]. Bell's palsy or weakness of the facial nerve, or cranial nerve VII, is unilateral lower motor neuron lesion of the facial nerve causing facial weakness, including loss of frontalis muscle on the affected side that is usually reversible [6]. It was first defined by Sir Charles Bell, who studied the facial nerve, in 1830 [7]. The incidence of Bell's palsy is indicated to be 45/100,000 pregnancies, but only 17/100,000 in nonpregnant women of childbearing age [8].

There is raised incidence during the third trimester (71-89.5%), followed by 9.8-21.3% in the puerperium, 5.3% in the first trimester, and 2.4-2.6% in the second trimester [9]. Shmorgun et al. [10] demonstrated higher rates of Caesarean section (43.6%), preterm delivery (25.6%), and low infant birth weight of less than 2.500 g (22.7%); all were comparably higher than expected for the general obstetric population. However, Katz et al. [11] did not find a relationship between Bell's palsy and adverse perinatal outcomes such as low Apgar scores (<7) at 5 minutes (4.8% vs. 3.1%), perinatal mortality (2.4% vs. 1.4%), or congenital malformations (4.8% vs. 5.7%) as the case in our case report.

In the general population, possible causal reasons include viral infection [herpes simplex virus-1 (HSV-1), varicella-zoster virus], vascular ischemia, autoimmune inflammatory disorders, diabetes, Lyme disease, stroke, cerebellar-pontine angle or other tumor, temporal bone trauma, acute or chronic otitis media, cholesteatoma, arterial-venous malformation, myasthenia gravis, mumps, and facial neuroma and hereditary factors [11]. Independent risk factors for the development of Bell's palsy in pregnancy include chronic hypertension and maternal obesity [11]. Other potential etiologies were also reported to be HSV-1 reactivation because of an increased maternal immunosuppression; the increased interstitial edema of the third trimester and postpartum period leads to a venous congestion in the temporal bone fallopian canal containing the facial nerve or perineural edema; maternal hypercoagulability with consequent thrombosis of the vasa nervorum, changing estrogen and progesterone levels; and increased cortisol levels in the third trimester [9].

Swelling of the facial nerve within the petrous temporal bone causes Bell's palsy. This usually occurs only on one side of the face and may happen abruptly or progress over a few days. Symptoms may initially include pain behind the ear or hearing hyperacuity and can progress to include change in taste and salivation, dry eyes due to generally

dysfunction of lacrimation and a consequent inability to lubricate with natural tears, and difficulty with smiling, moving the face, or closing the eye. Corneal abrasions might be also seen due to lack of natural eye protection. A diagnose of Bell's palsy could be often made clinically. Craniofacial and neuroimaging include computed tomography of the temporal bone or magnetic resonance imaging along the intracranial and extracranial path of the facial nerve or other diagnostic testing including Lyme titers, lumbar puncture, and electroneuronography should be also considered [12].

Bell's palsy tends to resolve spontaneously, might show improvements within 2 weeks of the onset of initial symptoms and return to normal function within 3 to 6 months. The use of prednisolone for its treatment within 72 hours of the presentation of symptoms has been supported by recent meta-analyses of research databases and trials [13]. Steroids might be highly effective and increase the probability of facial nerve recovery and shorter the recovery time with the initiation of steroids, within 24 h of onset of symptoms. Dosing is variable, but prednisolone at a dose 1mg/kg for 5 days followed by an appropriate taper has been recommended [4].

Antivirals, nucleoside analogues including valacyclovir and famciclovir which are classified as pregnancy category B and pose little risk to either mother or fetus, can be offered as another adjunctive treatment for Bell's palsy. Other symptomatic treatment is intended for protecting vision and ocular structure. Preservative-free artificial tears (methylcellulose drops) and overnight ocular lubricants should be used to prevent dry eye, exposure keratitis, and corneal abrasions [4, 14]. Surgical decompression of the facial nerve may be rarely necessary. This might occur during pregnancy or after induction and delivery if the pregnancy has reached 39 weeks gestation. In cases where function has not returned by 6 to 16 months, hypoglossal-facial nerve anastomosis may restore partial function [15].

The Bell's palsy has an excellent prognosis in pregnant and postpartum women, with the majority (70% to 90%) of patients experiencing a full recovery in 3 weeks, whereas the remaining 15% to 30% experiences a full to partial functional recovery within 3 - 6 months [15]. In the setting of complete facial nerve paralysis, full recovery may only be seen in 50% of cases. Recurrence has been reported as rarely [15]. The production of endogenous steroids in pregnant women might affect recovery. This case report shows Bell's palsy in a pregnant woman with Mona Lisa Syndrome at third trimester of pregnancy. Every obstetrician should be aware of Bell's palsy, an enigma, during pregnancy and the puerperium. For these women, a maternal surveillance can be recommended. The surveillance for preeclampsia should also be mounted. A fast and accurate diagnosis with a subse-

quent immediate treatment might be very important to minimize its associated long-term sequelae.

Conflict of interest statement

The authors declare no conflict of interest.

References

1. Adour KK. Mona Lisa syndrome: solving the enigma of the Gioconda smile. *Ann Otol Rhinol Laryngol* 1989;98:196-9.
2. Hellebrand MC, Friebe-Hoffmann U, Bender HG, Kojda G, Hoffmann TK. Mona Lisa syndrome: idiopathic facial paralysis during pregnancy. *Z Geburtshilfe Neonatol* 2006;210:126-34.
3. Alvis JS1, Hicks RJ. Pregnancy-Induced Acute Neurologic Emergencies and Neurologic Conditions Encountered in Pregnancy. *Semin Ultrasound CT MR* 2012;33:46-54.
4. Vrabec JT1, Isaacson B, Van Hook JW. Bell's palsy and pregnancy. *Otolaryngol Head Neck Surg* 2007;137: 858-61.
5. Klein A. Peripheral nerve disease in pregnancy. *Clin Obstet Gynecol*. 2013;56:382-8.
6. Ragupathy K, Emovon E. Bell's palsy in pregnancy. *Arch Gynecol Obstet*. 2013;287:177-8.
7. Bell C. *The Nervous System of the Human Body*. London: Longman, Rees, Orne, et al.; 1830:4-5.
8. Falco NA, Erikson E. Idiopathic facial palsy in pregnancy and puerperium. *Surg Gynecol Obstet* 1989;169:337-40.
9. Pearce CF, Hansen WF. Headache and neurological disease in pregnancy. *Clin Obstet Gynecol* 2012;55:810-28.
10. Shmorgun D, Chan WS, Ray JG. Association between Bell's palsy in pregnancy and pre-eclampsia. *QJM* 2002;95:359-62.
11. Katz A1, Sergienko R, Dior U, Wiznitzer A, Kaplan DM, Sheiner E. Bell's palsy during pregnancy: is it associated with adverse perinatal outcome? *Laryngoscope* 2011;121:1395-8.
12. Maloney WJ. Bell's palsy: the answer to the riddle of Leonardo da Vinci's 'Mona Lisa'. *J Dent Res*. 2011;90:580-2.
13. Charn TC, Subramaniam S, Yuen HW. Bell's palsy in Singapore: a view from the patient's perspective. *Singapore Med J*, 2013;54:82-5.
14. Lorch M, Teach SJ. Facial nerve palsy: etiology and approach to diagnosis and treatment. *Pediatr Emerg Care*. 2010;26:763-769. quiz 770-3.
15. Cohen Y1, Lavie O, Granovsky-Grisaru S, Abouafia Y, Diamant YZ. Bell palsy complicating pregnancy: a review. *Obstet Gynecol Surv* 2000;55:184-8.