Bell's Palsy as an Unfavorable Effect of Growth Hormone Therapy in a Pediatric Patient with Turner Syndrome

Turner Sendromlu Çocuk Hastada Büyüme Hormonu Tedavisinin İstenmeyen Etkisi Olarak Bell Felci

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Abstract

Bell's palsy is defined as a lower motor neuron facial palsy of acute onset unaccompanied by evidence of an aural, a neurological or a local cause. Bell's palsy is an uncommon cause of facial palsy in children under 10 years of age. Here, we report a 9-year-old girl presented with unilateral facial palsy as an unfavorable effect of growth hormone therapy. To our knowledge, this is the first case report of an association of Turner syndrome with Bell's palsy. Although this may be a chance association only, the condition raised the question of whether growth hormone therapy in Turner syndrome may contribute to Bell's palsy. *Turk Jem 2007; 11: 105-7*

Key words: Turner syndrome, Bell's palsy, growth hormone therapy

Özet

Bell felci aural, nörolojik ya da lokal bir sebebin eşlik etmediği ani başlangıçlı alt motor nöronun yüz felci olarak tanımlanmaktadır. Bell felci 10 yaş altındaki çocuklarda yüz felcinin nadir bir sebebidir. Burada büyüme hormonunun istenmeyen etkisi olarak tek taraflı yüz felci olan 9 yaşındaki kız olguyu sunuyoruz. Bilgilerimize göre bu olgu Bell felcinin eşlik ettiği Turner sendromlu ilk olgudur. Bu yalnız tesadüfi bir birliktelik olsa bile Turner sendromunda büyüme hormonu tedavisi Bell felcine katkıda bulunan bir durum olabilir. *Turk Jem 2007; 11: 105-7*

Anahtar kelimeler: Turner sendromu, Bell felci, büyüme hormonu tedavisi

Introduction

Growth hormone therapy has been clearly shown to improve height velocity during childhood in a variety of pediatric conditions in which growth is compromised. Currently, growth hormone therapy has been recommended for a number of licensed indications including growth hormone insufficiency, Turner syndrome, growth failure associated with chronic renal disease, and most recently, for the growth hormone deficiency associated with Prader–Willi syndrome (1). However, growth hormone therapy has some significant complications, most notably insulin resistance and hyperglycemia.

The administration of growth hormone to mostly non-growth hormone-deficient short children with Turner syndrome has resulted in increased growth velocities. Despite the apparent beneficial effects of growth hormone in Turner syndrome, a number of issues remain unresolved and requiring further research. The long-term administration of pharmacologic growth hormone doses to short, however mostly non-growth hormone-deficient children must be viewed with caution, as long-term complications can not as yet be fully evaluated.

Bell's palsy or idiopathic facial paralysis is defined as a lower motor neuron facial palsy of acute onset unaccompanied by evidence of an aural, a neurological, or a local cause (2). It is typically unilateral and can be complete or partial. Bell's palsy is an uncommon cause of facial palsy in children under 10 years of age (3). Here, we report a 9-year-old girl with Turner syndrome occurred Bell's palsy as an unfavorable effect of growth hormone therapy.

Case Report

A 9-year-old girl presented with unilateral facial palsy. The patient was first examined at the age of 7 years because of short stature. At that time, her height was 104 cm (Z-score -2.87), her body mass

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index 16.8 kg/m2, and her bone age corresponded to that of a 6 year old girl. Dopamine stimulation test and insulin tolerance test revealed growth hormone deficiency with a normal cortisol response. Her insulin-like growth factor-1 level (655 ng/mL, normal range: 140-308 ng/mL) was over the limits according to her age and sex. The patient's thyroid function was within normal limits. The rest of the laboratory investigations were unremarkable. Ultrasound sonography of the patient's pelvis had showed a hypoplastic uterus, whereas the ovaries were not possible to visualize. A karyotype analysis was done and this was 45, X with no evidence of mosaicism. At the age of 7 years, the diagnosis of Turner syndrome was made on the basis of the patient's clinical features, the high concentrations of basal gonadotrophins, the lack of ovarian ultrasonographic visualization, and the karyotype analysis.

Growth hormone substitution therapy was initiated and continued for a period of 1.5 years. The present examination in our institution when the patient was aged 9 years revealed a normal weight young girl (body mass index 15.9 kg/m^2), with a central pattern of fat distribution. The patient's height was 116.5 cm (Z-score – 2.06). She was noted to have facial asymmetry with an unilateral peripheral facial nerve deficit (Fig 1a). No history of trauma or seizure was obtained. All other cranial nerves were intact and her blood pressure was 110/70 mmHg. She did not have any symptoms indicating the existence of otitis media and there was no rash or vesicle on her face. In the rest of the physical examination, there was no any remarkable finding, except the physical stigmata of Turner syndrome.

The present laboratory examinations including complete blood count, serum electrolytes, fasting blood glucose and insulin, liver function tests, lipid profiles, the cerebrospinal fluid and coagulation tests were all normal limits. Cerebral axial computed tomographic scan of patient did not reveal any intracranial abnormality or evidence of mastoiditis. Axial T1- and T2-weighted magnetic resonance imaging of the brain performed was normal. Growth hormone was discontinued and the patient was treated with oral hydrocortisone (10 mg/day) for a period 2 weeks. Facial nerve palsy led to decreased tearing and blinking, both of which contribute to drying of the cornea. We applied the artificial lubrication in the form of wetting solutions or ointments. At the 2-months follow up, she was doing well without growth hormone therapy and there was partially her left peripheral facial palsy. Eventually, the follow-up showed a complete resolution of the facial nerve deficit at the 3 months after the cessation of growth hormone (Fig 1b).

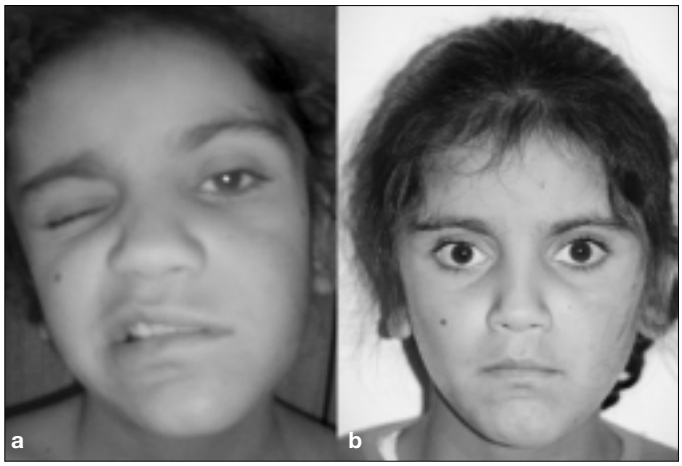

Figure 1. a) The characteristic face of Turner syndrome in the patient who had complete facial palsy with reduced elevation of left eyebrow and closure of left eye (with parents' consent). b) It is noted resolving the facial paralysis after the discontinued the treatment

Discussion

The safety profile of growth hormone treatment in Turner syndrome is good, with precautions related to risks of intracranial hypertension, type 2 diabetes, impaired glucose tolerance, and slipped capital epiphyses (4). However, two randomized controlled trials have demonstrated a doubling in the risk of otitis media with growth hormone treatment (5, 6). The patient had no physical finding of otitis media. This report described a patient with Turner syndrome occurred Bell's palsy as an unfavorable effect of growth hormone therapy.

Facial paralysis is uncommon in childhood and may be because of a divergent group of causes including otitis media, mastoiditis, abscess, osteopetrosis, trauma, tumor, Guillain-Barre syndrome, sarcoidosis, pseudobulbar palsy, leukemia, arteriovenous malformation, and Melkersson-Rosenthal syndrome (7-10). The patient presented with a rapid onset of unilateral facial palsy occurred over a few hours. There was no history of viral illness or trauma, and associated systemic symptoms to rule out traumatic, infectious, neoplastic or hematological causes. On the other hand, computed tomography and magnetic resonance imaging did not reveal any abnormality in our patient.

Bell's palsy is defined as an idiopathic peripheral facial nerve paralysis of sudden onset and is considered the most common cause of facial nerve paralysis. Generally, if the patient's presentation is typical and a thorough clinical examination does not reveal any obvious cause, no routine tests are needed to diagnose Bell's palsy. Investigations such as magnetic resonance imaging are needed only occasionally to rule out any other associated neurological problem. Facial nerve passes through the facial canal in the petrous temporal bone (intratemporal part) in close proximity to the medial wall of the inner ear and the mastoid cavity (3). Swelling and mechanical entrapment of the facial nerve may be contributing factors in the pathogenesis of facial nerve palsies. The facial nerve is most commonly damaged in the labyrinthine segment, where even mild edema may cause nerve compression (11). Growth hormone acts on the growth, metabolism and structure of bone and its effects can be direct or indirect, being mediated by insulin-like growth factor-I, which stimulates osteoblasts. Growth hormone therapy has different kinds of effects depending on the target structures (12,13). Rongen-Westerlaken et al. showed that growth hormone therapy particularly affects cartiloginous growth (14). Bell's palsy has been reported to be a complication of treatment with recombinant human insulin-like growth factor-I (15). The causes of facial palsy in our patient may be not only directly related the compression with the nerve caused by extrinsic swelling or pressure but also enlargement of bone structures due to growth hormone treatment.

The characteristic faces of a female with Turner syndrome is also primarily due to skeletal malformations. Many of the physical stigmata of Turner syndrome are a result of structural bone defects (16). However, the combination of estrogen replacement therapy

and growth hormone treatment results in a greater gain in bone mass (17,18). Sas et al. recently reported changes in body proportions of girls with Turner syndrome treated with growth hormone. They found that the increase in height after long term treatment was accompanied by an even higher increase in foot size and a moderate improvement of the disproportion between height and sitting height (19). In our patient, the disproportions in bone structures accompanied by growth hormone treatment might lead to a nerve compression in facial canal. However, further studies are necessary since it remains unclear. To our knowledge, this is the first case of Turner syndrome associated with facial palsy. Although this may be a chance association only, the condition raised the question of whether growth hormone therapy in Turner syndrome may contribute to Bell's palsy.

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