TEMPORARY MULTIPLE CRANIAL NERVE PALSY IN A PATIENT WITH TYPE 1 DIABETES MELLITUS

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SUMMARY - Remittent isolated palsy of peripheral or of upper cranial nerves in diabetic patients is well documented, but paralysis of a lower cranial nerve or an isolated branch of any cranial nerve has rarely been reported. In the case described, besides temporary hypoglossal and facial nerve palsies previously, unilateral temporary vocal cord palsy caused by right inferior laryngeal nerve (recurrent) paralysis associated with type 1 diabetes mellitus is presented. Hoarseness and vocal cord palsy of the patient, as in the case of her first admission with other complaints due to other cranial nerve palsies, totally remitted, presumably both owing to improved metabolic control.

Key-words: cranial nerve palsies, vocal cord palsy, type 1 diabetes mellitus.

RE´SUME - Paralysies multiples transitoires des nerfs crâniens chez un diabétique de type 1.
La survenue de paralysies isolées transitoires de nerfs périphériques ou de nerfs crâniens supérieurs chez le diabétique est bien documentée, mais celle d’un nerf crânien inférieur ou d’une branche isolée d’un nerf crânien quel qu’il soit a rarement été rapportée. Dans le cas présent, mise à part une paralysie du nerf hypoglosse et du nerf facial de survenue antérieure, nous rapportons la survenue d’une paralysie unilatérale transitoire d’une corde vocale causée par une paralysie du nerf laryngé inférieur droit (nerf récurrent) chez un diabétique de type 1. La dysphonie et la paralysie de la corde vocale, comme cela avait été antérieurement le cas des autres symptômes dus aux autres paralysies de nerfs crâniens, ont totalement régressé, probablement grâce à l’amélioration du contrôle métabolique.

Mots-clés : paralysie des nerfs crâniens, paralysie de la corde vocale, diabète de type 1.

This study was undertaken at Akdeniz University School of Medicine, Department of Pediatric Endocrinology, Antalya, Turkey.

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Neuropathy is one of the chronic complications of diabetes mellitus. Besides hyperglycemia and related metabolic changes, structural and functional disorders due to vascular and hypoxic causes in various parts of the nervous system are the mechanisms responsible for diabetic neuropathy [1]. Not only patients with peripheral nerve involvement, but also patients with isolated upper cranial nerve paralysis in diabetes are frequently reported [2-6]. The majority of cranial neuropathies affect the 3rd, 6th and 7th cranial nerves while the 4th cranial nerve is rarely affected alone [2-6]. Reports related to inferior laryngeal nerve paralysis are extremely rare [7-9]. There is no report in the literature related to the hypoglossal nerve paralysis in diabetic patients. In this article, the case of a diabetic patient with temporary hypoglossal, facial and recurrent nerve palsies which improved completely, presumably due to good metabolic control, is presented.

CASE REPORT

A 7-year-old girl with type I diabetes mellitus, who has been followed up for 6 years, was admitted to the hospital with complaints of facial numbness and difficulty in speaking. Right lower facial weakness, straightened nasolabial sulcus, inability to show her teeth on the right side, and rightward deviation of her tongue were detected upon neurological examination. Other neurological examinations were completely normal. Computed tomography (CT) of medulla oblongata and pons were normal. She was receiving conventional insulin therapy (CIT) (0.8 U/kg/day, twice daily). Mean HbA1c level for the previous year was 11.3%. To improve the metabolic control, we administered intensive insulin treatment (IIT) for 6 months. Follow-up HbA1c levels were 9.3 and 8.4%. She was not in compliance with IIT and refused to use this regimen. CIT (1 U/kg/day, twice daily) was restarted. Facial and hypoglossal nerve palsies recovered completely after 3 months.

After 2 years, she was readmitted to the hospital complaining of hoarseness lasting for 2 weeks. Laryngostroboscopic examination revealed right inferior laryngeal nerve paralysis (Fig. 1). Probable disorders of thorax and central nervous system that might cause vocal cord palsy were eliminated by normal thoracic and cranial CT. She had been receiving CIT (1 U/kg/day, twice daily). Mean HbA1c of the patient for the last year was 10%. Electroneuromyography of the peripheral nerves was normal. Following her glucose regulation, she was advised to come to visits regularly for check-ups and discharged from the hospital. Follow-up HbA1c levels were 8% and 7.4%.

Hoarseness of the patient disappeared completely after 6 months. Control laryngostroboscopic examination revealed normal vocal cord movements (Fig. 2).

FIG. 1. Incomplete glottal opening due to right inferior laryngeal nerve paralysis. There is vibration disorder because of the loss of muscular mass and firmness beneath the mucosa.

FIG. 2. Following the improvement of right inferior laryngeal nerve paralysis; complete glottal opening, and normal vibration of vocal cords as a result of normalization of muscular mass and firmness beneath the mucosa.
DISCUSSION

Although clinical diabetic neuropathy is rare in children, decreases in motor conduction rate and sensorial changes have been reported [10]. One of the largest published series reported a prevalence of diabetic neuropathy of 7.5% at the time of diagnosis of diabetes, with the prevalence increasing steadily thereafter with no apparent plateau [11]. Metabolic effects of hyperglycemia and ischemia in nervous tissue are thought to be the mechanisms responsible for diabetic neuropathy [1]. Factors implicated in the pathogenesis of diabetic neuropathy include the activation of polyol pathway, the activation of protein kinase C, increased oxidative stress, the impaired N-6 fatty acid metabolism, auto-oxidation of glucose, the formation of advanced glycation end products, and the reduced bioavailability of neurotrophic factors. Although the exact mechanisms of their action are not well understood, it is currently believed that these factors lead to reduced Na⁺, K⁺ ATPase activity and vasoconstriction, reduced endoneurial blood flow and nerve hypoxia. The latter changes then lead to reduced nerve conduction velocities, axonal loss, axonal demyelination, and nerve dysfunction. Other physiological changes that accompany the onset of diabetes may also contribute to peripheral neuropathy. In particular, decreased blood flow to these nerves is one of the earliest functional findings in the development or induction of diabetes. The resulting local hypoxia in the peripheral nerves is believed to be a major pathogenic factor, although impaired mitochondrial functions and apoptosis of neurons and Schwann cells also occurs with similar timing and may act independently of hypoxia to induce peripheral nerve dysfunction. In addition, reduction in neurotrophic factors responsible for the structural nerve axon integrity have also been implicated in the pathogenesis of diabetic neuropathy [12].

Diabetic neuropathy is seen in the form of symmetric polyneuropathies or focal neuropathies [5]. Mononeuropathies characterized by involvement of motor and sensorial fibers of a nerve appear more often as muscular weakness and usually improve in a short period of time [13]. Cranial neuropathies also belong to the category of mononeuropathies [5]. Watanabe detected 19 (0.97%) cranial nerve paralysis in 1961 diabetic patients, involving the 3rd, 6th and 7th cranial nerves and reported that occurrence of cranial nerve paralysis in diabetic patients was higher than in non-diabetic patients [6]. Facial paralysis due to 7th nerve palsy may occur with increased frequency in patients with diabetes mellitus, and the prognosis for recovery may be worse than that in patients without diabetes mellitus [5, 6].

The probable effect of hyperglycemia on neuropathy development and an increase in motor conduction rate following good metabolic control have been reported [14-16]. Because of the absence of any other cause except diabetes mellitus and of high HbA1c levels on each admission, the multiple temporary cranial nerve palsies of our patient were thought to be due to poor metabolic control of diabetes mellitus. An acute neuropathy rarely occurs early in the course of diabetes mellitus. However, our patient had a poorly controlled type 1 diabetes mellitus and diabetic ketoacidosis has occurred four times since the diagnosis of the disease. Uncommon early-onset neuropathy in diabetic patients has been reported in the literature [17]. It has been suggested that acute disequilibrium in the diabetic status may facilitate the occurrence of a variety of neuropathies. Alternatively, the autoimmune process which led to type 1 diabetes mellitus may also trigger an autoimmune neuropathy with vasculitis or demyelinating nerve lesions [17].

The Diabetes Control and Complications Trial (DCCT) demonstrated that IIT delayed or prevented electrophysiological abnormalities associated with diabetic neuropathy [18]. The first step in the treatment of diabetic neuropathy is the regulation of blood glucose [5]. In a limited number of cases with vocal cord palsies in the literature, partial or complete recovery within several months has been reported [8, 9]. In our case also, following the improvement of metabolic control, complete remission of clinical findings due to 7th and 9th cranial nerve palsies occurred in 3 months, while those due to right inferior laryngeal nerve paralysis remitted in 6 months.

In conclusion, the hoarseness and physical signs of vocal cord palsy of our patient, as on her first admission with other symptoms and signs due to other cranial nerve palsies, including the 9th cranial nerve involvement, which is probably unique in the literature, improved completely, presumably owing to good metabolic control.

REFERENCES

16 Young RJ, Ewing DJ, Clarke BS. Nerve function and metabolic control in teenage diabetics. Diabetes, 1988, 32; 142-147.