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Benign position paroxysmal vertigo with congenital nystagmus: A case report

Li GF *et al*. BPPV with congenital

Abstract

BACKGROUND

Benign position paroxysmal vertigo (BPPV) is a temporary vertigo induced by moving the head to a specific position. It is a self-limited, peripheral, vestibular disease and can be divided into primary and secondary. Congenital nystagmus (CN), an involuntary, rhythmic, binocular-symmetry, conjugated eye movement, is found at birth or within three months of birth. According to the pathogenesis, CN can be divided into sensory-defect nystagmus and motor-defect nystagmus. The coexistence of BPPV and CN is rarely seen in clinic.

CASE SUMMARY

A 62-year-old woman presented to our clinic complaining of a 15-d recurrent positional vertigo. The vertigo lasting less than one minute occurred when she turned over, sometimes accompanied by nausea and vomiting. Both the patient and her father had CN. Her spontaneous nystagmus was horizontal to right; however, the gaze test revealed variable horizontal nystagmus with the same degree when the eyes moved. The patient's Dix-Hallpike test was normal, except for persistent nystagmus, and the roll test showed severe variable horizontal nystagmus, which lasted for about 20 s in the same direction as her head movement to the right and left, although the right-side nystagmus was stronger than the left-side. Since these symptoms were accompanied by nausea, she was diagnosed with BPPV with CN and treated with manual reduction.

CONCLUSION

Though rare, if BPPV with CN is correctly identified and diagnosed, reduction treatment is comparably effective to other vertigo types.

Key Words: Congenital; Nystagmus; Benign position paroxysmal vertigo; Case report

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Core Tip: Benign position paroxysmal vertigo (BPPV) is defined as a disorder of the inner ear characterized by repeated episodes of positional vertigo. Congenital nystagmus (CN), an involuntary, rhythmic, binocular-symmetry, conjugated eye movement, is found at birth or within three months of birth. BPPV with CN is rarely seen in clinics. First, CN should be distinguished from other pathologic nystagmus types. Then, BPPV can be accurately determined through postural nystagmus. We report the characteristics of BPPV with CN, further explaining how to identify nystagmus.

INTRODUCTION

Benign position paroxysmal vertigo (BPPV) is defined as a disorder of the inner ear characterized by repeated episodes of positional vertigo[1]. BPPV is among the common diseases that cause aural vertigo, and 24.1% of patients with dizziness or vertigo are diagnosed with BPPV[2]. Spontaneous nystagmus refers to a continuous, involuntary, and rhythmic movement of the eyeball in the absence of inducing factors and is divided into congenital nystagmus (CN) and acquired nystagmus. The latter is a type of nystagmus commonly seen in clinic. CN is ocular motor disorders in which patients are afflicted by periodic involuntary ocular oscillations affecting both eyes[3,4]. develops during the first three to six months of a patient's life and has a prevalence of 14 per 10000 people in the United Kingdom[5]. The etiology of CN is largely unknown, but we know that most patients have lifelong nystagmus, although it can gradually decrease with age in some patients. Some BPPV patients present with spontaneous nystagmus, but BPPV with CN is rare: To date, we have not seen such cases reported. In this report, we present the case of a BPPV patient with CN and her nystagmus findings.

CASE PRESENTATION

Chief complaints

A 62-year-old woman presented at our clinic, complaining of positional vertigo that had recurred for 15 d.

History of present illness

Fifteen days previously, the patient's symptoms had begun with severe dizziness when she rose, which recurred when she rolled over or lied down. She sometimes experienced nausea and vomiting at the onset of these symptoms.

History of past illness

Physical health.

Personal and family history

The patient and her father had histories of CN.

Physical examination

The patient's physical examination revealed no abnormal findings.

Laboratory examinations

She didn't have laboratory examinations.

Imaging examinations

She didn't have imaging examinations.

FINAL DIAGNOSIS

The patient was ultimately diagnosed with BPPV with CN.

TREATMENT

The patient was prescribed a barbecue roll maneuver to treat her right, lateral, semicircular canal BPPV of the geotropic type. This treatment requires a patient to lie

down with the affected ear facing downward. Then, they roll over to the opposite side for 90 degrees until he returns to the original position. Our patient's vertigo symptoms disappeared after the therapy.

OUTCOME AND FOLLOW-UP

The patient's follow-up treatment comprised three telephone appointments, at one week, one month, and six months after her treatment. The patient was asymptomatic, without any recurrence of vertigo.

DISCUSSION

BPPV is generally categorized as posterior semicircular canal BPPV, anterior semicircular canal BPPV, or horizontal semicircular canal BPPV. Of these categories, posterior semicircular canal BPPV is the most common (affecting 80%-90% of patients), followed by horizontal semicircular canal BPPV (10%-20%), while anterior semicircular canal BPPV is rare (3%)[6,7]. The Dix-Hallpike maneuver is considered the gold standard test to diagnose posterior canal BPPV, and the supine roll test is considered the gold standard for diagnosing horizontal semicircular canal BPPV[8]. Upbeat-torsional nystagmus is provoked by vertical semicircular canal BPPV.

BPPV's pathogenesis remains unclear; however, risk factors include age, mental stress, osteoporosis, insomnia, and hypertension[9,10]. Currently, the following two theories are accepted. First, canalithiasis suggests that when the head is moved relative to gravity, otoliths residing on the macula utriculi migrate into the semicircular canal and are displaced relative to the semicircular canal wall because of gravity, causing endolymph flow and resulting in the deviation of cupula terminalis and, in turn, corresponding signs and symptoms. When the otolith moves due to gravity to the lowest point in the semicircular canal lumen, the endolymph stops, the cupula terminalis returns to its original position, and signs and symptoms disappear.

Second, eupulolithiasis suggests that the detached otoliths on the macula utriculi adhere to the cupula terminalis, changing the density of the latter relative to the

endolymph and making it sensitive to gravity, resulting in the corresponding symptoms and signs[11]. CN usually occurs at birth or within three months of birth. Although this nystagmus persists throughout most patients' lives, some patients' symptoms gradually decrease with age. CN is divided into two categories. The first is motional defect congenital motor nystagmus, in which eye movement includes fast and slow phases. The second is congenital sensory defect nystagmus, also known as "pendular nystagmus", in which the eye moves at one speed. CN is clinically rare, with an incidence of about 0.005%-0.286%[3]. To our knowledge, BPPV with CN has not been previously reported.

In the United States, according to statistics, about 5.6 million patients clinically complain of dizziness per year, and 17%-42% of patients with vertigo are diagnosed with BPPV[12-14]. BPPV treatment can be categorized as canalith repositioning maneuvers or vestibular rehabilitation[1]. Diagnosis of horizontal semicircular canal BPPV relies on a supine roll test. During examinations, clinicians should observe whether the direction of the nystagmus is geotropic or apogeotropic and which side of the nystagmus is stronger to enable identification of the patient's affected side. In our case, a Dix-Hallpike test of the patient showed the signs of horizontal nystagmus without vertigo and geotropic nystagmus, which was stronger on her right side during a roll test. Unlike other patients with BPPV, she exhibited persistent horizontal nystagmus on the right side after intense nystagmus lasting more than ten seconds and accompanied by vertigo. The lasting nystagmus was CN, which is similar to patients with spontaneous nystagmus but absent in BPPV patients without spontaneous nystagmus. Spontaneous nystagmus is very common clinically among patients with vertigo.

The following aspects should be used to distinguish CN from other central and peripheral spontaneous forms of nystagmus. The first and most important aspect is a patient's medical history. Nystagmus is always present during CN; someone in the patient's family will have CN because of the heritability of the disease, while other types of spontaneous nystagmus only appear at the onset of the disease. Second, the

direction of peripheral nystagmus is constant, while that of CN is variable. The test can be conducted with Frenzel glasses to observe the nystagmus accurately. The direction of the nystagmus will be seen to remain the same in the peripheral nystagmus; however, the direction of the nystagmus is consistent with the eye movement in central nystagmus and variable nyatagmus in CN. Third, a CN patient usually experiences horizontal nystagmus of variable intensity, while other pathologic central nystagmus types may entail vertical and horizontal nystagmus with generally persistent intensity.

CONCLUSION

CN is rare in clinic. If individuals experience spontaneous nystagmus with constant intensity and variable direction, a careful medical history should be taken to eliminate CN, which may influence the diagnosis. The treatment for BPPV with CN is the same as for BPPV. In the case reported here, the patient was diagnosed with BPPV with CN, and the result was good.

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