Downbeating Nystagmus and Postural Hypotension due to Basilar Invagination

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Abstract

Downbeating nystagmus is an involuntary vertical rhythmic eye movement with the fast component in the downward direction. The sign indicates a craniocervical disorder. The most common cause is the Arnold-Chiari malformation, followed by cerebellar degeneration. Basilar invagination is a rare cause of downbeating nystagmus. However, with appropriate treatment its prognosis is good. Here, we report a case of basilar invagination which presented with downbeating nystagmus and postural hypotension.

A 31 year-old Thai male patient had a 20 year history of postural hypotension. He had recurrent pneumonia and cough-induced syncope a year before admission. He complained of symptoms of an acute febrile illness and a productive cough. The physical examination showed high grade fever, postural hypotension and medium crepitation in the right upper lobe. The neurological examination showed downbeating nystagmus, atrophy and fasciculation of the right side of the tongue, atrophy of the right sternocleidomastoid muscle, mild weakness of the extremities and generalized hyperreflexia. The cervical spine X-ray revealed upward displacement of the vertebral bodies of C1 and C2, with a mild narrowing of the space between C1 and the occiput. The CT-myelogram and MRI showed upward displacement of C1 with overriding of the dens over the anterior lip of the foramen magnum ; this also compressed the medulla. Syringomyelia was seen at the C1-C5 level.

We report a patient who presented with postural hypotension, recurrent pneumonia and downbeating nystagmus due to basilar invagination. The symptoms were aggravated by cough which caused an increase in intracranial pressure. This resulted from medulla compression in the foramen magnum by the first cervical spine. The treatment of choice was surgical decompression.

Key word : Downbeating Nystagmus, Postural Hypotension, Basilar Invagination

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Downbeating nystagmus (DBN) is an involuntary rhythmic eye movement in the vertical direction from the primary position. Its fast component is seen when the patient looks down and it is accentuated on lateral gaze[1]. It is a specific neurological sign that indicates a craniocervical junction disorder[1]. About a third of the patients with this sign have the Arnold-Chiari malformation. Other causes are cerebellar degeneration, drug-induced DBN and basilar invagination. Basilar invagination is an uncommon cause but it has a good outcome if corrected surgically. We report a case of DBN and postural hypotension due to basilar invagination.

CASE REPORT

A 31 year-old married Thai male farmer complained of symptoms of a high grade fever and a productive cough. He had had the symptoms for seven days. In addition, when he sat up, he had generalized spasticity, blurred vision and syncope, all of which were made worse by coughing. The symptoms disappeared when he lay down, and were less severe when he slowly sat up or stood. Postural syncope occured four times a day and lead him to see a doctor.

Retrospectively, he thought that he had had postural symptoms for over 20 years, especially when changing from the sitting to the standing position. If he slowly stood up, this symptoms were less severe and he could carry out his normal activities. He had had no ataxia, vertigo or oscillopsia. Ten years previously, he discontinued driving because of dizziness. For four years he had slight dysphagia. One year previously he developed three episodes of pneumonia with frequent cough-induced bouts of postural syncope.

Physical examination showed a temperature of 39°C, respiratory rate of 24/min, supine blood pressure of 110/60 mmHg and standing pressure of 70/40 mmHg. The cardiovascular examination was normal, but the lung had medium crepitations in his right upper lobe and generalized rhonchi in both lungs. He was fully conscious. He had downbeating nystagmus in both eyes, atrophy and fasciculation in the right side of his tongue and atrophy of the right sternocleidomastoid muscle. His uvula was central and he had a normal gag reflex. He had mild weakness of the extremities, a positive Hoffman's sign in both hands and generalized hyperreflexia.

Laboratory investigations showed leukocytosis with a shift to the left and normal blood chemistry. Sputum culture yielded gram positive diplococci and a chest X-ray demonstrated alveolar infiltration with air bronchogram in the right upper lobe. Streptococcal pneumonia was diagnosed and successfully treated with penicillin. Because of the DBN, a pathological lesion at the craniocervical junction was suspected. A plain X-ray of the cervical spine showed upward displacement of the dens of C2 and anterior arch of C1, with mild narrowing of the distance between C1 and the occiput (Fig. 1). A CT-myelogram demonstrated upward displacement

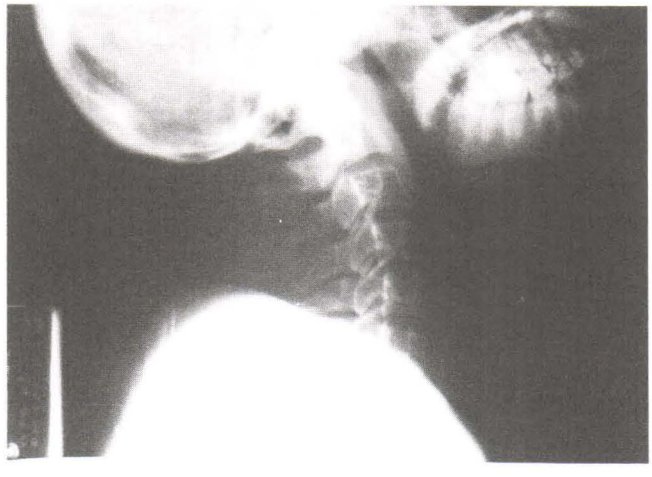

Fig. 1. Plain film of cervical spine shows upward displacement of dens of C2 and anterior arch of C1 with mild narrowing of distance between C1 and occiput.

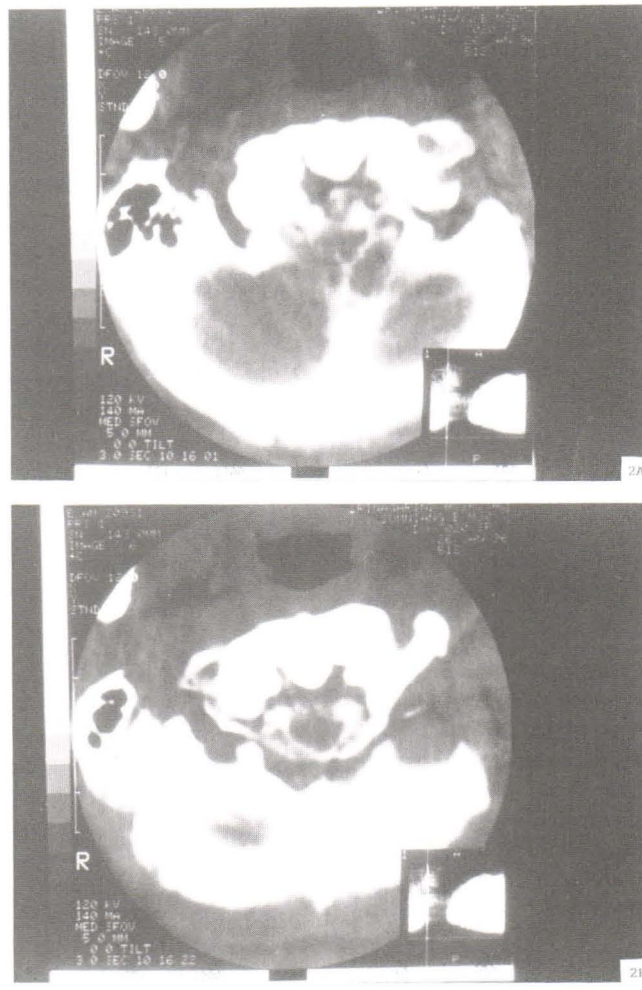

Fig. 2 A, B. CT-myelogram show upward displacement of C1 with overriding at anterior lip of foramen magnum over dens and compression on medulla.

of C1, with the dens overriding the anterior lip of the foramen magnum and compression of the medulla (Fig. 2 A, B). An MRI disclosed the same findings as the CT-myelogram, together with posterior displacement and angulation of the cervicomedullary junction of the spinal cord and brain stem and syringomyelia of the spinal cord (C1-C5). There was no evidence of tonsillar herniation. The diagnosis was "basilar invagination". The patient had foramen magnum decompression and, as a result, no further postural hypotension.

DISCUSSION

Downbeating nystagmus (DBN) indicates pathology at the craniocervical junction[1]. The more common causes are the Arnold-Chiari malformation, cerebellar degeneration, a posterior fossa tumor and basilar invagination. Less commonly DBN is caused[2-18] by drugs (lithium, carbamazepine, felbamate and amiodarone), severe magnesium depletion, excessive alcohol intake, vitamin B12 deficiency, demyelinating disease, acute cerebellitis, brain stem infarction and vertebral artery compression.

Basilar invagination is a developmental defect of the chondrocranium resulting in an elevation of the foramen magnum's rim. It is frequently associated with the Arnold-Chiari malformation, occipitalization of the atlas, syringomyelia and syringobulbia. It may give rise to a short neck and

low hair line. A patient may also have syringomyelia without evidence of the Arnold-Chiari malformation.

Da Silva et al reported 230 cases of basilar impression and the Arnold-Chiari malformation. He found concurrent autonomic nervous system disorders in 59.1 per cent of the cases. The most common symptom was sexual disorder (60.9%). The other symptoms resulted from urinary dysfunction, anhydrosis and hyperhydrosis[19]. Postural hypotension was not reported.

Our patient had underlying basilar invagination which produced a 20 year history of postural hypotension as a result of medulla compression. He also had gradual impairment of function of the ninth and tenth cranial nerves. He developed dysphagia and recurrent pneumonia from microaspiration. Repetitive cough produced an increase in intracranial pressure which resulted in descending of posterior fossa brain tissue through the foramen magnum[13]. This caused compression or parenchymal shear stress during movment through the foramen magnum, resulting in a pathological effect on the nuclei prepositus hypoglossi, prominent DBN and postural syncope.

Craniocervical anomaly surgery resulted in clinical improvement in about 72 per cent cases reported by Chopra et al[8]. Halmagyi et al[3] reported three cases of the Arnold- Chiari malformation with DBN treated surgically. Follow-up revealed that one patient had ataxia but no DBN four years after the operation; the second had unchanged neurological signs (quadriparesis, mild spastic ataxia and DBN) two years later; and the third was free of postural vertigo but had unchanged DBN two years after the operation.

We report a case of basilar invagination which presented with the unusual symptom of postural hypotension and prominent downbeating nystagmus accentuated by coughing. The craniocervical anomaly had a good outcome following surgical decompression.

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ภาวะตากระตุก Downbeating และภาวะความดันโลหิตต่ำจากการเปลี่ยนท่าเนื่องจาก Basilar invagination

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Downbeating nystagmus คือการเคลื่อนไหวที่ผิดปกติของตาโดยมีทิศทางของ quick phase ลงล่าง เป็นอาการแสดงที่บอกว่ามีความผิดปกติของสมองบริเวณรอยต่อระหว่างกระดูกกะโหลกศีรษะและกระดูกคอส่วนบน สาเหตุที่พบบ่อยคือความผิดปกติชนิด Arnold-Chiari และการเสื่อมของสมองส่วน cerebellum สาเหตุที่พบน้อยคือ basilar invagination แต่เป็นภาวะที่ได้ผลดีเมื่อรับการรักษาที่ถูกต้อง จึงขอรายงานผู้ป่วย basilar invagination ที่มาพบด้วย downbeating nystagmus และ postural hypotension

ผู้ป่วยชายอายุ 31 ปี มีประวัติ postural hypotension นาน 20 ปี 1 ปีก่อนเคยเป็นปอดอักเสบจากการติดเชื้อและเป็นลมเมื่อมีอาการไอ ผู้ป่วยมีอาการไข้ ไอมีเสมหะ ตรวจร่างกายพบไข้สูง postural hypotension เสียง crepitation ที่ปอดด้านขวา ตรวจร่างกายระบบประสาทพบ downbeating nystagmus กล้ามเนื้อลิ้นลีบและมี fasciculation ด้านขวา กล้ามเนื้อ sternocleidomastoid ลีบอ่อนแรงและรีเฟล็กซ์ไวทั่วๆไป เอกซ์เรย์กระดูกคอพบการเลื่อนขึ้นบนของกระดูกคอระดับที่ 1 และ 2 ระยะห่างระหว่างกระดูกคอระดับที่ 1 และ occiput แคบลง เอกซ์เรย์คอมพิวเตอร์ร่วมกับการฉีดสารทึบแสงและการตรวจคลื่นแม่เหล็ก พบการเลื่อนขึ้นของกระดูกคอระดับที่ 1 อยู่ที่ระดับของ foramen magnum และกดต่อสมองส่วน medulla นอกจากนี้พบ syringomyelia ที่ระดับกระดูกคอที่ 1 ถึง 5

ได้นำเสนอรายงานผู้ป่วยที่มีอาการ postural hypotension ปอดอักเสบจากการติดเชื้อและ downbeating nystagmus โดยมีสาเหตุจาก basilar invagination ซึ่งอาการเป็นมากขึ้นเมื่อไอ ซึ่งมีผลให้มีการเพิ่มขึ้นของความดันในกะโหลกศีรษะและมีผลให้มีการกดทับของสมองส่วน medulla มากขึ้น การรักษาคือการผ่าตัดแก้ไขความผิดปกติดังกล่าว

คำสำคัญ : ภาวะตากระตุก Downbeating, ภาวะความดันโลหิตต่ำจากการเปลี่ยนท่า, Basilar Invagination

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