



CASE REPORT OF CONGENITAL CERVICAL BASILAR INVAGINATION

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<p>Article Info Received 15/08/2015 Revised 27/08/2015 Accepted 05/09/2015</p> <p>Key words: basilar invagination.</p>	<p>ABSTRACT 12 years old female presented with headache , neck pain , neck stiffness & progressive Rt sided weakness for one month with signs of U.M.N lesions bilaterally with intact cranial nerves .diagnosis of congenital basilar invagination was done based on radiological finding.</p>
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INTRODUCTION

A 12 Years old from Abu-guta-Gazera state-Sudan. She was admitted at 26 January 2009 to Omdurman pediatrics teaching hospital, complains of headache and right sided weakness for one month. The condition started one month prior to admission with continuous, generalized, dull headache ,no relieving or aggravating factors, associated with vomiting once, which is small in amount, at morning not projectile. One week later she developed sever localized neck pain which is aggravated with flexion of the neck with no relieving factor.

After 5 days she developed neck stiffness then she became unable to move her right leg from side to side or to walk. Seven days later they noticed that she was unable to hold things with her right hand. Two days later she became unable to moves her right upper limb. She had no fever, blurring of vision, syncopal attacks, convulsion, aura, or trauma. No history of numbness sensation, parathesia or back pain. No symptoms related to cranial nerves involvement and no sphenteric disturbances.

Cardiopulmonary

She did not complain of cough, shortness of breath, palpitation, or chest pain

Gastrointestinal system

She had good appetite, No nausea or loss of weight, and normal bowel habits.

Genitourinary system

She had normal urine amount, color, and frequency.

Musculoskeletal system

She had No joint pain, swelling, skin rash, hypo- or hyper pigmented areas.

Past medical history (PH)

She had No PH of hospitalization. No PH of similar condition. No PH of trauma.No PH of hands and feet swelling, jaundice or blood transfusion. No history of ear discharge, facial trauma or infection.No history of contact with chronic cough.

She pass through normal developmental milestone. She is not in school (only khalwa* for 2yrs with good performance). She was Fully vaccinated according to old EPI program with BCG scar. She was on satisfactory balanced Sudanese diet.



Family History (FH)

No FH of similar condition. No FH of stroke or of sudden death. No FH of sickle cell anemia. No FH of migraine. They lives in their own house compose of 3 rooms and kitchen with no electrical supply, there is tap water. No animals in their house. Their Family is supported by her elder brothers. They were of poor socioeconomic status. The disease has negative impact on the family socially and financially because they left their home and came to Omdurman to treat their daughter. She is not known allergic to any medication and not on long term medications.

On examination

She looks unwell, not pale jaundiced or cyanosed. Has no dysmorphic features

Pulse Rate: 110 beat/min

Respiratory Rate: 20cycle/min

Temperature (axillary): 37.4C

Blood pressure: 90/60 (50th centille)

Weight was 30.5 kg at 3rd centile.

Height was 141cm at 10th centile.

Head circumference was 52 cm at 50th centile.

Normal eyes appearance no exophthalmia, squint or nystagmus. Normal fundus. There is neck stiffness on flexion no tenderness, no lymph node or thyroid enlargement& no bruit. Normal hair line.

Cardiovascular examination

Pericardium examination: normal appearance, no visible pulsations. Apex is at 5th intercostals space, midclavicular line, of normal character. No thrill or left parasternal heave. Second heart sound not palpable. First and second heart sound are normal, with no added sound, nor murmur. Normal chest, abdominal and musculoskeletal examination. Here Sexual maturity rate was at tanner stage 2.

Neurological examination

She was fully conscious. Well oriented for time and place. Normal memory and mood. She was cooperative and of average intelligence. Cranial nerves were intact. Fundoscopy was normal. Sustained ankle clonus was found bilaterally. Abdominal reflexes were present. Planter reflex was up going bilaterally. Sensation was intact. She had normal back examination with difficult to gait assessment.

Investigations

Urine and stool investigations were normal.

Complete Blood Count

HB: 12.3 g/dl

PCV: 39 %

MCH: 25pg

MCV: 86 fl

MCHC: 35 g/dl

Retics count: 1.2%

TWBC: 7700, POLY: 60%, LYMPH: 36%, MONO:3%

Platelet: 236000

ESR: 20 mm/hr

Comments of here complete blood count

RBCS: normocytic normochromic

WBCS: Normal morphology

Hb electrophoresis: A A.

Blood urea: 2.7 mmol/l (N 2.5 – 6.4)

Creatinine : 80 µmol/l (N<85 µmol/l)

Na : 135 mmol/l (N 135 – 146)

K : 3.5 mmol/l (N 3.5 – 5)

Ca : 2.3 mmol/l (N 2.1 – 2.5)

Rheumatoid factor: was negative

ANA profile: was -ve

Bleeding profile:- PT is 14.6 sec(13-18)sec

PTT is 29.3sec(23-33)sec

INR is 1.1

Chest x ray and cervical x ray were normal, abdominal ultrasound was normal. (Figure1 & 2.)

Cervical CT scan and MRI Reports:

Subluxation of the odontoid process with projection into the posterior cranial fossa, compression of the medulla oblongata and effacement of the lower part of the 4th ventricle. The presence of condylus tertius(an accessory ossicle posterior to the clivus) that creates pseudo-joint with the outer arch of atlas leads to diagnosis of congenital basilar invagination. In terms of anatomy:-the calivus is more horizontal than usual, there is an accessory bone seen in continuity with the posterior aspect of the clivus, the former is partially fused with the anterior arch of atlas (false joint). The angle or clivus canal angle is found to be <90degree where it should not be <150degree. (Figure 3, 4, 5, 6 & 7).

CONCLUSION OF THE REPORT

Congenital type of basilar invagination complicated with odontoid subluxation. The cause of invagination is condylus tertius

Treatment

Diagnosis of basilar invagination was done based on radiological findings, neurosurgery department at ALSHAAB hospital-Khartoum were consulted and patient was but on cervical collar with restriction of neck movement till the surgery but parents refuse the operation and accepted only the conservative treatment.

DISCUSSION

Basilar Invagination and Basilar impression (BI)

Basilar invagination and Basilar impression are uncommon syndrome that occurs when the superior part of the odontoid (part of the C2 vertebrae) migrates upward[1]. For the most part, the terms basilar invagination and basilar impression are often used interchangeably because in both cases there is upwards migration of the upper cervical



spine, but precisely, basilar invagination is defined as upward displacement of vertebral elements into the normal foramen magnum with normal bone, while basilar impression is a similar displacement due to softening of bones at the base of the skull [8]. Thus different terms are used according to whether bone is normal or not.

Platybasia, is a related term, indicating an increase in the basal angle of the skull -- basically a flattening of the base of the skull. Platybasia is diagnosed when, on a lateral skull film or sagittally formatted CT, the angle of the line drawn along the plane of the sphenoid sinus and along the clivus is greater than 145 degrees. Platybasia, per se, is not a disease. [2]

BI is uncommon but somewhat dangerous. It occurs both congenitally (i.e. basilar invagination associated with Down's syndrome, Klippel Feil syndrome, Chiari malformation (25-35% of BI),) and in persons with bone diseases (basilar impression), such as rheumatoid arthritis (also called cranial settling), hyperparathyroidism, Paget's disease, Osteogenesis imperfecta (also associated with hearing loss from otosclerosis), Rickets, Hurler's syndrome, and Hadju-Cheney syndrome. It may lead to static or dynamic stenosis of the foramen magnum, and compression of the medulla oblongata (lower brainstem). It can manifest as sudden death due to fatal brainstem compression, but more often it is asymptomatic. [3]

Basilar invagination from rheumatoid arthritis is due to loss of axial supporting structures in the upper cervical spine. It is estimated that about 10% of patients with rheumatoid arthritis are at risk for sudden death, but in reality, this seems to happen very rarely. Obstructive hydrocephalus or syringomyelia may also be seen because of direct mechanical blockage of normal CSF flow.

Diagnosis of Basilar Impression

Symptoms of BI generally become apparent when there is a great deal of flexion. It can present as posterior skull pain. According to Sawin and Menezes [7] series in persons with osteogenesis imperfecta , symptoms and signs included headache (76%), lower cranial nerve dysfunction (68%), hyperreflexia (56%), quadriparesis (48%), ataxia

(32%), nystagmus (28%), and scoliosis (20%). Downbeating nystagmus and postural hypotension has been reported [5]. A plain lateral x-ray, with odontoid views, is a good place to start, but is not 100% sensitive[6]. Chamberlain's line is drawn between the posterior hard palate to the posterior edge of the foramen magnum. If the dens is more than 3 mm above this line, the patient has basilar invagination. We sometimes find it hard to find the odontoid on lateral skull films, and have much less problems on sagittal MRI's. CT scans are also much easier to read.

McGregor's line is the line drawn from the posterior hard palate to the base of the occiput (lowest point). If the dens are more than 4.5 mm above this line, again basilar invagination is diagnosed. There isn't much difference between these two lines -- but McGregor's is easier to use as there is no judgment needed to find the posterior lip of the foramen magnum. McRae's line is the opening of the foramen magnum.

Treatment of Basilar Impression

Neurosurgery is recommended when neurologic symptoms and signs are present, and cord compression is confirmed by MRI. When these features are absent, a conservative approach may be pursued, such as a collar, nonsteroidal anti-inflammatory drugs (NSAIDs), and simple neck traction. In patients who are considered poor surgical risks, neurologic progression is likely and the one-year prognosis is poor.

Neurosurgical treatment is quite difficult. In the series of Sawin and Menezes [7], they suggest that ventral brainstem compression should be treated with ventral decompression, followed by occipitocervical fusion with contoured loop instrumentation to prevent further squamooccipital infolding. In persons with Chiari malformation, there may also be occipital decompression surgery. Ventral decompression means operating through the front of the head, through the mouth. Both the transoral and posterior surgery can be done in one sitting [9]. Another approach is atlantoaxial joint distraction and direct lateral mass fixation using screws [4].

Figure 1. Chest-x-ray

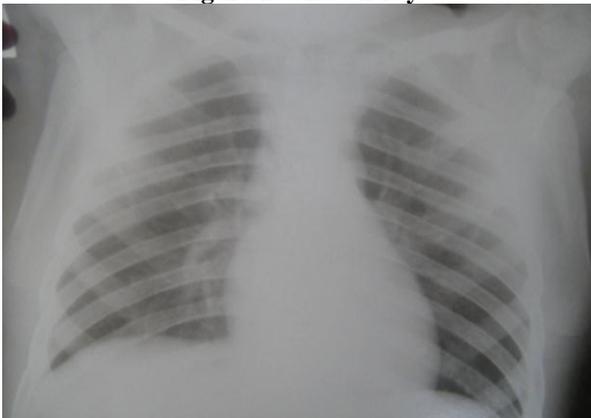


Figure 2. Cervical-x-ray



Figure 3. CT-Brain

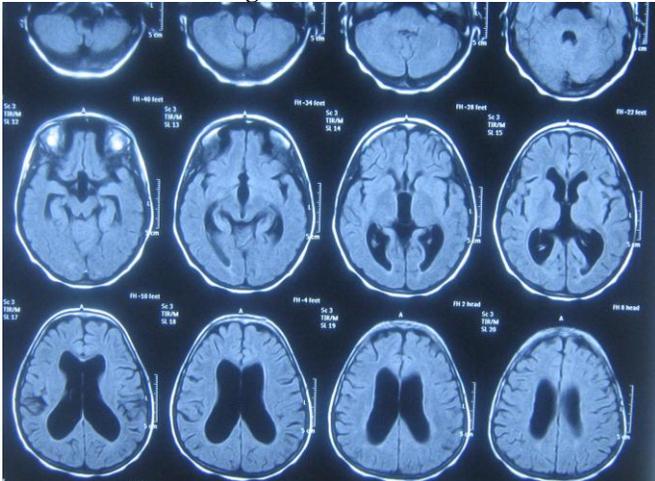


Figure 4. MRI-Neck & foramen magnum

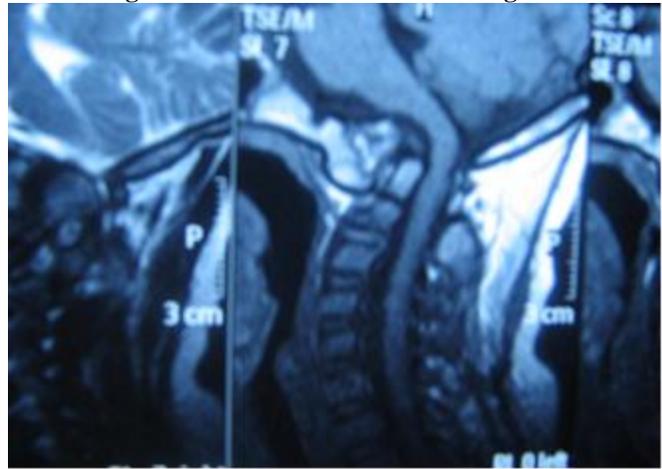


Figure 5. CT contrast Neck & foramen magnum

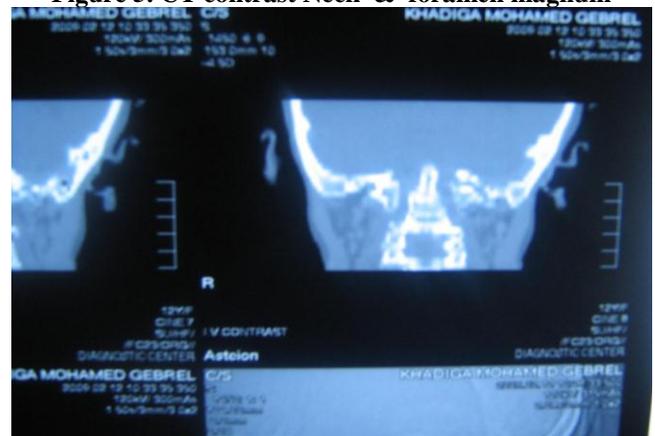


Figure 6. MRI neck & foramen magnum

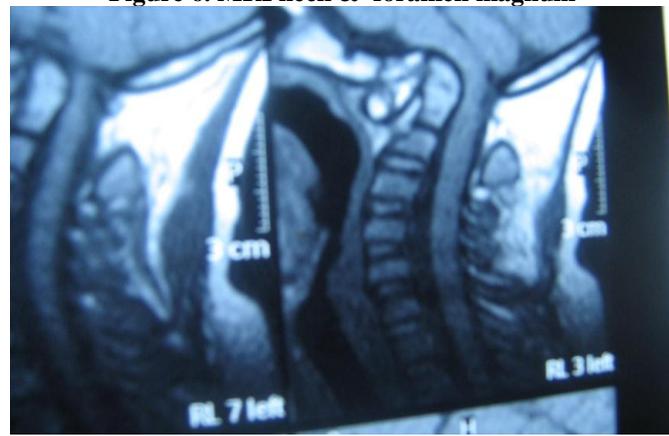
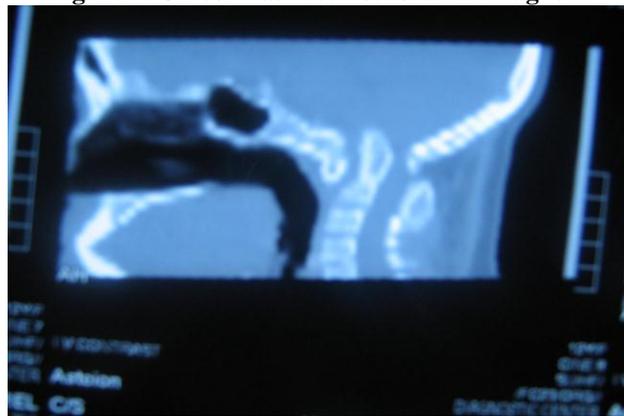


Figure 7. CT contrast neck & foramen magnum



CONCLUSION

Cervical abnormalities are not an uncommon problems specially in those with congenital anomalies in the neck. Radiological and clinical evaluation is needed in those with early symptoms.

ACKNOWLEDGEMENT

We would like to thanks our colleague Dr.Hanaa Faisal, Dr.Allaa Omer Alsamani Dr.Sumia Mohammed Ali, and Dr.Aarafat Ahmed Mohammed to their great effort with us during the investigations of our patient.

CONFLICT OF INTEREST: NIL



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