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Basilar Invagination: A Case Review

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Basilar invagination is a rare developmental defect involving the basiocciput. Often presents with severe neck pains, short webbed neck and difficulty in rotating the neck. We present a case of a 14 year old who presented with progressive neck weakness associated with pain and torticollis. Radiological investigations reviewed

Key words: Basilar, invagination, basiocciput.

Introduction

Basilar invagination is a developmental defect of chondrocranium thus involvement of basiocciput, exoccipital bone as well as squamou-occipital bone[1]. Consequently it results in cephalad migration of vertebrae into cranial cavity through foramen magnum. Other osseous abnormalities associated with it, are achondroplasia, occipital condyle and clivus hypoplasia, incomplete C1 ring and atlato-occipital assimilation[2, 3, 4]. Associated soft tissue dysgenesis include the chiari I malformation, syringomyelia, syringobulbia and hydrocephalus. These may be present in 35% of the patients[1]. The likely causes could be genetic, mechanical and infection [5, 6]. Goel et al thought that basilar invagination is secondary to malalignment of facets of atlas and axis which leads to slippage of atlas over axis with consequent invagination of odontoid process[7, 8].

There two types of basilar invagination[1]:

1. Anterior variety: - this is where there is shortening of basiocciput so that the clivus is short with resultant upward displacement of foramen magnum out of plane of vertebral column. There is high incidence of herniation syndrome due to compression of the posterior fossa
2. Paramedian variety:-there is associated hypoplasia of occipital condyles such that the clivus becomes dorsally displaced into posterior fossa. However this is ameliorated by the curving of lateral portion of squamo-occipital bones.

Basilar impression is an acquired form of basilar invagination resulting from softened basilar bones. The aetiological agents are infections, rickets, osteogenesis imperfecta, hyperparathyroidism, osteomalacia and Pagets disease. About 80% of those with basilar impression have no associated Chiari malformation, the remainder has both basilar impression and Chiari malformation. The principal pathological characteristic is observed to be direct brainstem compression due to odontoid process indentation in group I and a reduction in posterior cranial fossa volume in group II. Altered CSF dynamics, pressure or traction on the brainstem and possibly impaired vertebrobasilar blood supply are possible mechanisms[9].

Clinical features

Presentation varies depending on degree of compression and by structures affected. The most common manifestations include headache and neck pain due to compression of nerve roots. The pain is aggravated by any rough movements of the head and can be triggered by valsava maneuvers like sneezing, coughing and also bending forward[10]. Depending on abnormalities

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there may have short webbed neck, decreased range of motion and torticollis- incase of unilateral condylar hyperplasia[1].

Sleep apnea, opthalmoplegia, nystagmus, hoarness, dysarthria, and dysphagia, gait disturbances may ensue if there is compression of brain stem, cranial nerves or cerebellum. Vascular compression may cause syncope, vertigo, and loss of consciousness, visual defects or weakness especially with postural changes[10].

Diagnosis

Multi modal imaging facilitates diagnosis and planning of surgery[11], Acute or suddenly progressive deficits are an emergency, requiring immediate imaging. MRI allows for visualization of osseous, vascular and neural tissues like cerebellar, hindbrain, spinal cord, and syringomyelia. CT shows bone structures more accurately than MRI and may be done more easily in an emergency.

If MRI or CT suggests vascular abnormalities, magnetic resonance angiography or vertebral angiography is done. If MRI and CT are unavailable, plain x-rays—lateral view of the skull showing the cervical spine, anteroposterior view, and oblique views of the cervical spine are taken. Basilar invagination is suspected if on plain radiographs C1-C2 facet complex cannot be seen on open mouth view[12].

The following radiological lines are used to make diagnosis:
Chamberlain line- An extension of tip of odontoid beyond 5mm on a line joining the posterior edge of the hard palate to the dorsal lip(opisthion) of foramen magnum[9].
Mcgreror line – It's a line joining the back of hard palate to the lowest point of occipital squama. Tip of odontoid mat normally extend above it by 6mm in women 7mm in men thus beyond 7mm is abnormal[13].
Mcrae line extends from the anterior to posterior rim of foramen magnum, thus tip of odontoid should lie below this line. When there is narrowing of foramen to less 19mm in sagittal plane there is associated neurological deficits.

Treatment

Understanding the two types of basilar invagination is probably the most crucial factor in understanding the various involved management issues as their pathogenesis appears to be different in the two groups. Goel et al [14]stratified patients based on absence or presence of associated chiari malformation classified into Group I and II respectively. Essentially in Group I there is distancing of the odontoid process from the anterior arch, suggested presence of instability of the region and atlantoaxial dislocation. The angle of the clivus and the posterior cranial fossa volume were essentially unaffected in these patients. In Group II, on the other hand, the assembly of the odontoid process, anterior arch of the atlas and the clivus migrated superiorly in unison resulting in reduction of the posterior cranial fossa volume. Thus the Chiari malformation or herniation of the cerebellar tonsil[15].

More recently Goel et al came up with another classification which is based on parameters that determine treatment options[15]. In Group A basilar invagination there was a 'fixed' atlantoaxial dislocation and the tip of the odontoid process 'invaginated' into the foramen magnum and was above the Chamberlain line, McRae line of foramen magnum and Wackenheim's clival line. Group B basilar invagination was where the odontoid process and clivus remained anatomically aligned despite the presence of basilar invagination and other associated anomalies. In this group, the tip of the odontoid process was above Chamberlain's line but below McRae's and Wackenheim's lines. The odontoid process in Group A patients resulted in direct compression of the brainstem. Essentially, in Group A basilar invagination there was an element of instability of the region that was manifested by the tip of the odontoid process distancing itself from the

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anterior arch of the atlas or the lower end of the clivus. In some Group A patients there was Chiari malformation, and this feature differentiates the present classification from the earlier classification. In Group B, the atlantoaxial joints were normal and were normally aligned. In some patients the joints were entirely fused.

The modalities of treatment include; Reduction and immobilization, Sometimes surgical decompression, fixation, or both [10]. If neural structures are compressed, treatment consists of reduction (traction or changes in head position to realign the craniocervical junction and thus relieve neural compression). After reduction, the head and neck are immobilized. Acute or suddenly progressive spinal cord compression requires emergency reduction[10]. For most patients reduction involves skeletal traction with a crown halo ring or Gardner wells tongs. If reduction is achieved, the neck is immobilized in a halo vest for 8 to 12 wk; then x-rays must be taken to confirm stability.

If reduction does not relieve neural compression, surgical decompression, using a ventral or a dorsal approach, is necessary. If instability persists after decompression, posterior fixation is required. For some abnormalities external immobilization alone is rarely successful; if it is unsuccessful, posterior fixation or anterior decompression and stabilization are required. Several different methods of instrumentation like plates or rods with screws can be used for temporary stabilization until bones fuse and stability is permanent. In general, all unstable areas must be fused.

Preoperative traction

It is used achieve to immediate relief of symptoms and it also helps in planning surgical approach. It facilitates assessment of degree to which reduction of the invagination can be achieved. If there is noted reduction then posterior procedures will suffice but if there is no reduction then anterior decompression and posterior stabilization is necessary.

The direction of traction should be optimized to improve neural compression thus head should be elevated at least 15 degrees above the horizontal plane. Lateral cervical radiographs are taken serially as traction weights are increased to assess reduction. It's been noted that 80% of children below 14 years can be treated with traction [16]. Goel et al [14]reported that 82 patients without chiari malformation, 82% improved clinically after traction whereas of the 20 patients with associated chiari malformation who were put on traction only one (5%) patient improved.

The surgical approaches can be ventral where transoropharngeal approach is the most common form of decompression [17, 16, 18]. The posterior approach allows for foramen magnum and posterior cervical decompression as well as stabilization and fusion.

Case Presentation

This was 14 year old male teenager who has had neck pains over the last 18 months. Pain was sharp, initially was at occiput but radiated caudally on the neck. It was aggravated by movement and relieved by extension of the neck. Over last two months had worsened especially when walking down hill. No history of seizures, headache, paresthesia, visual difficulties nor trauma. Symptoms forced him to drop out of school. Physical examination revealed normal motor and s

Treatment

Based on above findings patient was put on traction using Gardner–Wells tong. The initial weight was 2kg and over next 48 hours was increased gradually by 0.5kg up to 6.5kg. During this period he was monitored for any neurological changes. X ray taken showed good reduction

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of the invagination. Traction was maintained during transfer to theatre. He was placed in prone position on May field frame after endoscopic intubation technique.

ensory function. Cranial nerves were unaffected, and reflexes were not exacerbated

Through a midline incision foramen magnum decompression and C1 laminectomy was done. Occipitocervical fusion was undertaken using reconstruction plates, occipital screws and C4 & C5 Lateral mass crews. Bone graft fusion was performed using iliac crest graft. Post operatively, he was put on Halo plaster jacket for five months and was removed after CT scan showed maintenance of the reduction and he did not have symptoms. Thereafter he was reviewed at three month intervals and at fourteen months he was well and had resumed school.

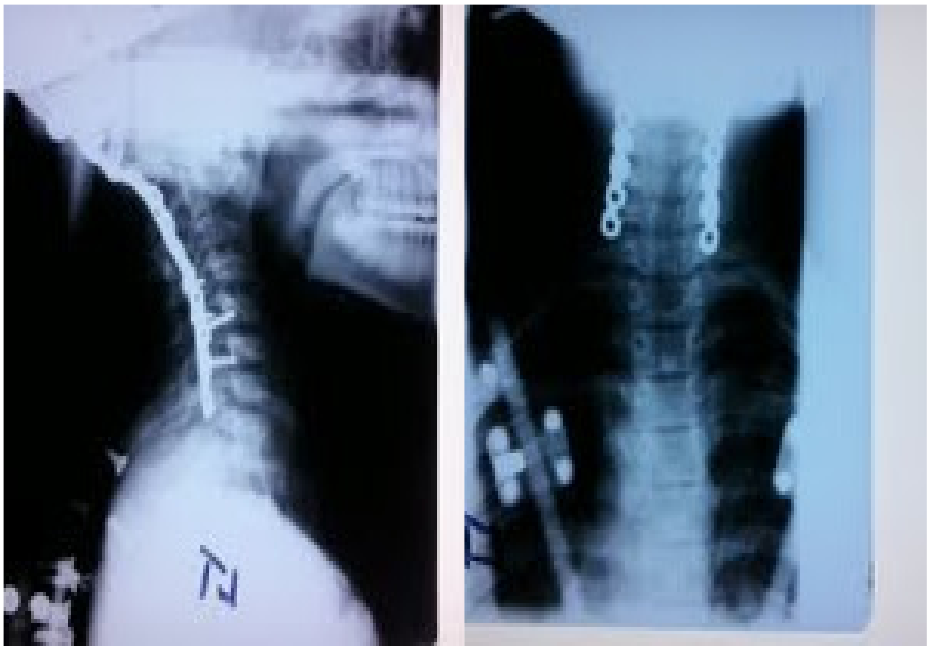

Figure 1. Radiographic examination Findings

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