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Atlanto-axial Dislocation - An Overview

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N H Wadia, - *Bombay*

Congenital cranio-vertebral (CV) anamolies have been known in the literature for a long time and the complications of AAD resulting in myelopathy-immediate, chronic and delayed has also been recognised almost since the turn of the century. So this problem of CV anamolies or AAD could be due to infective, traumatic and congenital etiology. We in India seem to come across many more of these patients compared to other parts of the world, although epidemiologically this fact has not been the proven. The reasons for this are not very clear and 25 years have gone by since we all have been looking at this problem. In late 1950's at J J Hospital attention was drawn to the fact that congenital AAD resulting in neurological deficits was not an uncommon entity, and the 1st paper was published in 1959-60 with first 6 cases. Subsequently there has been an explosion of interest in the subject and neurosurgeons all over the country were busy in operating as these patients were diagnosed. I would say there are few thousand patients on record with neurological complications. Myelopathy is essentially due to AAD and few may be due to basilar invaginations (BI). It has also been pointed very early that BI and AAD upto a point present with different neurological manifestations. If you are going to build surgery on very firm grounds, then we must understand, which of the two the patient is suffering from. So that wrong hope from surgery can be avoided. As you go along the course in the afternoon I hope we will be able to expose you to some points which are troubling, at least me, for the last 25 years in the subject and the approach of surgeons to this problem

The congenital anomalies of AA joint cause essentially a myelopathy, in my opinion. This produces compression. Acute compression will get one set of symptoms, chronic compression produce another set of symptoms. If it is acute on chronic compression a combination of symptoms will occur with both varieties. Each of them can exist independently of the other or collectively. When we take the history of these patients it is important to ask appropriate questions in the chronic cases to get the history of transitory attacks earlier. In an analysis of 106 patients, one fourth had transient attacks and 99 had progressive neurological deficits.

Interestingly, cervical symptoms are not very dominant, so they often come to neurologists or neurosurgeons than to orthopaedic surgeon.

Transitory attacks are diagnostic of AAD and I do not think it is seen in any other condition including basilar invagination. These attacks include unconsciousness, paraesthesia and cortical blindness. First these are self evident with a sudden jerk. One suddenly falls and becomes quadreparetic or paresthesias can occur due to rapid stretch on cervico medullary cord junction. Trauma is so minor that this can not be called traumatic. These are all congenital, simply because there is no significant trauma, no infection or rheumatoid disease. Trauma can be minimal like slight flexion of the neck. Other congenital anomalies are often association. So there is no doubt that they are congenital in nature. The transitory episodes last for less than 24 hours. Residual deficits may be seen clinically if examined after some time or we may see progressive neurological deficits. In transitory attacks, there are no physical signs in between the attacks, except perhaps brisk reflexes which almost always remain as tell tale evidence. Rarely extensor plantars and sensory disturbances and brisk knee jerks are seen. Unconsciousness may last for few minutes to few hours. One of our patient was unconscious for 4 days with quadreplegia, cortical blindness cleared in neat 3 - 4 weeks and she was duly corrected by surgery and had no subsequent symptoms.

Pyramidal signs were prominent in 104 patients. Nearly less than half were asymmetrical. Do not miss hemiplegic

presentation of AAD as cortical lesion, as brisk reflexes are always seen of the otherside, so it is quadreparesis with much greater account on one side of the body

Paraesthesias were spontaneous in 25 patients. Post column deficit was seen in 20. Column of Burdagh is more affected (upper limb) than Gall and can be mistaken for postero lateral sclerosis. Occasionally localised wasting, urinary symptoms are seen in more acute or severe form. Syringomyelic defect can cause sensory deficit on the trunk, quite often immediately after the transitory attack. Cerebellar signs were noted in twelve. There was no dysarthria. Nystagmns of high spinal variety was seen in 12. This set of symptoms can not be mistaken for basilar invagination which should present with lower cranial nerve palsies, increased ICP, pyramidal signs, cerebellar signs. There is a zone of demarcation but it is not always easy.

Forty one patients had pain in the neck, but not as presenting symptoms. Restriction of movements mostly of rotation is a prominent feature. Neck movements were severely restricted in 27. Cervical symptoms are not dominant in AAD. Forty seven had low hair line. But it is seen in one variety of AAD, where there is short neck. This is predominantly seen in men with a mean age of 27 years.

Most of them presented within the first 5 years of symptoms. 35 took more than a year. One came after 35 years with slowly progressive neurological deficits in the last 5 years. Always look for the other congenital anomalies which serve as marker in these cases.

Table 1 - Associated congenital anamolies - Clinical (65 cases)

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Table 2 - Associated congenital anamolies - Radiological (65 cases)

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Management:

In the irreducible variety the surgeons should address to the problem to remove the compressive element. If it is reducible, it can be fused.

1st variety dislocation:

They are fixed variety of true dislocation, with occipitalisation and C 2-3 fusion, where neck is short. Others had small odontoid, occipitalisation and C 2-3 fusion where Mcgregors line and Chamberlins line might suggest BI. But at surgery or at post-mortem it is not BI. Mcgregor and Chamberlin drew their lines in normal skulls and spines. Various anomalies present they were not taken into account seriously. In abnormal necks these lines give wrong impression of BI and lead to wrong type of surgery.

2nd variety of subluxation:

There is free movement of vertebrae. Odontoid is not seen separate. The dislocation reduces in neutral position. As the neck is further extended it goes right back across the wide arc of the circle, through which the cord is embarassed by the movements of a neck. Here surgical procedure required is only fixation.

3rd variety:

Everything looks normal except dislocated odontoid in good shape. There transverse ligament is not

well developed. This is usually seen in adults with no history of injury. Another variety is the one which I would like to call as "vertical dislocation" or BI. The C 2-3 may be united in such cases. Occipitalisation may be present. Odontoid is seen right inside the foramen magnum. Surprisingly there will not be any cranial nerve deficits. With traction it can be brought down. Similar thing does not happen in BI which is a fixed situation. CT scan shows this in lateral reconstruction. Myelogram can be dispensed with CT scan, being available.

Spinal cord pyramidal tract on one side may be demyelinated, other side being intact. Column of Gall may also be demyelinated. Distortion of the cord and unilateral damage will produce asymmetrical pyramidal signs. It does not mean that other side is not affected but it is minimal.

Transitory attacks should be differentiated from transient ischemic attacks in elderly, epileptic drop attacks, multiple sclerosis and hysteria. Progressive myelopathy can occur in cerebral palsy in young kids which starts on 1st day of line, whereas atleast 5 years later is onset in children with AAD. Posterior-lateral sclerosis was the popular diagnosis in 1960. In other words AAD produces cervical cord syndrome and BI leads to post fossa syndrome.

Post decompression resulted in haematomyetia in the past. The whole problem is an enigma. In some patients though the dislocation is partially reduced they walk around with just brisk reflexes only. Nervous system seems to take a lot of strain, which we got to understand yet. In AAD cord is squeezed and something else may also be happening. CT scan excels in delineating exact findings and the degree of compression. Unless the compression is demonstrated therapy can not be planned. If patient has not improved, I feel it has not been sufficiently decompressed.