



ORIGINAL RESEARCH PAPER

Radiology

CRANIO-VERTEBRAL ANOMOLIES :HISTORY AND REVIEW OF LITERATURE

KEY WORDS: Craniovertebral anomalies,Diagnostic Modalities**Dr. Sushil Sikchi**

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ABSTRACT An exhaustive review of literature is performed which reveals that It definitely helps in diagnosis of different anomolies and leads to surgical correction in many cases which was thought to be incurable before the advent of Radiological investigations and merely treated on physical examination. Diagnostic modalities changes the visual acceptance of various cranio vertebral anomolies and pave the way for newer ways of treating the symptomatic patients . Our Fellow researchers efforts are recognized for their great contribution in evaluating and simplifying these complex region.

INTRODUCTION

Before the advent of the diagnostic radiology , the diagnosis of the cervical vertebral anomalies were rarely made antemortem. But with the institution of routine vcervical spine roentgenography they are being diagnosed readily.

For many years anomalies in the region of foremen magnum were of interest mainly to anatomists . in later half of the 19th century they began to be described in the pathological literature and finally it appeared in the British radiological literature.

There are number of abnormalities of craniovertebral area which may produce significant neurological symptoms or be entirely asymptomatic. The severity , duration and nature of the signs and symptoms are dependent on location of lesion and degree of cord or brain- stem compression . Root compression, cord compression, myelopathy, vascular disorder, and internal hydrocephalus all can result from cervical and craniovertebral anomalies. Radiological evaluation is necessary to localized the bony abnormalities and normal data must be available to determine whether the findings seen on radiograph are responsible for patient's symptoms. In many patients diagnosis was not clear on merely physical examination. And number of others were thought to have disorders for which no curative treatment existed (like syringomyelia, brain-stem tumor). It is very important to recognize the craniovertebral anomalies as in some cases surgical correction is possible. This radiology plays an essential part in the diagnosis of craniovertebral anomalies. The importance of the radiological examination is to determine the amount of spaces available for the cord and the brain-stem. Since space varies with the position of the head , dynamic studied may be necessary for complete evaluation. These anomalies were first found to be common in the west. But studies have revealed the high frequency of craniovertebral anomalies in India than in west. Possible explanation for this higher incidence may be related to high incidence of antenatal infections and malnutrition in under-developed countries.

Hence it is necessary to carefully evaluate the craniovertebral junction radiologically and ascertain whether the patient is symptomatic or asymptomatic.

REVIEW OF LITERATURE

Even before the advent of radiology, different craniovertebral anomalies were described in literature.

In 1767 occipito- atlantal fusion was first described by Morgagni.

Basilar impression was not a new disease having been described by ackerman in 1970.

First case of altanto- axial dislocation was reported by Sir Charles Bell in 1830.

First attempt to classify altanto- axial dislocation was made by malgaingol in 1855.

One of the most complete and accurate contribution is that of Boogard in 1865 in whose article many of the recently discovered

facts about the basilar impression may be found.

Boogard in 1865, described the basal angle & also two other measurements which, though not used, are still perfectly valid indices. They are : (1) Fora men Magnum Clivus angle & (2) Boogard's line.

Giacomini (1886) described the type II odontoid dysplasia although he used the term Os odontoideum. He described the first case of congenital altanto- axial dislocation.

A theory advanced by Chiavugi (1895) and later developed by fl man (1905) attribute failure of distal occipitoblast to fuse with others as the cause of abnormal bone formation on the external surface of the skull around the occipital foramen. This phenomenon is called the manifestation of the occipital vertebra.

In 1895 Wilhelm Konard Roentgen, a German physicist discovered the X- rays which was the most important discovery of the Century, which has revolutionized the medical science completely . After the discovery of X-rays many authors focused their attention in studying craniovertebral region.

Dwight (1904) reported the first case of Os odontoideum, and further reported the second case of type II odontoid dysplasia.

Basilar impression was first diagnosed clinically by shuller in 1911.

In 1912, Klippel & Feil described massive fusion of cervical vertebrae in shortneck of French tailor.

Robert (1933) is credited with reporting the first case of agenesis of nteh entire odontoid. The first case of actual absence of odontoid ws reported by weler (1942).

Coutte (1934) stated- the normal altanto- odontoid interval does not exceed 1-2mm.

Chamberlain (1938) described the Chamberlain line for the diagnosis of basilar impression.

Walsh (1940) was the first author since Boogard to draw attention to the fact that basal angle was indeed a bad and inconstant index for basilar impression.

Bull (1946) pointed out that the planes passing by the hardpalate and by the atlas are parallel or they slightly intersect. This relation being altered when there is basilar impression.

McGregor (1948) reviewed the istorical development of measurements for diagnosis of basilar impression and described McGregor line which is the most satisfactory available system for measurement on lateral cervical spine projection.

Type II ossiculum terminals first coined by Hafley in 1948.

Jackson (1950) stated- In adult the altanto odontoid interval not to be greater than 2.5mm and in children as there is a movement of

C1 on C2 with flexion altanto-odontoid interval may be as large as 4.5mm in this position.

Fischgold (1952) described the diagastric line on antero-posterior or laminogram to evaluate basilar impression.

McRae (1953) reported many cases of craniovertebral anomalies of which six cases are of atlanto-axial dislocation, 25 cases of occipitalization of atlas and 11 cases of separate odontoid process of axis and a few cases of basilar invagination.

McRae (1953) stated that if the antero-posterior diameter of spinal column behind the dens is 19mm or less, neurological signs are always present. He considered 20mm to be minimal normal canal depth.

Greenfield (1953) reported another case of os odontoideum. McRae (1953) also reported examples of this condition calling it a separate odontoid process of axis.

McRae(1953) described McRae line for basilar invagination and foramen magnum stenosis.

McRae (1953) observed fusion of C2 & C3 vertebrae in 17 out of 25 cases studied for occipitalization of atlas and also atlanto-axial dislocation in 50% of cases.

Boijesen (1959) stated normal range for measurement of diameter behind dens in male was 19- 32 mm and in female 19-30mm.

Wolf, Khilnani Matis (1956) repeated Boijesen's work and found that average diameter behind dens was 22mm with a range of 16-30mm. If the diameter is between 10-13mm cord compression may or may not be present.

Spillane et al (1957) reported occipitalization 29.2%, atlanto-axial dislocation 8.4% and basilar impression in 24 cases.

Greenberg (1958) stated normal range of sagittal diameter of canal at dens in males is 20.0 to 26.0mm and in females it is 19.0 to 25.0mm.

Cord compression	Behind dens	C2 to C7 (mm)
Always	14 or less	10 or less
Possible	15 to 17	11 to 13
Never	18 and above	19 and above

Wadia M.H (1960) was first to point out that the high frequency of craniovertebral anomalies in India than in West.

In 1960 Hinck & Hopkin revised following formula for atlanto-odontoid interval:
 $Dm = 2.05 - 0.02 A + 1.00mm$
 $Df = 1.24 - 0.0074 A + 0.90mm$
 (A= Age, m= Male, F= Female)

Hope & hopkin (1961) stated McGregor baseline and Fischgold's line seems to be best measurement on lateral and antero-posterior radiograph respectively.

Lombardi (1961) roentgen examination of the craniovertebral junction in consecutive series of 4,000 patients has revealed manifestations of the occipital vertebra in 19 patients (0.47%).

Schmidt & Fischer remarked that there is hardly a variant of occipital bone or of cranial part of the atlas which has not been attributed to it.

McRae and Branum observed- 5 para condylar processes in 25 cases of assimilation of atlas.

Barucha & Dastur (1964) and Singh reported the common abnormalities to be - occipitalization, while atlanto-axial dislocation as next common. Basilar impression and cervical canal stenosis were rare.

Dieckman (1966) in his series of 112 patients found that occipitalization of atlas was in 35% cases and also found evidence of basilar impression in 98 of the 133 patients (other series) of craniovertebral anomalies occurring either alone or in association with other malformations.

In 1966 Rougemont also reported a case of os odontoideum.

Singh S. Datta A. K. Gupta S. (1969) and Patel M.B. & Yajnik V. H. (1971) reported the craniovertebral anomalies in 4.7% and 8.71% cases of spinal cord compression respectively.

Sharma S. R., D.Janki, M. S. Gulati, Dr. S. R. Jayaram, Dr. J. Dar and Dr. S. Gupta, reported that occipitalization of atlas is most common craniovertebral anomaly and the most common combination is that of occipitalization of atlas with basilar invagination and remaining were without basilar invagination. He also stated that although basilar invagination is common, atlanto-axial dislocation and foramen magnum stenosis is more responsible for producing neurological symptoms.

In 1981 R. Shukla, D. Nag, N. N. Gupta and B. N. Lal reported radiological abnormalities seen in 28 patients of clinically suspected cervical cord compression due to craniovertebral anomalies. Occipitalization of atlas was commonest (64.2%) while atlanto-axial dislocation seen in 50% cases. Fusion of cervical vertebrae was next common anomaly. Basilar impression and platybasia is uncommon and multiple abnormalities seen in 32.2%.

N. K. Singh, D. N. Verma, S. K. Gupta, B. C. Katiyar and S. Mohanti (1986).

Roentgen findings in 40 patients of craniovertebral anomalies have been discussed. Various anomalies observed in order of frequency were occipitalization of atlas in 80%; atlanto-axial dislocation in 62%; Klippel-Feil anomalies in 50%; dens dysplasia in 48% and other minor anomalies. Multiple anomalies were present in 80% of patients of which most frequent combination of anomalies was occipitalization, C1 C2 fusion, dens dysplasia and atlanto-axial dislocation. The atlanto-axial dislocation as seen in full flexion view in 25 patients was completely reducible in 12 patients during extension of head. Thus the importance of the routine use of full flexion film has been emphasized. There was statistically significant narrowing of cervical canal from foramen magnum down to C3 level.

RN Hensinger. Clin Orthop Relat Res (264), (1994) in their review on Congenital Anomalies of the Cervical Spine concluded that the majority of afflicted individuals are asymptomatic or have only mild restriction of neck motion. If symptoms develop, they are usually due to cervical instability or degenerative osteoarthritis. Patients with upper cervical anomalies such as atlantooccipital fusion, anomalies of the odontoid, or the transverse atlantal ligament have a great propensity to develop early instability and neurologic problems secondary to minor traumatic events.

Tiwari, A. et.al, in their paper Congenital Abnormal Cervical Vertebrae-A Case Report Abstract(2002). Concluded that Normal typical cervical vertebrae from 3rd to 6th are characterised by small body, triangular spinal canal, foramina transversarium, superior articular facet directed backward and upward, inferior articular facet directed forwards and downwards.

Myung-Sang Moon et.al, in their paper on Radiographic assessment of congenital C2-3 synostosis (2010) evaluated the morphologies of congenital C2-3 synostosis in the cephalad and caudal adjacent discs and facet joints of 25 patients.

Veena Vidya Shankar* and Roopa R Kulkarni in their paper titled, Block vertebra: fusion of axis with the third cervical vertebra - a case report (2011) Concluded Skeletal abnormalities at the craniocervical junction or cervical region may result in severe neck pain and sudden unexpected death.

In their review paper titled Congenital fusion of cervical vertebrae: a review on embryological etiology, Mohammad Mardani (Ph.D) et.al , (2016),detailed Scrutiny of embryological etiology of anomalies plays an important role in reducing the incidence of this anomalies.

Buse Kayhan1 et.al, (2015), Occipitocervical synostosis: case report Occipitocervical synostosis is clinically significant as it may cause narrowing of foramen magnum which may compress the brainstem, vertebral artery and cranial nerves. It should be noted that this pathology may cause a wide range of clinical features, and neurosurgeons, orthopedic surgeons, radiologists and anesthesiologists should be aware during their interventions.

Mohammad Javad et.al, their review paper titled Congenital fusion of cervical vertebrae: a review on embryological etiology,(2016) Congenital fusion of cervical vertebrae is a rare anomaly. In this condition, two fused vertebrae appear structurally and functionally as one. This anomaly may be symptomatic or asymptomatic.

Atlanto-occipital fusion and its neurological complications: a case report by the authors

Campos, D.1,2*, Silva, TH.3, Ellwanger, JH.4, Goerck, ML.5, Kipper, JF.3, Piazza, JL.3 and Kraether Neto, L.3(2016) During routine activities in the Laboratory of Human Anatomy at the University of Santa Cruz do Sul – Brazil, an atlanto-occipital fusion was observed in a Caucasian cadaver skull. The skull used in our study had complete fusion of the occipital bone with the atlas vertebra, except in traffic areas of the vertebral arteries. Some important neurological disorders seem to be related with atlanto-occipital fusion.

CONCLUSION

With above discussion of review that since last four decades there is significant evolution in radiological diagnosis of numerous varieties of cranio vertebral and cervical anomalies, Fellow researchers have contributed a lot in differentiating and diagnosing the different types of cranio vertebral anomalies in patients with associated minor anomalies whether they are symptomatic or asymptomatic in patients. Gradual evolution helps in projecting the newer and multiple anomalies at cranio vertebral and cervical region.It definitely helps in diagnosis of different anomalies and leads to surgical correction in many cases which was thought to be incurable before the advent of Radiological investigations and merely treated on the basis of physical examination.Diagnostic modalities changes the visual acceptance of various cranio vertebral anomalies and pave the way for newer ways of treating the symptomatic patients. Our Fellow researchers efforts are recognized for their great contribution in evaluating and simplifying this complex region.

It is remarkable that all are in the view that , occipitalisation of Atlas , congenital fusion of vertebra, atlanto axial dislocation ,basilar impression or invagination are the most common cranio vertebral anomalies followed by , os odontoideum and platybasia. Even they noticed that multiple other bony minor anomalies like homovertebral bone , sprengele shoulder,hemivertebra etc. are associate with primary anomaly.

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