Bipartite Atlas –A Rare Entity, a Study of Its Incidence in North Indians.

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ABSTRACT

Background: Combined anterior and posterior C1 bifidity i.e BIPARTITE ATLAS or SPLIT ATLAS is a rare entity. Our goal in this study was to examine the incidence of this anomaly in North India. review the literature and the clinical implications of this cleft defects.. Methods: It is retrospective study of NCCT evaluation in 1735 patients who underwent CECT neck [900 patients] NCCT with spine evaluation [835 patients] in Panaj diagnostic set up between Jan 2012 to December 2016, and department of radiodiagnosis Sarojni Naidu Medical college Agra during the time interval between Dec 2016 to July 2017 the images were reviewed retrospectively to identify patients with BIPARTITE ATLAS defects. Posterior arch defects of the atlas were grouped in accordance with the classification of Currarino et al. anomalies were subsequently grouped. Results: We found only 4 (4/1735) patients of bipartite atlas i.e. 2% of total. anterior arch defects were midline and have a reported width ranging from 1-5 mm The type A posterior arch defect was found in 3 patients and the type B posterior arch defect was found in one patient. No type C, D, or E defects were observed in these bipartite atlas defect The CT scans of the patients show midline clefts of the anterior and the posterior arches of C1 with similar imaging features: smooth margins lined by cortical bone and no lateral offset. The patients had no neurological symptoms relating to the C1 abnormality, and no follow-up was performed. Conclusion: Bipartite atlas is a rare entity as it’s incidence was found to be just 2%. knowledge is essential as it can predispose to certain neurological compressive disorders, awareness helps in clinical radiological diagnosis, management in patient of trauma as appearance of cleft simulate Jefferson fracture.

Keywords: Bipartite atlas, cleft defect, Arch of C1, CT scan.

INTRODUCTION

A split or bipartite atlas is a congenital abnormality that results in a coexisting anterior and posterior schisis due to the failure of fusion of the anterior and posterior arches [Figure 1]. This complete bipartition of the atlas has been reported in only 0.1% of the population (Hummel et al., 2013) (Tachibana et al).¹ ²

In cervical spine trauma, profound knowledge of congenital atlas defects is crucial. Malformations, where C1/C2 junction might be compromised, have to be distinguished from fractures. SPINE SURGERIES on one hand where they are helping patient in allaying their problems, on other hand they are increasing morbidity if not properly done like C1 laminectomy procedures increases the risk of anterior arch fractures, and if this is associated with posterior arch defect it simulate bipartite atlas so knowledge of this variant of atlantal arch defect is essential. Currarino et al.³ and Geipel⁴ reported the incidence of atlantal arch defects by a cervical radiographic study and a cadaveric study, respectively. Few cadaveric and imaging studies have been reported on the BIPARTITE ATLAS anomaly detection of these anomalies is clinically important because they can cause neurologic compressive disorders and so we took this study.

Figure 1: Bipartite atlas in a 12 year old boy CT a.axial view   b.THREE dimensional view.
MATERIALS AND METHODS

The records of 1735 patients who presented with various medical problems visiting a major diagnostic set up PANKAJ CT SCAN b/w jan 2012 to dec 2016 and in dept of radiodiagnosis SNMC agra during the time interval b/w between december 2016 to july 2017 were reviewed retrospectively. The craniovertebral computed tomography (CT) scans done in these patients were evaluated to see the cervical spine morphology.

Table 1: Classification of congenital anomalies of the posterior arch of the atlas according to Currarino et al.[3]

| Type | Discription |
|------|-------------|
| A    | Failure of posterior midline fusion of the two hemarches |
| B    | Bilateral defect |
| C    | Bilateral Defect |
| D    | Absence of the posterior arch, with persistent posterior tubercle |
| E    | Absence of the entire arch, including the tubercle |

Table 2: Type of posterior arch defect in bipartite atlas found, and associated anomaly present in these patients.

| Patient No. | S e x | Age (yr.) | Type of posterior arch defect | Anterior arch defect | Associated anomaly | SYMPTOMS | DIAGNOSIS |
|-------------|------|-----------|-----------------------------|---------------------|-------------------|----------|-----------|
| 1           | M    | 12 yrs    | A                           | Midline anterior cleft | cleft palate       | headache, neck pain precipitated due to somersaults | cervical sprain |
| 2           | F    | 16 yrs    | A                           | Midline anterior cleft | absence of pedicle of C7 | neck pain | cervical pain |
| 3           | M    | 35 yrs    | A                           | Midline anterior cleft | palpable neck mass | Rough voice | larynx cancer |
| 4           | M    | 39 yrs    | A                           | Midline anterior cleft | none               | cervical sprain |

The patients clinical history had varied medical problems, including weakness, palpable neck mass, posterior neck pain, radiolucopathy due to degenerative disease or after traffic accident, arm pain, sore throat. When a congenital defect of the atlas arch was identified on a CT scan, the patient's medical record was reviewed to determine his or her neurological status. An associated anomaly was searched by retrospective review of X-ray and CT findings. Exclusion criteria were incomplete visualization of the CVJ [cervicovertebral junction], radiologic evidence of traumatic lesion at CVJ, severe degeneration, previous operation, known skeletal dysplasia, and maturation delay.

We evaluated consecutive neck (3 mm interval image from lower occiput to second thoracic vertebra) and cervical spine (2 mm interval image from lower occiput to second thoracic vertebra) CT scans. At our institution, CT was done on GE OPTIMA 660, 64-slice CT scanner. Imaging parameters were as follows: 2.75 mm slice thickness, 0.75 s/rotation, 120 kV, and 300 mA. Reconstruction was done with a slice thickness of 1.0 mm.

At Pankaj diagnostic set up CT machine used wasGE Bright speed 16 slice, imaging parameters were2.5mm slice thickness, 120Kv;160mA reconstruction was done with slice thickness .6 mm. Posterior arch defects of the atlas were grouped in accordance with the classification of Currarino et al.[3] Who have divided posterior arch defects anomaly into five types [Table 1], depending on the extent of absence of the posterior arch and the presence or absence of the posterior tubercle.

In all cases anterior ossification center did not develop and lateral masses did not fuse anteriorly resulting in anterior cleft [Figure 3], the majority of anterior ach defects are midline and have a reported width ranging from 1-5 mm.[9]

Clinical Implications and Incidence of Bipartite Atlas

Atasoy et al. reported the first case of bipartite atlas with os odontoideum causing spinal stenosis.[6] Garg et al.[7] presented a report of bipartite atlas with anterior arch aplasia associated to an os odontoideum. They found a small projection on the anterior surface of the dens and concluded that the ossification centre of anterior arch of atlas may fail to separate from future dens resulting in anterior arch aplasia with a small tubercle attached to the anterior surface of the dens. Patients with a bipartite atlas are usually
asymptomatic and these are often detected incidentally during an imaging procedure (Childers et al., 1971).[9] at times they are discovered during the workup for neck pain (Phan et al., 1998).[9] When symptomatic, patients can present with chronic symptoms of neck pain or neurological abnormalities; in case reports by Sharma et al.[10] 2000 Klimo et al. 2003 Caminos et al., 2008,[11,12] patients of bipartite atlas presented with cervical myelopathy, which occur with anomalies involving types C and D of the Currarino classification These types contain the posterior osseous fragment that may be mobile and possibly displaces forward during extension of the neck, thereby interacting with the cervical cord and producing symptoms; Klimo et al.[11] established this mechanism using fluoroscopy in a teenage patient that presented with loss of sensation in both lower limbs and left arm accompanied by L’Hermitte’s sign. After surgically removal of the fragment, the patient’s symptoms completely resolved.[13] Type E arch defect was observed in a child of nine year old boy by Corominas et al., 2010 who reported with 2 month history of headache localized to the occipital region. Also had neck pain increased by hyperextension.[13] Tachibana et al.,[2] reports a case of 3year and 11 months old girl that presented with a two–week duration of persistent torticollis, head tilt and neck rotation, with a characteristic cock–robins position after sustaining a minor fall, torticollis was due to a specific C1 dislocation with a congenital split atlas after minor trauma. The cervical range of motion was severely restricted due to severe neck pain, but there were no neurological abnormalities. Imaging revealed malformed anterior and posterior arches in the midline region (split atlas) as well as a slight leftward shift of the odontoid process (Tachibana et al., 2010) A rare case of bipartite atlas vertebra in a 10 years old boy,with fusion of C2 and C3 and cord compression was reported by bala chandran, MR images showed subluxation of anterior atlanto-axial joint,extreme narrowing of cervical cord, Posterior arch of atlas was seen compressing the cord reconstructed coronal images revealed split atlas. The cord compression was removed at surgery and patient has had a dramatic improvement.[14] Anthony I petralgia reports a case of Bipartite atlas in a collegiate football player of 19 years with neck pain and upper extremity paresthesias after sustaining a tackle that forced neck hyperextension.[15] In more severe cases, it can present acutely after trauma to the head and neck region as neurological symptoms (Pasku et al., 2007).[16] Hummel et al. 2008, 3 patients of split atlas presented after being involved in a traumatic injury involving the head and neck. It was discovered that all the patients had an incidental finding of complete failure of fusion of both the anterior and posterior arches of the atlas. There is a relationship between certain congenital diseases that appear to have an impact on connective tissue, such as Down syndrome, and the presence of a split atlas, which can be a result of ligamentous laxity. This theory attributes that the ligamentous laxity causes a greater amount of range of motion during gestation that may lead to failure of fusion of the arches (Hummel et al., 2013).[11] Galindo and Francis reported the incidence of anteroposterior spondyloysis of the atlas in normal individuals as 0.3%.6 In a study of atlantal arch defects Jong Kyu Kwon et al reports one patient with a type A posterior arch defect had an anterior atlantal-arch midline cleft (1/1153, 0.087%).[17] Sebastian Guenkel, et al states that “A bipartite spondyloysis was present in 2 cases of our cohort (4.8% of all atlas arch defects, 0.2% of all patients,”[18] as shown in [Table 3] NOT much literature exist till date about the incidence of bipartite atlas usually case reports are present our study appears to be 1st in north India.

| S.N | Author | Region | Year of Study | Incidence of Bipartite Atlas |
|-----|--------|--------|---------------|-----------------------------|
| 1   | Galindo and Francis[6] | KOREA | 1983 | 0.3% |
| 2   | Jong Kyu Kwon, M.D et al[12] | KOREA | 2009 | 1/1153, 0.087% |
| 3   | Sebastian Guenkel et al[11] | zurich | 2013 | 0.2% |
| 4   | PRESENT STUDY | NORTH INDIANS | 2016 | 4/1735=2% |

RESULTS & DISCUSSION

We found only 4 patients of bipartite atlas i.e .2% of total Bipartite atlas or split atlas defects were found in 4 (4/1735,0.23%) of the 1,735 patients.similar to finding of Sebastian guenkal et al.[18] The type A posterior arch defect was found in 3 patients and the type B posterior arch defect was found in one patient. No type C, D, or E defects were observed in these bipartite atlas defect. Congenital anomalies of the atlantal arch frequently occur with various combinations of bone and spinal cord abnormalities, suggesting an interrelationship exists.[19] In our study [Table 2] shows the associated anomalies One patient (patient 1) had associated cleft palate, patient 2 also had absent seventh cervical spine pedicle, while 3rd pt showed partial posterior posticulus None of the reviewed patients of bipartite atlas had neurological deficits because of atlantal arch anomalies but off and on cervical neck pain with radiation in arm, relieved by analgesics was common. The CT scans of the patients show midline clefs of the anterior and the posterior arches of C1 with similar imaging features: smooth margins lined by
cortical bone and no lateral offset. The patients had no neurological symptoms relating to the C1 abnormality, and no follow-up was performed. The clefts at level C1 are the result of the failure of three ossification centers to fuse properly. Anterior and posterior clefts are caused by hypoplasia of the hypochondral bow and lateral parts of the C1 sclerotome respectively.[20] Because of the risk of instability, assessing atlantoaxial stability is advised.

Clefts of the atlas are known to simulate fractures. However, because the two lateral masses in a bipartite atlas are essentially loose bones connected by incompetent soft tissues,[21,22] axial loading forces can split them apart similar to the mechanism of a Jefferson’s fracture hence BIPARTITE ATLAS can create an image on X-rays similar to that of a Jefferson fracture.[23] There are differences in these two conditions that enable us to distinguish one from the other. Generally, the lateral translation of the lateral masses with a congenital anomaly is 1 to 2 mm, and with a Jefferson fracture they are more than 3 mm.[24] In a young population (3 months to 4 years old), the lateral masses of the atlas commonly extend 1-3 mm beyond the margins of the axis, secondary to different growth patterns of the vertebral bodies. This is called pseudospread of the atlas.[21] The classic radiographic feature of a Jefferson fracture is a bilateral atlanto-axial lateral offset of 3-9 mm.[23] A lateral translation of more than 7 mm is an indication of transverse ligament damage, which causes instability in the upper cervical complex.[24] A CT with three-dimensional reconstruction is extremely helpful in evaluating the integrity of the atlas and differentiating an acute injury from developmental cleft. On imaging, the fractures show irregular edges with associated soft-tissue swelling, while congenital clefts are smooth with an intact cortical wall and have no associated soft-tissue swelling, in congenital bipartite atlas lateral offset, if present, is not more than 1-2 mm while in case of fracture it is usually >3 mm. In all 4 patients of our study there was no lateral off set [Figure 4]

Cervical instability is suggested by history, symptoms, and radiographic evidence of non-physiologic motion between C1 and C2 [disturbed atlantoaxial stability] and 3 mm or more lateral displacement of the lateral masses compared with the dens. However, the imaging findings in the patients of bipartite atlas in our study indicated that no acute fracture or instability existed, no lateral offset and confirmed the presence of a congenital nonunion of the anterior and posterior arches of C1 with fibrous nonunion of the anterior arch. However, in patients with neurological symptoms, we believe that MRI should be performed to evaluate the spinal cord and adjacent neural structures adequately[17] and to identify the transverse atlantoaxial ligament between C1-2, which when intact indicates stability of atlantoaxial joint and neurological symptoms are not associated. but importance of this anatomical variant can not be overlooked as it can predispose to neurological compressive disorders.

CONCLUSION

Our study reports an incidence of BIPARTITE ATLAS to be .2% similar to Sebastian Guenkel, et al. Awareness and knowledge of this anomaly of atlas i.e bipartite atlas is important as our eyes sees what our mind knows, so knowing the catastrophic complications bipartite atlas can cause one will surely give eye to this anomaly of cervical region, this will surely add to helps in clinicoradiological diagnosis and management of patients of trauma as appearance of bipartite atlas simulate fracture.

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