Case Report

Cervical Vertebrae Anomalies—Incidental Findings on Lateral Cephalograms

Priyavrat Soni[a]; Vineet Sharma[b]; Jaideep Sengupta[c]

ABSTRACT

Three cases of abnormal incidental findings on lateral cephalogram are presented. These patients reported for orthodontic consultation in their adolescence. While studying the patients' cephalograms, abnormal radiographic findings were discovered in their cervical vertebrae. Because the patients were asymptomatic, early diagnosis based on these radiographic findings made the patients aware of the situations. Lifestyle changes were instituted with specialist consultation in two patients to prevent or delay the onset of symptoms of an underlying pathology. Patients were educated about the likely future course of these findings. Specialist follow-up was advised to all the patients.

KEY WORDS: Atlanto-occipital assimilation; Block vertebra; Os Odontoideum; ADI; Basilar invagination

INTRODUCTION

The lateral cephalogram is the most common diagnostic radiograph used in clinical orthodontics. The cervical spine area present in lateral cephalograms is generally omitted in cephalometric tracings. Though the cervical vertebrae maturation index (CVMI)[1] is now commonly used to interpret the growth potential of young patients, adequate attention is not paid to the radiological and morphological anatomy of this region. Cephalograms must be studied comprehensively and traced in a stepwise manner so that any deviations from normal in the cervical spine area can be detected.[2]

Cattel and Filtzer[3] studied 160 children and found variations from normal due to displacement of vertebrae that resembled subluxation, spasm, and ligamentous injury related to skeletal growth centers resembling fractures. Farman and Escobar[4] described the radiographic appearance of congenital anomalies of the vertebral bodies due to defects in fusion or normal segmentation, occipitalization of the atlas, basilar impression, odontoid malformations, atlas malformations, spina bifida, and abnormal ossifications. These findings can be of great importance for patients in whom these anomalies assume clinical significance necessitating management or lifestyle changes to prevent or delay aggravation of the pathology.

Roentgenographic examination of the cervical spine may reveal a pathologic disorder in asymptomatic as well as in symptomatic subjects. Orthodontists do not have to be experts in cervical vertebrae abnormalities, but they must be aware of the normal radiological anatomy of the cervical spine on the lateral cephalogram. Many abnormalities of the cervical spine do not manifest themselves symptomatically until adolescence or young adulthood, and the orthodontist may be the first person to detect some of these. Progressively degenerative defects, if discerned early, may help in mitigation of the severity of their consequences.

CASE REPORT 1

A 14-year-old girl of average build reported for orthodontic treatment of her forwardly placed and spaced teeth. During the diagnostic work-up the lateral cephalogram showed a skeletal Class II pattern with abnormalities in the cervical spine area. The posteroanterior (PA) cephalogram showed asymmetrical facial structures. To get more details, radiographs of the

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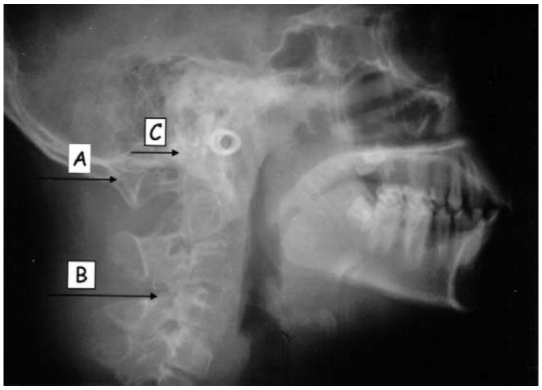

Figure 1. Lateral cephalogram showing occipitalization of the atlas along with block vertebra C2–C3 and basilar invagination.

cervical spine in lateral flexion and extension and a PA cephalogram in the open mouth position were taken with a radiologist's consultation. These radiographs showed atlanto-occipital assimilation. In addition, there was basilar invagination with block vertebra of C2 and C3 and multiple craniovertebral junctions and cervicovertebral anomalies (Figure 1).

Assimilation of Atlas to Occiput (Occipitalization of the Atlas, Block Atlas)

Occipitalization is a congenital synostosis of the atlas to the occiput caused by a failure of segmentation and separation of the most caudal occipital sclerotome during the first few weeks of fetal life.[5] Columbo first described the condition in 1577, followed by Rokitansky in 1844 and Macalister in 1892. The first radiological demonstration of occipitalization was performed by Schuller in 1911. There may be varying degrees of bony fusion between atlas and occiput; complete and partial assimilation have been described. In a majority of cases, assimilation occurs between the anterior arch of the atlas and the anterior rim of the foramen magnum and is associated with other skeletal malformations such as basilar invagination, occipital vertebra, spina bifida of atlas, or fusion of the second and third cervical vertebrae (Klippel-Feil syndrome).[6] In complete occipitalization the anterior arch of the atlas fuses with the lower end of the clivus, and the posterior arch is often fused with the squamous occiput.[7]

Etiology. Atlas assimilation is defined as failure of segmentation between the last occipital and first cervical sclerotome. Given the described genetic background, revealed by experimental analysis of transgenic mice model systems, assimilation of atlas into the occiput might be interpreted as an anterior homeotic transformation.[6]

Clinical findings. A short neck, low hair line, and restricted neck motion are found in more than two-thirds of the cases. There is male dominance of up to 5:1, with a population incidence as high as 0.75%–3%.[5,8] Generally, young patients are asymptomatic, but the risk for significant neurological dysfunction increases with age, usually beginning in the third or fourth decade of life. Repetitive microtrauma, a minor injury, or even a trivial event such as sneezing may initiate symptoms.[8–10] When the odontoid process encroaches into the foramen magnum, anterior neuroaxial compression and associated neurologic problems are common. The following most common signs and symptoms occur, in decreasing order of frequency: pain in the occiput and neck, vertigo, unsteady gait, paresis of the limbs, paresthesias, speech disturbances, hoarseness, double vision, syncope, auditory disturbances, interference with swallowing, occipital headaches, blurring of vision, neurological deficit, and, rarely, sudden death.[5,7,11,12]

Radiological findings. Most commonly, the anterior arch of the atlas is assimilated into the occiput. Up to 70% of occipitalizations have an accompanying fusion or block vertebra of C2–C3.[8] Cervical radiographs usually demonstrate assimilation of the atlas to the basiocciput, and flexion-extension views may detect instability at the C1–C2 articulation, especially in cases with associated C2–C3 fusion.[6] In 90% of the cases, details of the fused posterior arch of atlas can be discerned at the occiput on a plain film. MRI and CT may be necessary to clarify the pathologic condition.[5]

Treatment. Nonoperative measures such as cervical collars, braces, plaster, and traction should be attempted initially in some symptomatic patients because aggressive management may be hazardous. Surgical intervention carries a much higher risk of morbidity and mortality.[12] Awareness and monitoring of complications need to be instituted. Appropriate counseling on the management of known risk factors, including therapeutic options that can precipitate complication should be encouraged.[5] Patients are advised not to expose themselves to undue trauma. Extension and rotational maneuvers may place the spinal cord and vertebral artery at risk.

Basilar Invagination (Basilar Impression)

Basilar invagination describes the condition of a relatively cephalad position of the upper cervical vertebra to the base of the skull.[5] Chamberlain first described the condition in 1939. There are two types of basilar impressions: (1) primary, a congenital abnormality often associated with other vertebral defects such as atlanto-occipital fusion, hypoplasia of the atlas, bifid posterior arch of the atlas, Klippel-Feil syndrome, and Goldenhar syndrome; and (2) secondary, a developmental condition usually attributed to softening of the osseous structures at the base of the skull, with the

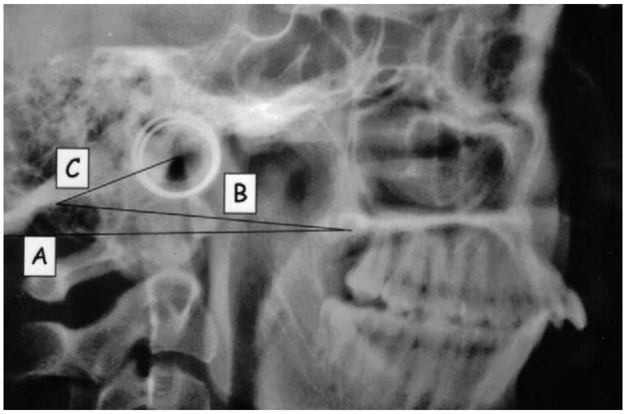

Figure 2. McGregor line (A), Chamberlain line (B), and McRae's line (C) can be easily drawn on a lateral cephalogram in a suspected case.

deformity developing later in life. This is occasionally seen in conditions such as osteomalacia, rickets, Paget disease, osteogenesis imperfecta, renal osteodystrophy, and rheumatoid arthritis.[12]

Radiological findings. Basilar invagination is difficult to diagnose, though radiological diagnosis can be established with the help of three constructed lines on the lateral cephalogram.[5,6,12] These lines denote the extent of protrusion of the tip of the odontoid process into the foramen magnum. A person is said to have basilar impression if the lateral cephalogram (Figure 2) shows the tip of the odontoid process to be:

- More than 5 mm beyond the McGregor line (from hard palate to the lowest part of the occiput);
- More than 2.5 mm beyond the Chamberlain line (from hard palate to the posterior rim of the foramen magnum); or
- Lying above McRae's line (from the anterior to the posterior rim of the foramen magnum).

Clinical findings. Common signs and symptoms include muscle weakness, neck pain, posterior column dysfunction, bowel and bladder disturbance, and paraesthesia.[13] Basilar impression can also lead to sudden hearing loss, pyramidal tract signs, and wasting of the upper limbs. On recognition, one must consider MRI to assess for neurological and vascular complication.[5]

CASE REPORT 2

A 13-year-old girl reported to the orthodontic department with a chief complaint of an irregular arrangement of teeth. On routine cephalometric examination, an abnormality was noted in the cervical spine region. On radiological consultation, an MRI scan was done and a definitive diagnosis of os odontoideum with increased atlantodental interval (ADI) was made. She also had atlanto-occipital assimilation (Figures 3 through 5).

Os Odontoideum

Os odontoideum is defined as nonunion of the dens with the axis body.[5] The absence of a united odontoid process results in a separate ossicle with smooth circumferential margins that has no connection with the body of C2, leaving a variable gap between itself and the small odontoid process.[6] It was first described in 1886. Clinically, it usually is associated with atlantoaxial instability, local mechanical irritation (neck pain and torticollis), progressive myelopathy, or transient neurologic symptoms secondary to vertebral artery compression.[14]

Etiology. The etiology of os odontoideum is controversial, although most authors now agree that it most likely results from an unrecognized injury to the odontoid process in childhood.[15–17] Because os odontoideum is generally not seen in children younger than 5 years of age, it is unlikely that it is congenitally acquired.[6]

Radiological findings. The ossicles may be readily overlooked or obscured and detection requires scrupulous attention to detail.[5] In adults the diagnosis of os odontoideum is suggested by observing a radiolucent defect between the dens and the body of the axis.[12] The os odontoideum may be in the normal position of the odontoid process (orthotopic), displaced cranially (dystopic), or fused to the clivus.[6] The line separating an os odontoideum from the body of the axis usually is cranial to the level of the superior articular facets of the axis. Flexion-extension cervical spine radiographs usually demonstrate instability and movement between the os and body of the axis. An anteroposterior open-mouth view, CT with a bony window, and two- or three-dimensional reconstruction readily identifies the abnormality. An MRI allows visualization of the cervical spinal cord and myelomalacia secondary to atlantoaxial instability.[6] Os odontoideum should be differentially diagnosed from the fracture of the dens.[11]

Clinical findings. Familial incidence has been reported, and associated conditions include twins, Down syndrome, occipitalization, block vertebra, and Klippel-Feil syndrome.[5,18] The majority of the patients are asymptomatic and the anomaly is found incidentally on radiographs taken for unrelated conditions. The average age of discovery is between 19 and 30 years, and there are no gender differences.[15,16] Suboccipital pain and neuralgia, audible crepitus, and jerky motion on sagittal flexion-extension may be present.[5]

Treatment. Instability of C1 on C2 secondary to os odontoideum carries the risk of damage to the spinal

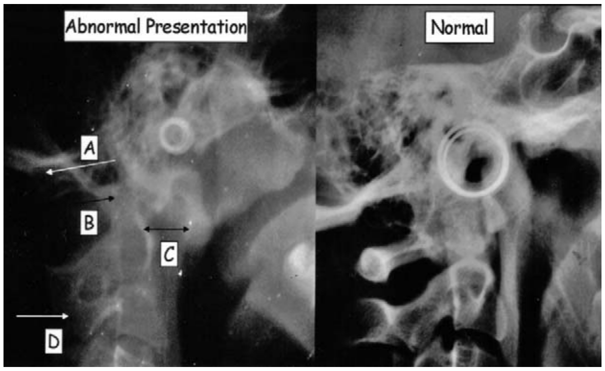

Figure 3. (a) Abnormal presentation in a lateral cephalogram with atlas occipitalization (A), os odontoideum (B), increased atlantodental interval (C), and block vertebra C2–C3 (D). (b) Normal presentation of the cervical spine.

cord or vertebral arteries. High-velocity spinal manipulative techniques are contraindicated in patients with these conditions. Anesthetic risk during intubation with neck extension needs to be considered.[5] Atlantoaxial instability, if present, requires some form of posterior stabilization. The best fusion rates have been reported with C1–C2 transarticular screw fixation. In the presence of os odontoideum, even minor trauma may produce symptoms and upper cervical cord compromise; therefore, preventive stabilization should be considered.

Atlantodental Interval (ADI)

The atlas-dens interval is the space seen on the lateral cephalogram between the anterior aspect of the dens and the posterior aspect of the anterior ring of the atlas (Figures 3 through 5). In children the ADI should be no greater than 4 mm. The upper limit of normal in adults is less than 3 mm.[16] Fielding[15] and others noted that for this interval to increase, the transverse ligament must be attenuated. When the ADI increases to 5 mm or greater, the transverse ligament and accessory ligament, as well as the accessory stabilizing ligaments have been ruptured.[15,19]

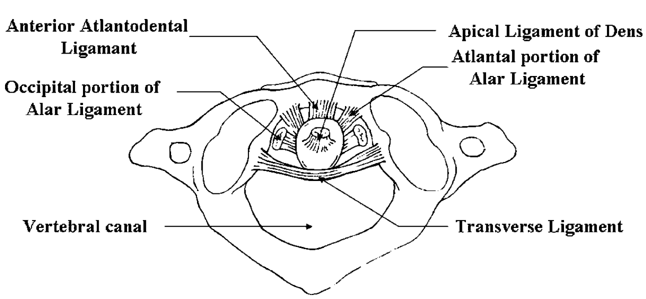

Figure 4. Relationships of the odontoid process in transverse plane. Note that the odontoid process is supported by the transverse ligament posteriorly. If the atlantodental interval increases, it leads to rupture of the transverse ligament and other supporting ligaments.

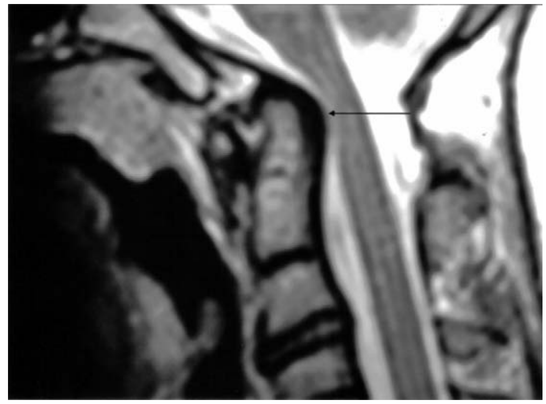

Figure 5. MRI scan of the same patient showing rupture of transverse ligament and compression of the dura mater space.

CASE REPORT 3

A 17-year-old girl reported for orthodontic correction with a periodontally compromised dentition with pathologic migration and a traumatic bite. On lateral cephalogram she presented with block vertebra C2–C3 (Figure 6).

Block Vertebra (Congenital Synostosis, Congenital Vertebral Fusion)

Fusion of one or more contiguous vertebral segments results from the embryological failure of normal spinal segmentation. This most likely is the result of a local decreased blood supply during the third to eighth week of fetal development.[5] C2–C3 is the most common site, and the incidence is 0.4% to 0.7% with no sex predilection. Patients are generally asymptomatic,[5] but increasing age and injury may precipitate symptoms. Premature degenerative changes at adjoining motion segments are common as this condition results in greater biomechanical stress in the adjoining segments. Discal tear, rupture of the transverse ligament, fracture of the odontoid process, and spondylosis are common consequences.

CONCLUSIONS

This case series emphasizes the importance of not overlooking abnormal cervical spine findings. By the

Figure 6. Lateral cephalogram showing the block vertebra C2–C3.

time this article was submitted, our first patient had already started developing temporal and occipital headaches which are a manifestation of occipitalization. Concurrent appearance of symptoms may lead to the patient blaming the symptoms on the initiation of orthodontic treatment. Early diagnosis of these pathologies on cephalograms not only gives patients a chance to live life normally by changing their lifestyles, but it can also provide useful documentation of change due to an injury, aging, or progression of a degenerative process. This consequently may influence a medical diagnosis or litigation.

REFERENCES

1. Hassel B, Farman AG. Skeletal maturation evaluation using cervical vertebrae. *Am J Orthod Dentofacial Orthop.* 1995; 107:58–66.
2. Vastardis H, Evans CA. Evaluation of cervical spine abnormalities on cephalometric radiographs. *Am J Orthod Dentofacial Orthop.* 1996;109:581–588.
3. Cattell H, Filtzer DL. Pseudosubluxation and other normal variations in the cervical spine in children. A study of one hundred and sixty children. *J Bone Joint Surg Am.* 1965;47: 1295–1309.
4. Farman AG, Escobar V. Radiographic appearance of the cervical vertebrae in normal and abnormal development. *Br J Oral Surg.* 1982;20:264–274.
5. Guebert GM, Rowe LJ, Yochum TR, Thompson JR, Masia CJ. Congenital anomalies and normal skeletal variants. In: Yochum TR, Rowe LJ, eds. *Essentials of Skeletal Radiology.* 3rd ed. Baltimore: Lippincott Williams & Wilkins; 2005: 257–403.
6. Dávid KM, Crockard A. Congenital malformations of the base of the skull, atlas, and the dens. In: Clark CR, ed. *The Cervical Spine.* 4th ed. Philadelphia: Lippincott Williams & Wilkins; 2005:420–425.
7. Spillane JD, Pallis C, Jones AM. Developmental abnormalities in the region of the foramen magnum. *Brain.* 1957;80: 11–48.
8. Bharucha EP, Dastur HM. Craniovertebral anomalies: a case report of 40 cases. *Brain.* 1964;87:469–480.
9. Bassi P, Corona C, Contri P, Paiocchi A, Loiero M, Mangoni A. Congenital basilar impressions: correlated neurological syndromes. *Eur Neurol.* 1992;32:238–243.
10. Sherk HH. Lesions of the atlas and axis. *Clin Orthop Relat Res.* 1975;109:33–41.
11. Holt RG, Helms CA, Munk PL, Gillespy T 3rd. Hypertrophy of C-1 anterior arch: useful signs to distinguish os odontoideum from acute dens fracture. *Radiology.* 1989;173: 207–209.
12. Hensinger RN. Congenital anomalies of the cervical spine. In: Rothman RH, Simeone FA, eds. *The Spine.* Vol 1. 4th ed. Philadelphia: WB Saunders Company; 1999:232–245.
13. Goel A, Bhatjiwale M, Desai K. Basilar invagination: a study based on 190 surgically treated patients. *J Neurosurg.* 1998;88:962–968.
14. Pizzutilo PD, Horn BD. Congenital anomalies of the odontoid. In: Clark CR, ed. *The Cervical Spine.* 3rd ed. Philadelphia: Lippincott-Raven; 1998:334.
15. Fielding JW, Hensinger RN, Hawkins RJ. Os odontoideum. *J Bone Joint Surg Am.* 1980;62:376–383.
16. Hukuda S, Ota H, Okabe N, Tazima K. Traumatic atlantoaxial dislocation causing os odontoideum in infants. *Spine.* 1980;5:207–210.
17. McRae DL, Barnum AS. Occipitalization of the atlas. *Am J Roentgenol Radium Ther Nucl Med.* 1953;70:23–46.
18. Morgan MK, Onofrio BM, Bender CE. Familial os odontoideum. Case report. *J Neurosurg.* 1989;70:636–639.
19. Fielding JW, Griffin PP. Os odontoideum: an acquired lesion. *J Bone Joint Surg Am.* 1974;56:187–190.