CASE REPORT

What the mind does not know, the eyes do not see: a rare congenital fusion of the odontoid process to the atlantal hemiarch

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SUMMARY

Congenital spinal fusion of an odontoid process to an atlantal hemiarch is very rare. The unfamiliarity of the medical fraternity with this congenital malformation can easily be mistaken for an acute fracture, chronic infection or inflammatory disease. We present our experience of managing an adult who presented with neck pain after a motor vehicle accident. Radiological investigation revealed congenital fusion of the odontoid process to the atlantal hemiarch. The prevalence, embryology and clinical significance of this anomaly are discussed. As the natural progression of this anomaly is not well documented, we suggest periodic follow-up to monitor the progression of degenerative changes and instability of the occipitoatlantal junction.

produced similar radiological images. Our examination did not reveal any syndromic features.

TREATMENT

The patient was discharged with a soft cervical collar and, 2 weeks later, reassessed with dynamic lateral cervical spine X-rays, which were found to be normal.

DISCUSSION

Fusion of the atlas and the odontoid process is probably the rarest among cervical spine anomalies.¹ Cave¹ described three types of atlantoaxial fusion: (1) fusion of a separated odontoid process with the anterior atlantal arch; (2) complete bilateral fusion of atlas and axis and (3) incomplete

BACKGROUND

Congenital spinal fusion of an odontoid to an atlantal hemiarch is very rare. It can be mistaken for an acute fracture, chronic infection or inflammatory disease. We present our experience of managing an adult who presented with neck pain after a motor vehicle accident, with X-rays suggestive of a fracture of the atlas. The literature review records this as the seventh reported case.

CASE PRESENTATION

An 18-year-old man presented at our accident and emergency department after a motor vehicle accident. He was an unhelmeted pillion rider on a motorcycle. He reported mild upper neck pain with slight restriction of motion. There was no obvious tenderness observed. Neurological examination was unremarkable. He had a history of childhood epilepsy.

INVESTIGATIONS

The patient was referred on the suspicion of a C1 fracture, which was detected in a normal cervical spine X-ray. In view of a lack of clinical signs to support the diagnosis of C1 fracture, we ordered a CT of the cervical spine. The result confirmed our suspicion of a congenital defect of the anterior arch of the atlas. Incidentally, this patient also had bony fusion of the odontoid process to the C1 anterior arch (figure 1).

DIFFERENTIAL DIAGNOSIS

The patient disclaimed previous episodes of inflammatory arthritis or infection; these might have

Figure 1 CT scan showing a small defect with smooth corticated margin of the anterior arch of the atlas, with fusion of the odontoid process to the atlantal hemiarch.

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Figure 2 Ossification centres of the anterior arch of the atlas (above). Resegmentation of the fourth occipital, and first and second cervical sclerotomes, forms the primitive vertebral bodies of the atlas and axis (below).

(unilateral) fusion with or without some degree of assimilation (other congenital abnormalities). Wackenheim² was the first to describe fusion of a non-separated odontoid process to atlas. The presence of a non-separated odontoid fusion to atlas without assimilation, such as in our patient, fits into type 3 of Cave's classification, which is rarer.³

The prevalence of fissures in the posterior atlantal arch is 4%, as reported by Geipel.⁴ An anterior cleft is very rare, with prevalence of 0.09–0.1%.^{5–8} Usually, the anterior cleft is associated with posterior arch anomaly. The C1 clefts could be associated with Down syndrome, Arnold-Chiari malformation and gonadal dysgenesis. Bipartite atlas, an association of both fissures, anterior and posterior, has been described in the literature.^{9–13} An anterior defect alone is very rare and has been seen in only a few patients.¹⁴ Clefts of the anterior arch are usually very narrow, and may be median or paramedian.¹⁵

So, the unilateral fusion of non-separated odontoid to atlantal arch with median cleft is very rare. Our literature review indicates that this is only the seventh reported case.¹⁶ The unfamiliarity of the medical fraternity with this congenital malformation can easily lead to it being mistaken for an acute fracture, chronic infection or inflammatory disease.

Embryologically, the caudal part of the fourth occipital sclerotome and cranial part of the first cervical sclerotome contribute to the formation of the atlas (figure 2). The caudal portion of the first cervical sclerotome and cranial portion of the second give rise to the axis. The odontoid process, which is the centrum of the atlas, descends and fuses with the vertebral body of the axis. This fusion between the axis and odontoid process could be explained as a segmentation defect in the sclerotomic material of the first cervical somites.

Three primary ossification centres of the atlas normally appear. The two lateral ossification centres usually unite with the anterior centre of the anterior arch at 5–9 years of age. The absence of an anterior ossification centre or failure of fusion of two lateral centres could cause a defect over the anterior arch (figure 2).

The atlanto-odontoid fusion could be due to congenital or acquired conditions. Therefore, acquired conditions such as tuberculosis, rheumatoid arthritis and trauma, and the accompanying clinical symptoms, should be ruled out.

Since this condition is very rare, there is a paucity of knowledge regarding the progression and degenerative changes accompanying it. Patients should be assessed regularly for signs of myelopathy (weakness and paraesthesia of the upper limbs and occasionally the lower limbs) and occipitoatlantal junction instability (headache, dizziness, neck stiffness and pain).

The anterior atlantal arch cleft could be confused with a fracture. There is rarely complete aplasia of the anterior arch, which is associated with craniocervical instability.¹⁷ A congenital cleft of the atlas arch can be determined in CT scan images, which demonstrate a small defect with smooth corticated margin. Although harmless, a variant of this, the split atlas, has been associated with asymptomatic atlantoaxial subluxation.¹⁸

Learning points

- Congenital spinal fusion of the odontoid process to the atlantal hemiarch is very rare.
- The unfamiliarity of the medical fraternity with this congenital malformation can easily be mistaken for an acute fracture, chronic infection or inflammatory disease.
- Periodic follow-up is required to monitor the progression of degenerative changes and instability of the occipitoatlantal junction.

Competing interests None declared.

Patient consent Obtained.

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REFERENCES

- 1 Cave AJ. On fusion of atlas and axis vertebra. *J Anat* 1930;64:337–43.
- 2 Wackenheim A. C1–2 block vertebra. *Neuroradiology* 1978;16:416–17.
- 3 Pérez-Vallina JR, Riaño-Galán I, Cobo-Ruisánchez A, *et al.* Congenital anomaly of craniovertebral junction: atlas–dens fusion with C1 anterior arch cleft. *J Spinal Disord Tech* 2002;15:84–7.
- 4 Geipel P. [Studies on the fissure formation of the atlas and epistropheus. IV]. *Zentralbl Allg Pathol* 1955;94:19–84.
- 5 Garg A, Gaikwad SB, Gupta V, *et al.* Bipartite atlas with os odontoideum: case report. *Spine* 2004;29:E35–8.
- 6 Martich V, Ben-Ami T, Yousefzadeh DK, *et al.* Hypoplastic posterior arch of C-1 in children with Down syndrome: a double jeopardy. *Radiology* 1992;183:125–8.
- 7 Senoglu M, Safavi-Abbasi S, Theodore N, *et al.* The frequency and clinical significance of congenital defects of the posterior and anterior arch of the atlas. *J Neurosurg Spine* 2007;7:399–402.
- 8 Torriani M, Lourenco JL. Agenesis of the posterior arch of the atlas. *Rev Hosp Clin Fac Med Sao Paulo* 2002;57:73–6.
- 9 Teichert G. Isolated fissures in the anterior atlas arch; case report. *Zentralbl Chir* 1956;81:316–18.
- 10 Childers JC, Wilson FC. Bipartite atlas. *J Bone Joint Surg [Am]* 1971;53:578–82.
- 11 Desgrez H, Gentaz R, Cheverl JP. Anomalies congénitales des arcs de l'atlas. *J Radiol Electrol* 1965;46:819–26.
- 12 Friedman MB, Jacobs LH. Case report: computed tomography of congenital clefts of the atlas. *Comput Radiol* 1985;9:185–7.
- 13 Haakonsen M, Gudmundsen TE, Hestøl O. Midline anterior and posterior atlas clefts may simulate a Jefferson fracture. A report of 2 cases. *Acta Orthop Scand* 1995;66:369–71.
- 14 Lipson SJ, Mazur J. Anteroposterior spondyloschisis of the atlas revealed by computerized tomography scanning. *J Bone Joint Surg [Am]* 1978;60:1104–5.
- 15 Chambers AA, Gaskill MF. Midline anterior atlas clefts: CT findings. *J Comput Assist Tomogr* 1992;16:868–70.
- 16 Tubbs RS, Tyler-Kabara EC, Salter EG, *et al.* Unusual finding of the craniocervical junction. *Clin Anat* 2005;18:449–5.
- 17 Brocher JE, Masset A. Aplasia of the anterior atlas arch. *Fortschr Geb Rontgenstr Nuklearmed* 1965;102:465–7.
- 18 Park SY, Kang DH, Lee CH, *et al.* Combined congenital anterior and posterior midline cleft of the atlas associated with asymptomatic lateral atlantoaxial subluxation. *J Korean Neurosurg Soc* 2006;40:44–6.

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