■ CASE REPORT

Congenital absence of the posterior arch of the atlas associated with a fracture of the anterior arch

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Structural defects of the posterior arch of the atlas are rare, and range from clefts of variable location and size to more extensive defects such as complete agenesis. These abnormalities are usually incidental radiological findings. We present a case of a fracture of the anterior arch of the atlas associated with a congenital abnormality of the posterior arch.

Congenital partial or complete aplasia of the posterior arch of the atlas is usually an incidental finding^{1,2} during the investigation of neck pain, radiculopathy, swelling in the neck or trauma. Careful differentiation between an acute burst fracture and a congenital defect is essential. A CT scan is helpful in evaluating the integrity of the atlas and differentiating an acute injury from a developmental cleft.³

It is unclear as to whether this is a hereditary defect, although two reports have documented the existence of cases involving a mother and son.^{1,4} The incidence between genders is similar and the clinical presentation is variable. Patients are most commonly asymptomatic, although the defect can cause chronic cervical pain, headache and L'Hermitte's sign.⁵

Despite being well-documented, the rarity of the condition leads to errors in diagnosis, and to the lack of confirmation of stability which can result in increased neurological morbidity.⁶ Identifying the transverse atlantoaxial ligament between C1-2 in cases of complete agenesis of the posterior arch is important, since its absence can lead to neurological sequelae, such as atlantoaxial instability or transient quadriplegia.⁷

We present a patient with a previously unreported combination of a fracture of the anterior arch and complete absence of the posterior arch.

Case report

A previously healthy, right-handed, nine-year-old boy presented with a history of occipital headache for two months after axial compression of his neck while performing somersaults. He reported that the pain increased with hyperextension of the neck. He had no other symptoms. On examination, there was tenderness and localised swelling at the occipitocervical junction and slight restriction of

flexion of the neck. Neurological examination was normal and there were no other musculoskeletal injuries.

Radiographs demonstrated a defect in the posterior arch of the atlas, with no evidence of atlantoaxial instability as assessed by flexion and extension views (Fig. 1). A helical CT scan confirmed complete absence of ossification of the posterior arch of C1, corresponding with type E of the Curriano classification.^{1,8} A three-dimensional reconstruction gave an additional perspective (Fig. 2). The CT scan also showed appearances suggestive of a vertical fracture through the anterior arch of C1, with early callus formation. It appeared as a cleft with sclerotic margins. MRI of the cervical spine confirmed the existence of the posterior ligament between C1 and C2, providing evidence for the stability of the congenital aplasia (Fig. 3).

The patient was treated conservatively, with non-steroidal anti-inflammatory drugs and a cervical collar for three months. He was referred for physiotherapy and was asked to avoid contact sports and athletic activities. On his last follow-up after 18 months, he was asymptomatic and fully active.

Discussion

The atlas can be divided anatomically into three parts: the anterior arch, the lateral masses, and the posterior arch. The anterior arch ossifies from one or two centres or, in the absence of a separate centre of ossification, by extension of the lateral masses. Ossification is usually complete by ten years of age. It begins in the posterior arch during the seventh week of intrauterine life, proceeding perichondrally from two centres located in the lateral masses.⁹ The laminae arise from buds in these growth

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Fig. 1a

Fig. 1b

Flexion (a) and extension (b) radiographs showing no atlantoaxial instability.

Fig. 2a

Fig. 2b

Fig. 2c

Three-dimensional reconstruction of C1 and C2 showing absence of the posterior arch and a fracture of the anterior arch of C1 as seen from a) superior, b) posterior and c) lateral views.

centres and extend dorsally, becoming nearly fused by birth except for several millimetres of cartilage. Complete fusion of the posterior arch is expected between three to five years of age. In about 2% of the population an additional centre may occur posteriorly in the midline, forming the posterior tubercle of the atlas during the second year of life.¹⁰

At least two different anomalies can develop during ossification, namely median clefts of the posterior arch of C1 and varying degrees of aplasia of the posterior arch.^{11,12} Based on studies conducted by Geipel¹³ and Currarino et al,¹ these findings have been classified into five different

Fig. 3

Cervical MR image showing the intact transverse atlantoaxial ligament (white arrow).

subtypes. In type A there is a median cleft of the posterior arch of C1. The cleft is unilateral in type B and bilateral in type C. In type D there is absence of the posterior tubercle and in type E there is total agenesis of the posterior arch. Type A clefts occur in about 4% of the population and represent 97% of all posterior defects, whereas types B to E are reported to occur in 0.69% of the population.¹⁴ The presence of an isolated posterior tubercle in types C and D is clinically important because these anomalies can often cause acute neurological deficits such as transient quadriplegia, paraparesis and L'Hermite's sign, chronic neck pain and headache.¹⁵ These clinical deficits, have been attributed to anomalies in the cartilaginous development of the posterior arch rather than to disturbances of ossification.¹⁶

Total or partial aplasia of the posterior atlas arch is rare. Although generally asymptomatic, it may cause neck pain and there is a very rare subset of young patients who develop chronic neurological deficits and atlantoaxial instability.¹⁷ Defects of the posterior arch commonly occur as isolated abnormalities, yet they may be accompanied by other malformations of the cervical spine, mostly in the atlantoaxial region. A cleft in the midline of the anterior arch of C1 associated with a posterior midline cleft, also known as bipartite atlas, is rare.¹⁸ This was not observed in our case, although there was a cleft with sclerotic margins causing pain, stiffness and muscle spasm of the neck.

Clefts of the atlas are known to simulate fractures.^{17,19} 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. CT with three-dimensional reconstruction is extremely helpful in evaluating these spinal anomalies. However, in patients with neurological symptoms, we believe that MRI should be performed to evaluate the spinal

cord and adjacent neural structures adequately²⁰ and to identify the transverse atlantoaxial ligament between C1-2.

We have presented a case of absence of the posterior arch of the atlas associated with fracture of the anterior arch. Treatment of these anomalies should depend on the presence or absence of atlantoaxial instability, with or without neurological symptoms. MRI should be performed to assess the stability of the upper cervical spine, and whether the agenesis is combined with a vertical fracture of the anterior arch.

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