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CASE REPORT

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Aplasia of the posterior arc of the atlas with persistent posterior tubercle: a case report

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Abstract An unusual case of partial aplasia of the posterior arc of the atlas, with persistent posterior tubercle, is presented in a previously healthy individual who sustained a neck trauma. Both plain X-rays and CT findings specified the lesion. Dynamic X-rays in flexion and extension showed an immobile posterior tubercle. The patient did not develop neurological symptoms at any stage during follow-up (1 year).

Keywords Aplasia · Posterior arc · Atlas · Posterior tubercle

Case report

A 24-year-old man was admitted after a road traffic accident. He was a motorbike helmet-protected driver, riding at 50 km/h on a wet road, when the bike was hit by a car and skidded off the road. He was ejected from the bike and landed on the ground. When he arrived in the emergency department, he complained of pain at the back of his neck, but he had no neurological symptoms from the limbs. On physical examination, he had generalized tenderness over his cervical spine in the midline and slight restriction of flexion, without evidence of neurological signs. He had no other injuries to the locomotor system, the chest or to the viscera.

Plain X-rays of his cervical spine revealed a free bony fragment in the lateral aspect of the posterior arc of the atlas that was initially interpreted as a fracture (Fig. 1). On detailed examination, the lesion was found to have smooth edges, whereas the posterior arc of the atlas could not be clearly visualized. A computed tomography scan of the C1/C2 region was therefore obtained, to further delineate the pathology. This confirmed the presence of posterior arch aplasia, a butterfly-shaped atlas and a persistent posterior tubercle. A helical-CT reconstruction with 1.5-mm-thick sections of the cervico-occipital region provided an additional perspective of this anomaly (Fig. 2). No evidence of involvement of neural structures was found. The patient was admitted for observation and reassessment. A Philadelphia collar was applied and nonsteroidal anti-inflammatory drugs (NSAIDs) were administered. The patient was asymptomatic the day after, and dynamic plain radiographs of his cervical spine, both in flexion and extension, were performed to assess the potential mobility of the bony fragment (Fig. 3). As the fragment was stable both in flexion and extension, and the patient did not develop neurological symptoms at any stage during follow-up (1 year), magnetic resonance imaging was not performed.

Anatomic features

The atlas (C1) is anatomically divided into three parts; the anterior arch, the lateral masses and the posterior arch. Ossification begins from the two lateral masses at

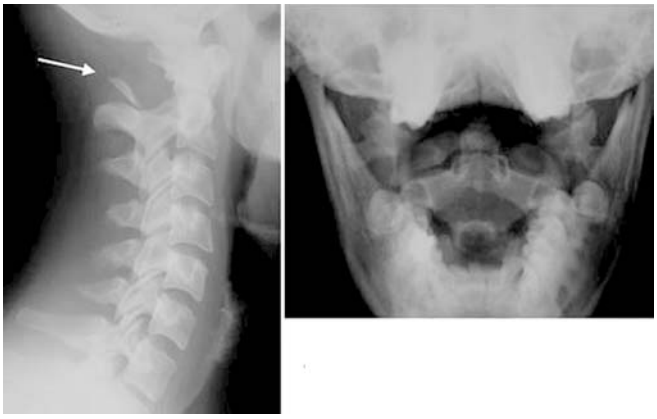

Fig. 1 Initial radiographic control, showing a bony fragment in the lateral aspect of the atlas (*arrow*)

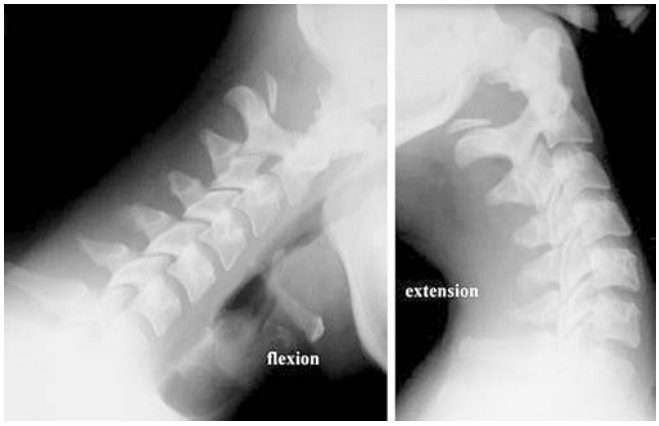

Fig. 3 Dynamic plain films in both flexion and extension showed no movement of the persistent posterior tubercle

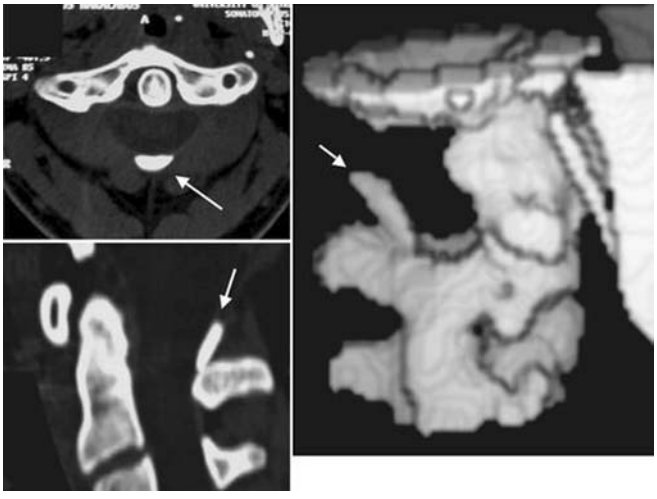

Fig. 2 CT scanning of the C1–C2 region showing aplasia of the posterior arc, butterfly-shaped body of the atlas and persistent posterior tubercle (*arrows*). A helical-CT reconstruction confirmed the diagnosis

the seventh week of intrauterine life, extending dorsally. Between the two bony posterior hemi-arches, there is a remnant cartilaginous cleft, which begins to get ossified during the second year after birth and is completed around the fourth year [10]. This region may arise from a separate ossification center in 2% of the population, where it forms the posterior tubercle in the second year of life [1].

Anomalies of the posterior arch have been reported to occur in 4% of 1,613 dissections [4] and can be of two types: median clefts or hypoplasia. Currarino et al. [2] proposed a classification in five types (A–E) for congenital defects of the posterior arch of C1. Posterior midline fusion defects with a remaining small gap represent the commonest type, A, and are considered to result from a failure in local chondrogenesis. Unilateral clefts with defects range from a small gap to a complete absence of the hemi-arch (type B). Bilateral clefts with preservation of the most dorsal part of the arch (type C) and complete absence of the posterior arch, with (type D) or without (type E) a persistent posterior tubercle, have a combined incidence of 0.69% among the general population. Patients with lesions type C and D, like our patient, have a free-floating posterior tubercle at the apex of the arch. It is hypothesized that to form this anomaly, the patient has both an error of chondrogenesis as well as the rare fourth ossification center, as described previously.

Clinical presentation

Partial or complete aplasia may be asymptomatic, discovered incidentally. In other cases, such anomalies may be presented with transient neck pain or even different degrees of cord compression, including myelopathy [1, 6]. Congenital aplasia or hypoplasia of the posterior arch of the atlas may be associated with several diseases, such as the Arnold-Chiari malformation, gonadal dysgenesis, Klippel-Feil syndrome, and Down and Turner syndromes [9].

Among the different types of posterior arch defects, types C and D are more prone to cause symptoms spontaneously or after trauma [3]. Defects of the posterior arch of the atlas may increase the risk of atlantoaxial subluxation. However, it has been shown, by autopsy studies and surgical findings, that the gap in the posterior arch of the atlas of these patients is bridged by connective tissue [7]. This tissue, with the cleft that may exist, moves in dissociation from the anterior arch. Extension of the neck reduces the distance between the occiput and the spinous process of the axis, displacing the bony fragment anteriorly. This causes impingement of the cord during extension of the neck [8]. Klimo et al.

[6] reported in 2003 a case of type C aplasia of the posterior arch associated with neurological symptoms of sensory deficit in distal lower extremities, transient quadriparesis after a minor fall and positive Lhermitte sign only with neck extension. Their review of the literature revealed another 16 cases that presented with symptoms of myelopathy related to an isolated posterior tubercle (types C and D). Of these, only three patients had documented movement of this tubercle during extension. Our patient had no neurological symptoms and the tubercle remained stable during extension of the cervical spine.

Imaging studies

Plain X-rays (lateral C-spine) will demonstrate bilateral defects (types C–E), whereas oblique views can demonstrate a unilateral defect (type B). Other abnormalities, including clefts of the anterior arch, atlantoaxial rotatory subluxation or hypertrophic downward projection of the posterior border of the foramen magnum, have been also described in the literature [5, 9]. Flexion/extension views can demonstrate anterior displacement of the persistent tubercle and exclude atlantoaxial instability. Computed tomography scanning combined with three-dimensional reconstruction will demonstrate the true nature of the defect and can resolve the diagnostic confusion that is created by plain radiographs. In the presence of neurological deficit, magnetic resonance imaging should be performed to address potential cord contusion. Focal-increased T2 signal within the cord, at the level of the anomaly, can be observed.

Treatment

Currarino et al. [2] concluded that patients with posterior arch defects could be presented in one of five ways:

- Group 1, the lesion is discovered with imaging studies obtained for unrelated reasons in asymptomatic patients
- Group 2, patients presented with neck pain related to trauma and undergo imaging studies
- Group 3, patients who develop sudden neurologic symptoms after head or neck trauma
- Group 4, patients who had a variety of neurologic symptoms before the discovery of the anomaly
- Group 5, the lesion is discovered during a workup for chronic neck pain

The treatment of patients who present with neurological deficit has to be surgical, with excision of the persistent posterior tubercle and posterior ligament between C1 and C2, after ruling out atlantoaxial instability [3, 6]. If the later is present, posterior C1–C2 fusion using interarticular screws or posterior occipitoaxial fusion, are suggested.

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