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Case Report

“Split atlas” in a trauma and nontrauma patient: two different case reports for a rare congenital malformation [☆]

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ABSTRACT

Congenital atlas abnormalities are rare and often asymptomatic findings, accidentally detected in trauma and, more rarely, in nontrauma patients. Rachischisis in both anterior and posterior atlas arches, condition defined as “split atlas,” is extremely uncommon and it may well be confused with fracture. Being able to discriminate between these 2 conditions is an essential step in patient care management. In this article, we report 2 cases of split atlas ascertained in both trauma and nontrauma patients. The first concerning a 54-year-old man fell from a 2 m scaffold, and the second related to a 25-year-old woman suffering from treatment-resistant headaches. Subsequently we proceed to analyse the embryology of these abnormalities, and later to discuss pitfalls, tips and tricks useful to a correct diagnosis, in order to achieve an accurate management of split atlas. Specifically, we outline the crucial radiological features to identify, that are beneficial to an efficient differential diagnosis between congenital atlas abnormalities and fracture. These include smooth corticated margins of the cleft, and <3 mm lateral displacement of C1 lateral masses.

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Introduction

Congenital atlas abnormalities are rare malformations. They include aplasia, hypoplasia, and various arch clefts, such as split atlas. They may be asymptomatic or associated to neurological symptoms, and they represent a diagnostic challenge because can mimic C1 fractures [1]. In this article we report two cases of split atlas in a trauma and in a nontrauma patient, and subsequently we define methods to differentiate

cleft from a fracture, to achieve an accurate patient care management.

Case report 1

A 54-year-old man was admitted to emergency department after a 2-m fall. The patient was immobilized in a Philadelphia cervical collar and in a long spine board. He was awake,

[☆] Written informed consent for publication of their clinical details and clinical images was obtained from the patients.

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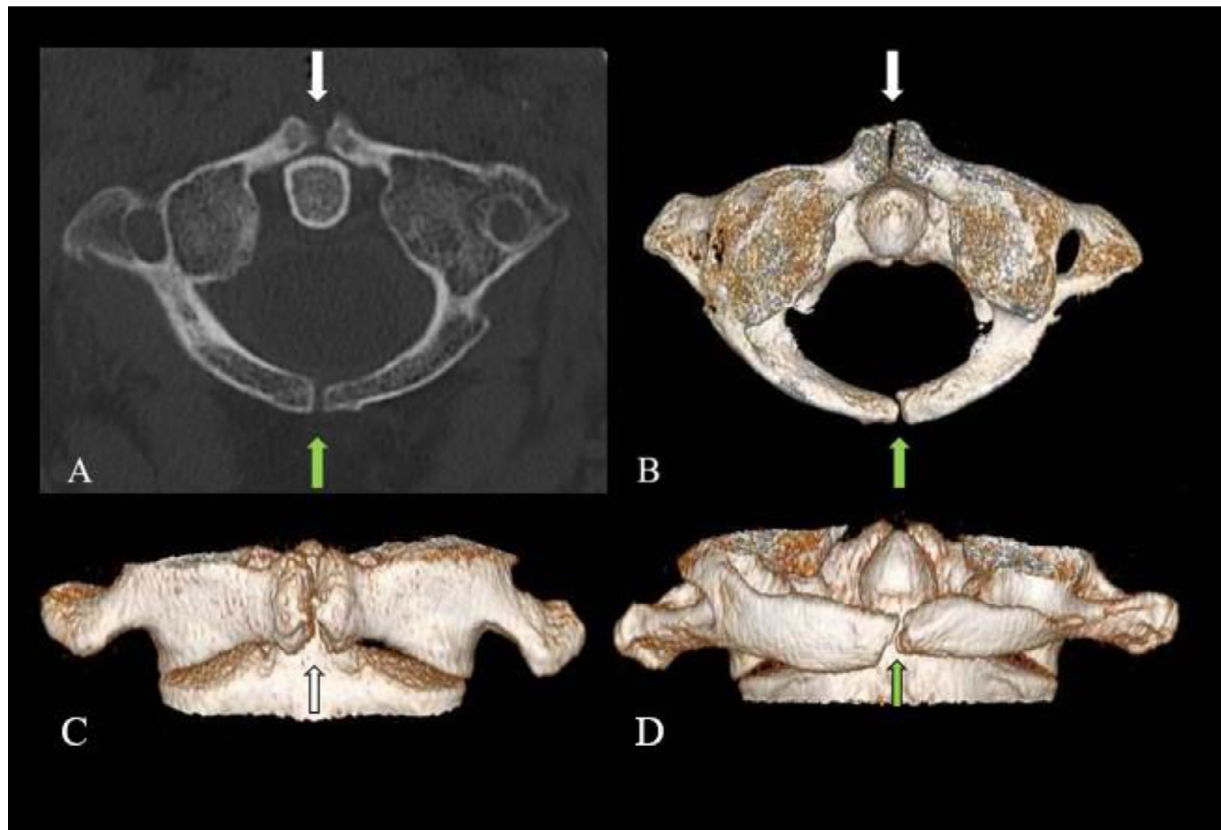


Fig. 1 – Case report 1: Trauma patient. Noncontrast computed tomography (NCCT) of cervical. Axial (A) and three-dimensional (B, axial; C, anterior; D, posterior) reconstruction of atlas CT showed rachischisis in both anterior (white arrow) and posterior (green arrow) atlas arches with smooth corticated margins and displacement <3 mm of C1 lateral masses. (Color version of figure is available online.)

alert, oriented and he reported head, neck, and left anterior hemithorax pain. The abdominal examination was negative. The neurological examination was negative. The patient denied any history of surgical procedures or previous traumas. Plain film cervical and chest X-ray were not performed due to the obvious technical limitations caused by the immobilization. Noncontrast computed tomography (NCCT) of head, cervical, and thorax was performed. Focused assessment with sonography in trauma was used for the evaluation of any abdominal injury.

Head NCCT scan was negative for traumatic injury. Thorax NCCT scan showed multiple left anterior rib fractures (V-VI-VII-VIII ribs).

Cervical NCCT showed a midline rachischisis in both anterior and posterior atlas arches, along with smooth corticated margins and displacement <3 mm of C1 lateral masses (Fig. 1). No spinal cord damage estimated at NCCT. Philadelphia cervical collar and spine board were removed, and the patient was kept under observation for 24 hours.

Case report 2

A 25-year-old woman was admitted to emergency department for treatment-resistant headaches she suffered from since

few days. The patient denied previous traumas. The neurological examination was negative. A head NCCT scan was performed and excluded spontaneous intracerebral hemorrhage or expansive lesions. As a collateral finding, it was observed a midline anteroposterior rachischisis, with well-corticated border and displacement <3 mm of atlas lateral masses (Fig. 2). In this case, the diagnosis of split atlas has been more immediate compared to the previous case. The young woman was discharged after the prescription of different classes of analgesics.

Discussion

Congenital atlas abnormalities are rare and include aplasia, hypoplasia, and various arch clefts, such as split atlas. They are considered by some to be benign variations, accidentally discovered during radiological evaluations mainly performed on trauma patients [1]. Isolated posterior defect is the most frequent abnormality (5.16%) followed by combined anterior and posterior defects (0.46%). One isolated anterior arch defect is the rarest (0.031%). Females have a higher prevalence than males [2]. Rachischisis in both anterior and posterior atlas arch is usually called “split atlas” [1].

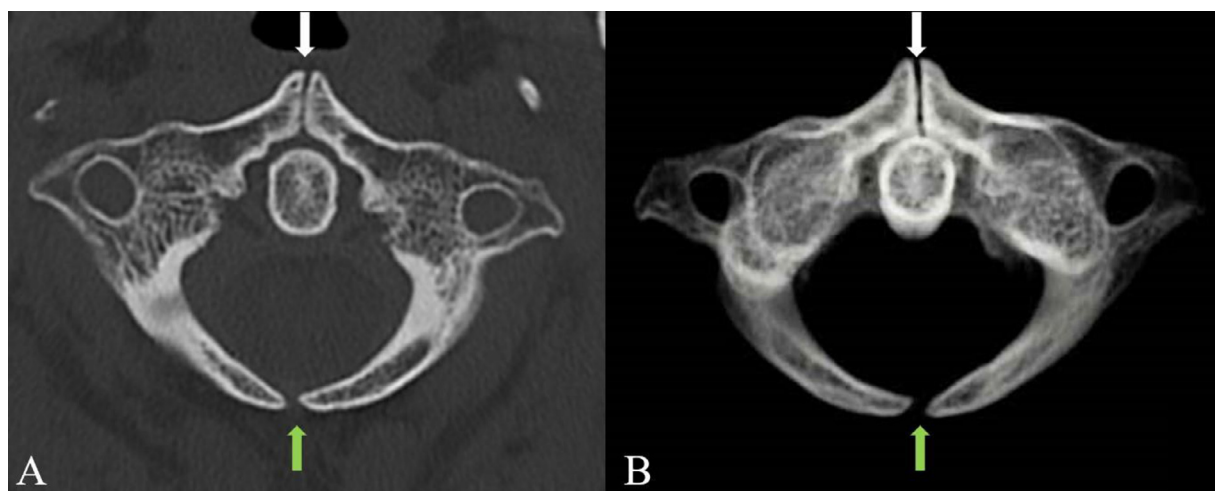


Fig. 2 – Case report 2: Nontrauma patient. Noncontrast computed tomography (NCCT) of head. As incidental findings axial (A) and three-dimensional (B) CT scan showed rachischisis in both anterior (white arrow) and posterior (green arrow) arches with smooth corticated margins and displacement <3 mm of C1 lateral masses. (Color version of figure is available online.)

The aetiology of congenital atlas abnormalities remains unclear. During the embryonic period, 3 primary ossification centres of the atlas usually appear. The earliest, anteriorly, appears during the first year of age. It develops into the anterior arch and tubercle and it usually unite with the 2-lateral ossification centers, showing up during the seventh week of intrauterine life, at 5-9 years of age. The 2-lateral ossification centers extends posteromedially, fusing in the midline and forming the posterior arch. The ossification process is completed between 3 and 8 years of age [3–5]. Anterior atlantal arch defect could be developed in absence of the anterior ossification centers, or whenever 2 lateral ossification centers anteriorly develop, but they do not fuse. Similarly, the lack of fusion of the 2-lateral ossification centers posteriorly, may develop a posterior cleft or hypo/aplasia [6,7].

Posterior defects were classified by Currarino et al. into 5 types: type A, failure of posterior midline fusion of the 2 hemiarches, the most frequent; type B, unilateral cleft in one of the 2 arms or the posterior arch; type C, bilateral cleft with preservation of the most dorsal part of the arch; type D, absence of the posterior arch with persistent posterior tubercle; type E, absence of the entire arch including posterior tubercle [8].

Congenital atlas abnormalities frequently occur with gonadal dysgenesis, Klippel-Feil syndrome, Arnold-Chiari malformation, Turner syndrome, Down syndrome, and thalassemia minor [9–11].

Posterior arch defects may be asymptomatic or associated with neurological symptoms such as weakness in the 4 limbs, paraesthesia in the 4 limbs, in the upper limbs only, or in the ipsilateral upper and lower limbs. In type C and D of Currarino et al. classification (persistent posterior tubercle and bilateral cleft, or absence of the posterior arch [8]) cervical trauma often cause transient quadriplegia, paraparesis, Lhermitte's sign, chronic neck pain, and headache. Type A and D of Currarino et al. classification (failure of posterior midline fusion of the 2 hemiarches) [8], may become symptomatic without exacerbating factors such as atlantoaxial instability, degenerative

change or trauma [3]. Anterior isolate arch defects are rare. Rachischisis in both anterior and posterior atlas arch (“split atlas”) can be associated with asymptomatic lateral atlantoaxial subluxation [12].

Split atlas can mimic a 4-point C1 fracture, known as Jefferson fracture, therefore is extremely important to recognize this congenital abnormality in trauma patient [13]. Computed tomography (CT) scan is the best diagnostic tool for the study of bone tissue. In the first case CT scan showed a small defect in the arch with smooth corticated margins, while sharp edges without cortex suggest a fracture. Lateral translation of C1 lateral masses is >3 mm in Jefferson fracture, whereas is usually <3 mm (1-2 mm) in congenital atlas abnormalities [6]. In addition to Jefferson fracture, at least other four C1 fracture patterns may be described: posterior arch fractures, anterior arch fractures, lateral mass fractures, and comminuted fractures of lateral mass [14]. There are not any reported cases in literature of 2-point midline traumatic fractures. Generally, a congenital defect, unlike a fracture, occurs in the midline position [15,16]. Clinical findings may be similar in both conditions [17].

In conclusion, congenital atlas abnormalities may be incidentally found during the examinations performed after trauma, neck pain, neck mass, radiculopathy or, like in Case report 2, even more casually during CT head scan for treatment-resistant headaches. Arch cleft can mimic fracture. It is essential to differentiate cleft from a fracture to achieve an accurate patient care management. Clinical findings may not help. In Case report 1 rachischisis in both anterior and posterior atlas arches is characterized by smooth corticated margins and displacement <3 mm of C1 lateral masses: these findings suggest split atlas, despite trauma. In Case report 2 we found same radiological features of Case report 1, but it was easier to diagnose split atlas because the patient was a non-trauma patient. CT was extremely helpful in differentiating C1 fracture from C1 congenital anteroposterior cleft, because split atlas is generally difficult to diagnose on plain radio-

graphs. Smooth corticated margins of the cleft, and 1-2 mm (<3 mm) lateral displacement of lateral masses, represent the most important radiological features in the differential diagnosis between congenital atlas abnormalities and fracture.

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