

Case Report

Complete absence of the posterior arch of C1: Case report

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Abstract

Posterior atlas arch anomalies are relatively common, but have a variety of presentations ranging from partial clefts to complete agenesis of the posterior arch. Partial clefts are prevalent in 4% of patients and are generally asymptomatic. However, complete agenesis of the posterior arch is extremely rare. We report the case of a 46-year-old man who presented with upper cervical spine and occipital pain as well as left sided headaches. Imaging revealed congenital complete absence of the posterior arch of C1 (Type E) without any radiographic evidence of instability. We discuss our case in light of other reported cases and detail its management.

Key words: Agenesis, atlantoaxial instability, bracing, C1, posterior atlas arch

INTRODUCTION

Congenital absence of the posterior atlas arch is rare. Cases may appear as partial clefts or even total agenesis of the posterior arch.^[1] They are mostly found incidentally in asymptomatic patients, but symptoms vary from mild neck pain to neurologic deficits after traumatic injury.^[1,2] Partial clefts are prevalent in 4% of patients.^[2-5] In contrast, complete agenesis of the posterior arch is very rare, with reported prevalence of 0.15%.^[2-4] We report the case of a 46-year-old man who presented with upper cervical spine and occipital pain as well as left sided headaches. Imaging revealed congenital complete absence of the posterior arch of C1 (Type E) without any radiographic evidence of instability. We discuss our case in light of other reported cases and detail its management.

CASE REPORT

A 46-year-old man presented to clinic with worsening upper cervical spine pain, occipital pain, and radiating pain from the base of the skull down into the left side of his head. He reported having the pain since childhood, but it has been worsening in frequency and severity. Additionally, with sudden movements he experienced episodes of presyncope. More recently, the patient had been having balance problems and aching and fatigue in both upper extremities. After a period of conservative management with physical therapy, he was referred to our institution for evaluation. On examination, the patient had a motor power of 4/5 in the left deltoids, left biceps, left triceps, and left intrinsic hand muscles, but normal motor power (5/5) in his right upper extremity and bilateral lower extremities. Sensory exam was symmetric, and intact to light touch and pin prick in the bilateral upper and lower extremity dermatomes. Deep tendon reflexes were symmetric and physiologic, and ambulation was normal. There was significant decrease in the range of motion of the neck.

His cervical computer tomography (CT) scan, revealed total absence of the posterior elements of C1 [Figure 1]. Flexion/extension X-rays of the cervical spine similarly revealed absence of the posterior elements of the C1 arch [Figure 2]. Magnetic resonance imaging (MRI) of his cervical spine showed

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absence of the posterior elements of C1 and severe central canal stenosis at C3-4, primarily due posterior disk osteophyte complexes [Figure 3]. There was a focal area of T2/STIR signal hyperintensity in the right cord at this level consistent with myelomalacia. Further evaluation with a flexion-extension MRI and magnetic resonance angiography of the cervical spine revealed no craniocervical junction instability [Figure 4].

Since the patient had myelopathy, the patient underwent a C3-C4 ACDF, but the decision was made to undergo non-surgical management for the posterior C1 arch defect because there was no radiographic evidence of instability. Instead, he was prescribed rigid collar bracing (Miami-J Collar).

At 6 month follow-up, his myelopathic symptoms had resolved, and his headaches have improved. However, when the patient reported that upon removal of the neck collar, he had tremendous lightheadedness and episodes of presyncope. Thus, the patient currently wears the Miami-J Collar most of the time.

DISCUSSION

Posterior atlas arch anomalies are found mostly coincidentally, and are much more frequent than anterior atlas arch

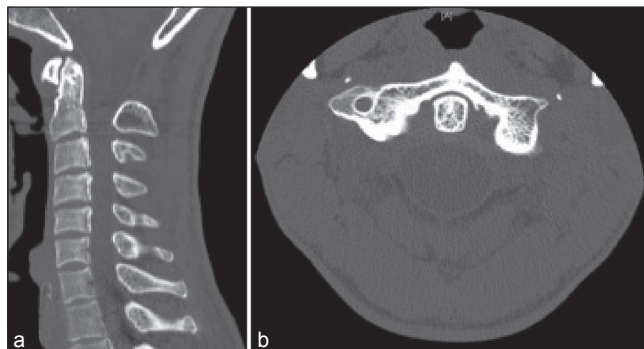


Figure 1: Sagittal (a) and Axial (b) CT scan demonstrating total absence of the posterior arch of C1



Figure 3: Lateral T2 weighted MRI demonstrating total absence of the posterior arch of C1 as well as C3/C4 stenosis as well as signal cord change at that level

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anomalies.^[6] An understanding of embryological development is crucial to understanding congenital atlas arch defects. C1 is derived from the primitive fourth occipital and first cervical sclerotomes. There are three or more primary ossification centers in the embryological period: A midline center that builds the anterior arch in the seventh week of gestation and two lateral ossification centers which form an anterior center for formation of the anterior tubercle and two lateral centers from which the lateral masses and posterior arch form. The pathogenesis of atlas abnormalities is not currently known, but unification between the ossified atlas parts occurs between the ages of five and nine.^[3,7]

According to the classification of Currarino *et. al.*,^[8] there are five types of anomalies. Type A defects are defined as failure of posterior midline fusion of the two hemiarches; Type B, unilateral defects; Type C, bilateral defects; Type D, the absence of the posterior arch with preservation of the posterior tubercle, and Type E, the complete absence of the posterior arch including the tubercle. Based on the literature, roughly 4% of patients present with congenital atlas arch defects, with the predominant type being Type A.^[2-4] In contrast, Type E defects are very rare. One study found two patients (0.18%),^[2] while

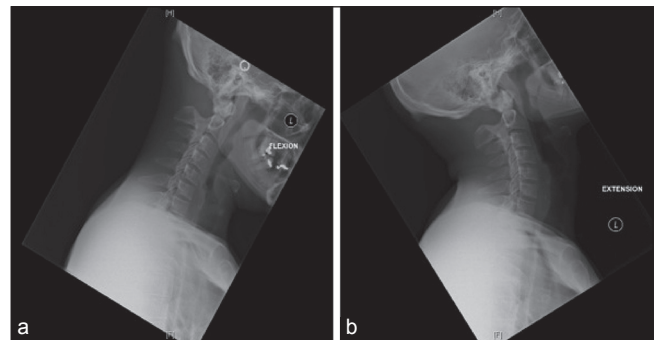


Figure 2: Flexion (a) and Extension (b) lateral cervical spine x-rays demonstrating dynamic instability over upper and subaxial cervical spine

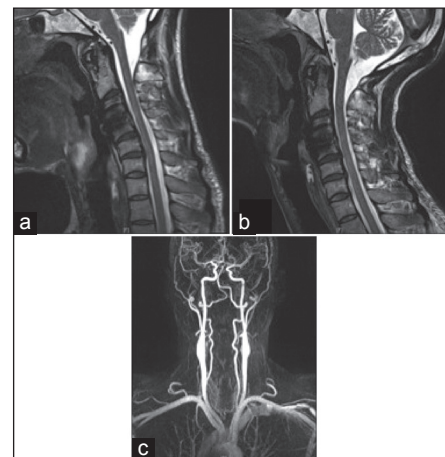


Figure 4: Flexion (a) and extension (b) sagittal T2 weighted MRI showing instability at the craniocervical junction. It also shows postoperative C3/4 anterior cervical fusion and the resolution of the stenosis. MRA of the cervical spine (c) demonstrating normal vasculature

two studies didn't find any patients with type E defects and had patient populations of 1069 and 1153 respectively.^[3,4]

Anomalies of the posterior arch are generally asymptomatic, but there have been case reports of partial aplasia with isolated posterior arch tubercles and C1 canal stenosis causing myelopathy.^[9] There have only been sporadic case reports of patients with Type E posterior arch anomalies. In general, such abnormalities are asymptomatic, but there have been case reports of patients with neck pain and neurological deficits due to structural instability.^[10] To detect instability and determine if management is needed, imaging including dynamic flexion/extension is needed.^[2] In cases of atlantoaxial instability, posterior fusion involving the occiput and lower spinal segments can be performed to immobilize the upper cervical spine.^[1]

Our patient, diagnosed with myelopathy and type E defect of the posterior atlas, responded very well to a C3–C4 ACDF, and had all myelopathic symptoms resolve. However, symptoms due to the congenital absence of his posterior atlas remain, but resolve when stabilized with a Miami J neck collar. Since there was no evidence of atlantoaxial, or occipitocervical instability, the decision was made not to undergo surgical management given the high morbidity of an occipitalcervical fusion. The patient's only current complaints are that the symptoms return when his neck collar is removed, but he has no other symptoms.

Our findings are different than previous reports in the literature. There have been very few recorded cases of Type E posterior arch anomalies, and to the best of our knowledge, there is no other case report detailing the symptoms described here. Imaging remains the first option for investigation, but surgery should only be reserved for cases of atlantoaxial instability. We propose that for symptomatic patients without any radiographic

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evidence of instability, a non-operative treatment plan is initiated before surgical intervention. In this case, a Miami J collar was used with success.

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