Congenital Absence of the Posterior Arch of the Atlas – A Strange but Benign Anomaly

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ABSTRACT

Congenital partial aplasia of the atlas with a posterior arch remnant is rare. It may be found as an incidental radiological finding or patients can present with neurological signs and symptoms after head or neck trauma. A 36-year-old female presented with a 3-day history of right sided neck pain radiating down the right arm. Radiographs of the cervical spine showed a radiolucent area in the region of the posterior arch of the atlas. Computed tomography subsequently revealed partial absence of the posterior arch of the atlas with a defect at the anterior ring. Her clinical condition subsequently improved with rest, analgesia and physiotherapy. This case report illustrates a situation where congenital defect of the posterior arch of the atlas was revealed in a patient with neck pain. Awareness of the existence of this condition will help clinicians avoid misdiagnosis and excessive investigations.

Key Words:

Posterior arch of the Atlas, Congenital absence, Posterior tubercle

INTRODUCTION

Congenital partial aplasia of, or clefts in the arch of the atlas are rare abnormalities. Presenting clinical features vary from mild neck pain to neurological deficits occurring after minor cervical spine or head injury¹. A patient who suffered mild neck pain was found to have a radiolucent area in the region of the posterior arch of atlas. Further investigation showed partial absence of the posterior arch of the atlas. We are reporting this case in order to enhance awareness of the anomaly so that misdiagnosis as a spinal lytic lesion can be avoided.

CASE REPORT

A 36-year-old female presented with a 3 day history of right sided neck pain radiating down the right arm. The patient

had no history of trauma and denied any weakness or numbness. Physical examination was negative for any neurological signs. Neck movement was within normal limits.

Radiograph of the cervical spine showed radiolucency in the posterior arch of the atlas (Fig. 1). Computed tomography demonstrated partial absence of posterior arch of atlas with a gap in the anterior ring (Fig. 2). The patient was treated with a cervical collar and analgesics and her neck pain subsided after 2 weeks. She remained asymptomatic when she was last reviewed at six month.

DISCUSSION

Congenital defects of the atlas are rare¹. Embryologically, the atlas is formed from three ossification centers during the first seven weeks of gestation. Two centers from the lateral masses extend posteromedially to form the posterior arch. The ossification of the anterior arch involves one or two ossification centers that extend posterolaterally to fuse with the lateral masses. A fourth ossification center exists in about 2% of fetuses that forms the posterior tubercle. Failure of fusion or absence of this particular fourth ossification center leads to clefts or aplasia of the atlas².

Curriano et al, have classified this congenital defect into five categories: Type A, failure of posterior midline fusion with a small gap remaining; Type B, unilateral cleft, Type C, bilateral defects with preservation of the most dorsal part of the arch, Type D, complete absence of the posterior arch with persistent isolated tubercle, and Type E, complete absence of the posterior arch including the tubercle³. Our patient was found to have a type C defect. Type A defects have been reported in 4% of the population and account for over 90% of all posterior arch aplasias. It is estimated that 0.69% of the population have type B, C, D and E defects³. Cervical spine radiography with flexion/extension views may help to detect cases with mechanical instability where there is increased risk of spinal cord injury.

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Fig. 1: Lateral radiograph of cervical spine reveals partial absence of the posterior arch of C1 vertebra and isolated posterior tubercle.

Patients with posterior arch defects typically present in one of five ways: asymptomatic with the anomaly discovered as a result of radiolucency imaging undertaken because of unrelated indications; patients with neck pain after trauma who undergo imaging; patients found to have neurological deficits secondary to trauma; patients with neurological symptoms prior to diagnosis; and patients who present with and are investigated for chronic neck pain³. Congenital partial aplasia of the atlas with posterior tubercle can be managed conservatively or by surgical resection. Patients with this anomaly should be advised to avoid contact sports and seek medical treatment should they experience

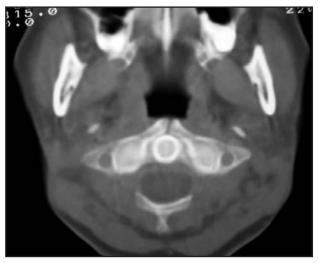


Fig. 2: Axial CT image of C1 vertebra shows agenesis of posterior arch and cleft at the anterior arch.

neurological symptoms³. Presence of a posterior tubercle remnant in Type C and D potentially causes transient quadriparaesis. Surgical removal of the tubercle and ligament between C1 and C2 is preferred in these groups if there is no evidence of atlanto-axial instability⁴.

Congenital partial aplasia of the atlas with posterior tubercle is a rare anomaly and is a 'benign' condition. It should be considered when there is a defect over the posterior element of atlas. Consulting a radiologist could help achieve diagnosis and avoid unnecessary diagnostic and therapeutic procedures.

REFERENCES

- 1. Sharma A, Gaikwad SB, Deol PS. Partial aplasia of the posterior arch of the atlas with an isolated posterior arch remnant: findings in three cases. *AJNR Am J Neuroradiol.* 2000; 21:1167-1171.
- Atasoy C, Fitoz S, Karan B. A rare cause of cervical spinal stenosis: posterior arch hypoplasia in bipartite atlas. *Neuroradiology*. 2002; 44:253-255.
- 3. Curriano G, Rollings N, Diehl JT. Congenital defects of the posterior arch of the atlas: a report of seven cases including an affected mother and son. *AJRN Am J Neuroradiol*. 1994; 15: 249-54.
- 4. Klimo P, Blumenthal DT, Couldwell WT. Congenital partial aplasia of posterior arch of the atlas causing myelopathy: Case report and review of the literature. *Spine*. 2003; 28: 224-8.
- 5. Garg A, Gaikwad SB, Gunta B, Mishra MK. Bipartite atlas with os odontoideum. Spine. 2004; 29: 35-8.