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Posterior Hypoplastic C1 Arch with Neurological Deficits – A Case Report

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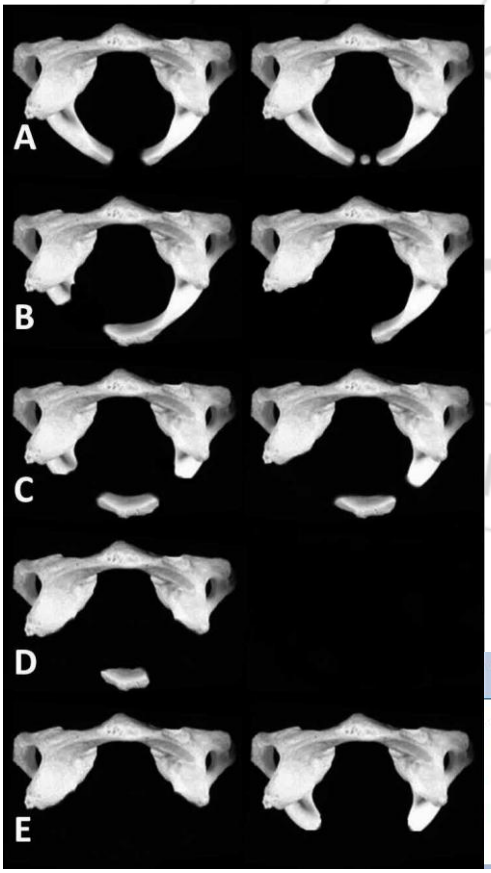
Abstract: ***Hypoplastic C1 posterior arch, a rare anomaly ranges from partial cleft to total agenesis which is a result of developmental failure of chondrogenesis, which is always missed as a diagnosis until only when symptomatic and is usually surgically treated. However, preventing cumulative trauma to the cord by surgery at early stage is debatable. We present a case of 36 years female patient with paraesthesia in all four limbs and weakness of right hand grip and triceps. Patient was investigated with dynamic X ray of CVJ, CT scan with 3D reconstruction and MRI. Posterior hypoplastic arch excision was done without a need for stabilisation as there was no segmental instability.***

Keywords: hypoplastic posterior C1 arch

1. Introduction

Congenital hypoplastic c1 posterior arch is a relatively rare with structural defect in posterior arch of atlas which ranges from partial cleft to total agenesis. It is classified into five types by Currarino. Hypoplastic posterior arch of atlas is due to developmental failure of chondrogenesis[1], which is always missed as a diagnosis as it is usually asymptomatic.It is diagnosed only when symptomatic and is usually surgically treated. It is usually associated with neurological deficits and segmental instability at C1-C2. It may be associated with Arnold-chiari malformation, Klippel feil syndrome[4], Down's and Turner's syndrome. When asymptomatic, it is radiologically diagnosed. Embryologically, C1 is formed from three ossification centers. Posterior arch and lateral masses are developed from two lateral ossification centers.



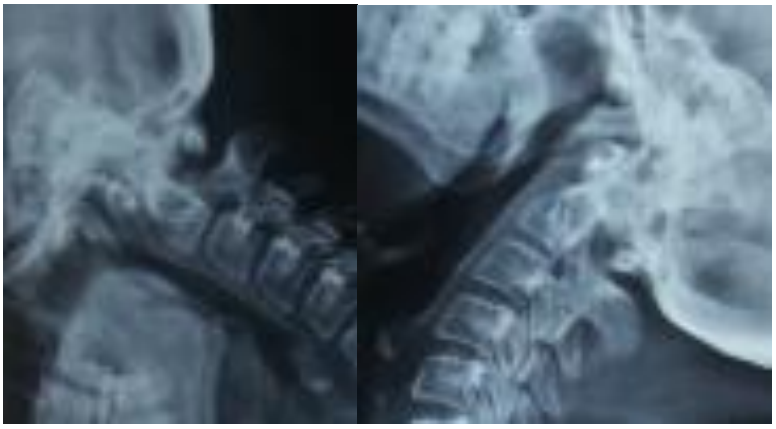
Type	Description
A	Failure of posterior midline fusion of the two hemiarches
B	Unilateral defect
C	Bilateral defects
D	Absence of posterior arch, with persistent posterior tubercle
E	Absence of the entire arch, including the tubercle

Reference: Congenital Hypoplasia of the Posterior Arch of the Atlas, Turkish Neurosurgery 2011, Vol: 21, No: 1, 97-103

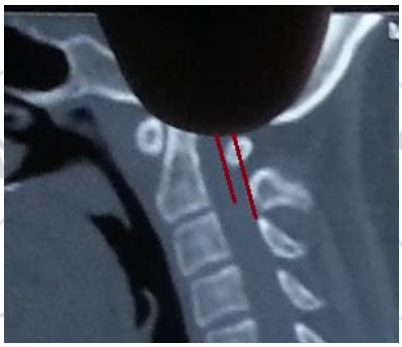
2. Case Report

A 36 years old female patient presented with history of chronic headache, neck pain and giddiness since two months and associated with paraesthesia in all four limbs since 1 month. On examination, patient had right hand grip and triceps weakness with significant decrease in activity of daily living. She had no bowel and bladder incontinence and no posterior column signs with normal deep tendon reflexes and plantars are equivocal on both sides. Patient was

investigated with X ray, CT scan with 3D reconstruction and MRI. Radiologically the patient was diagnosed to have a hypoplastic posterior C1 arch without atlanto axial instability.

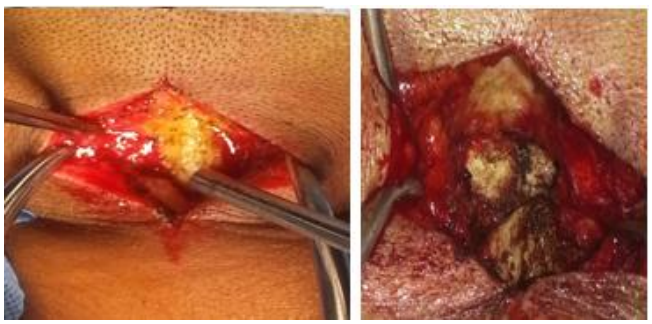


X ray (dynamic views) shows no segmental instability (AAD)



CT scan with 3D reconstruction shows narrowed canal diameter and hypoplastic posterior arch of atlas

After clinic radiological evaluation of the patient, she was planned for surgical excision of posterior C1 arch. Under general anaesthesia, posterior C1 arch was removed and significant decompression of the cord was achieved. However, there was no need for stabilisation as there was no atlanto axial instability (which was confirmed on dynamic x ray).



1. Midline exposure via ligamentum nuchae
2. Posterior arch of C1 exposed
3. Posterior C1 arch excision

Exposure of C1 posterior arch was limited to 1.5cm on both sides to prevent vertebral artery injury. Patient was discharged on 2nd postoperative day and was reviewed on 10th postoperative day and after 1 month. Patients neurological symptoms improved.

3. Discussion

Congenital hypoplastic posterior C1 arch is a rare condition where the diagnosis is most of the time missed as the patient is asymptomatic most of the time and is an incidental finding till the patient is symptomatic. The patient usually presents after a trivial trauma with neurological symptoms. Unless the medical personal has a thorough knowledge, the radiology is always misleading. However, clinical symptoms are highly variable from asymptomatic(most of the times) to tetraparesis. Patients falling under group C and D are usually symptomatic. In our case, unlike the Currarino's classification, the posterior arch was complete with significantly narrowed canal diameter[3]. She was never diagnosed even after she had neurological symptoms and she was on anti-depression drugs. After the relevant clinicoradiological examinations, the patient was diagnosed to have hypoplastic posterior arch of atlas. Her dynamic x ray of CVJ(cranio vertebral junction) showed no instability(atlanto axial dislocation)[2]. After excision of posterior hypoplastic arch of atlas, patient symptoms

improved. The treatment modalities of patients with hypoplastic posterior C1 arch, includes no treatment or follow up for asymptomatic cases. In asymptomatic patients of subgroup C and D, they are advised not to go for contact sports. Prophylactic surgery for such cases are debatable. However in symptomatic cases, surgery is the treatment of choice.

4. Conclusion

Congenital hypoplastic posterior C1 though rare, should always be suspected in a patient with neurological symptoms post trivial trauma with no obvious cervical spine fractures. Most of the patients though asymptomatic, needs surgical decompression when symptomatic and a stabilisation along with decompression in cases with segmental instability (AAD)[2].

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