

Proatlas Segmentation Anomalies: Clinical Presentation, Radiological Features & Surgical Management Natarajan Muthukumar MCh FACS FICS FAANS Department of Neurosurgery, Devadoss Multispeciality Hospital, Madurai, India

Introduction

Developmental abnormalities of the craniovertebral junction (CVJ) due to defective segmentation of the Proatlas sclerotome are known as Proatlas Segmentation anomalies (PSA). In this report, clinical & radiological findings and management of nine patients with PSA are presented.

Methods

Between Jan 2012 and June 2016, nine patients with PSA were encountered. Plain radiographs, thin section CT & MRI were obtained for evaluation. Surgical management varied according to the nature of the underlying pathology.



Ventral brainstem compression with basilar invagination

Results

Four patients had cervical myelopathy. One had myelopathy with cerebellar signs, three had only neck pain & vertigo and in one patient the diagnosis was made during radiological examination after trauma. Radiologically, one patient had assimilation of anterior arch of atlas, platybasia, partial assimilation of posterior arch, basilar invagination and an accessory ossicle behind the flat clivus. The second patient had a prebasicoccipital arch, basilar invagination, completely assimilated atlas, unilateral occipital condyle hypoplasia and Klippel-Feil anomaly. The third patient had Os Avis or dystopic os odontoideum. The fourth patient had pre-basioccipital arch and atlanto-axial subluxation. The fifth patient had severe platybasia, retroflexed odontoid compressing the cervicomedullary junction, tonsillar ectopia up to C 2 and cervical syrinx; patients six and seven had retroflexed odontoid with lateral proatlas failure, patients eight and nine had prebasioccipital arch. Six of the nine patients underwent surgery. Three patients underwent craniovertebral realignment and occipitocervical fusion. Two patients underwent Goel-Harms fusion and one patient underwent Goel-Harms fusion with distraction using spacers

in the atlantoaxial joint and foramen



Horizontally oriented clivus, basilar invaginatiion with accessory ossicle behind the clivus.

Figure 3



Pre & Postoperative Sagittal CT showing the descent of the odontoid with increased O- C2 distance

Conclusions

PSA is a rare cause of CVJ compression and/or instability. Careful CT evaluation is the prerequisite for proper diagnosis. The management of PSA is aimed at: 1. Decompressing the cervicomedullary junction if there is compression and, 2. Stabilization of the craniovertebral junction, if there is instability.

Learning Objectives

"By the conclusion of this session, participants should be able to : 1. know the diverse radiological & clinical presentations of proatlas segmentation anomalies and, 2. know the treatment modalities for various Proatlas segmentation defects.

References

1.Menezes AH, Fenoy KA : Remnants of occipital vertebra: Proatlas Segmentation Anomalies. Neurosurgery 64: 945 -954;2009

2.Pang D, Thompson DNP: Embryology and bony malformations of the CVJ. Childs Nerv Syst 27:523–564; 2011.



Coronal CT showing unilateral occipital condyle hypoplasia with absence of lateral mass of atlas on one side with C 2 articulating directly with hypoplastic occipital condyle