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Introduction

This retrospective study analyzed 46 patients with cranio-vertebral junction anomalies and hindbrain herniation and or syringomyelia. The presence of syringomyelia is well known however the presence of bony cranio-vertebral junction anomalies is much rare, with hind brain herniation.

Methods

Forty six patients with bony CVJunction anomalies with hind brain herniation with or without syringomyelia who were operated at AIIMS Delhi between 2005 and 2010 were analyzed retrospectively. History and clinical examinations, and radiology studies including X rays, Ct scans and MRI were studied from charts and old records. The operation notes and follow-up charts were also studied.

Results

Forty- six patients were studied. Mean age was 27 years(range 3-52 years). The duration of symptoms ranged from 3 months to 3 years(mean 21 months). Sub-occipital headaches and neck pain were the commonest symptoms. More than 36 % had motor weakness. patients were categorized as Group I - in whom the bony anomaly was reducible, Group II where the anomaly was irreducible and Group III patients with syndromic musculo-ligamentous instability like MPS. The most common anomaly was Atlanto-axial dislocation (AAD) seen in 95%, 65% of these were irreducible. More than 65 % had basilar invagination . Chiari was seen in all and syringomyelia in 58%. Eight percent had syndromic abnormality like MPS, Downs etc. All patients underwent posterior fixations, 96% also had posterior decompression, and in 63% ventral decompression was done in addition to posterior procedures. CSF leak occurred in 6 patients , all of them underwent intradural procedures. Eight % patients needed prolonged ventilatory support and were also tracheostomised. Patients treated with ventral and dorsal decompression along with posterior fixation had better clinical outcomes in comparison to patients treated only with dorsal decompression and fixation(82% versus 77% at 6 months, n= 34 and 86% vs. 72% at 1 year, n= 22), on follow-up, respectively.

Conclusions

Patients with developmental bony CV junction anomalies and chiari malformation with or without syringomyelia require anterior decompression if ventral compression is present. These patients also require posterior decompression, if posterior compression is present.

Learning Objectives

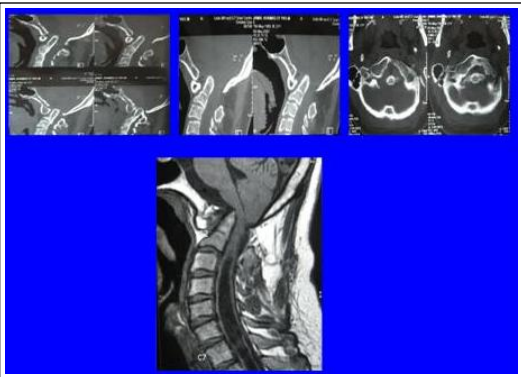
By the conclusion of this session, participants should be able to identify this less common but complex spinal problem. Should be able to discuss the management principles, and the complications encountered. Also the participants should be able to identify that some rare syndromic diseases like Down's and MPS may be associated with these clinical conditions.

References

1. Milhorat et al Neurosurgery 1999.
2. Tubbs et al JNS 2003.
3. Goel et al BJNS 1995.

Goel et al JNS 1998

Menezes et al Pediatric JNS 2005.



surgical lesions

Surgical Lesions

- Bony & soft tissue lesion
- Mobility at the Cranio-vertebral Junction
- Syringomyelia

Inclusion criteria

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1. Chiari I/Hind Brain Herniation:
Cerebellar tonsillar descent below the level of Foramen magnum.
2. Syringomyelia
3. Bony anomaly – OA, Fused C2 C3, BI, AAD

1 or 2 and 3

Meterail & Method

Material & Method

- All patients with both bony and soft tissue CVJ anomalies
- Operated between March 2005 and February 2010
- Evaluation of case records, discharge records, follow-up visit records
- Follow-up between 3 months and 5 years