

Oral presentation

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Symptomatic cerebellar tonsillar ectopia in lipomyelomeningocele patients

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Background

To study the incidence of cerebellar tonsillar ectopia and Chiari type symptoms in a population of LMM patients.

Materials and methods

The records of patients with a diagnosis of LMM between the years of 1978 and 2003 (130 patients) at Children's Hospital and Regional Medical Center (CHRMC) in Seattle, Washington were reviewed. The presence and degree of tonsillar ectopia was determined by reviewing total spine and brain MRIs (sagittal T1 sequences) from these patients. We defined significant ectopia as greater than 5 mm below the foramen magnum. We limited the study to the 46 out of 130 patients who had previous MRIs. Clinic notes from yearly clinic visits were also reviewed in order to determine the presence of symptoms such as headaches, shoulder pain and bulbar symptoms. We used Fisher's exact test of proportions.

Results

We identified cerebellar tonsillar ectopia greater than 6 mm in 14/46 (30%) of the patients. The degree of tonsillar ectopia ranged from 6 to 13 mm below the foramen magnum. Review of the medical records of all 46 patients which included a yearly clinic visit until the age of 21, revealed that 6/14 patients with significant tonsillar ectopia had frequent headaches associated with cough or valsalva, while only 1/32 patients without significant ectopia had headaches ($P = 0.002$). Patients with significant ectopia were not more likely to have multiple detethering operations (4/14 (29%) in the ectopia subgroup versus 5/32 (16%) in patients without significant ectopia) ($P =$

0.3). There was no correlation between the number of detethering operations and degree of tonsillar ectopia within each group. Also, no correlation between the age of the patient at initial repair and the degree of tonsillar ectopia was observed.

Conclusion

This study has revealed a 42-fold increase in the incidence of pathologic cerebellar tonsillar ectopia in LMM patients in comparison with historical controls. In addition we have identified symptoms suggestive of Chiari type I malformation in greater than half of the patients within this subgroup.