ABNORMAL RHOMBOID LIP IN CHILDREN WITH COCHLEAR NERVE DEFICIENCY SUBMITTED TO ABI

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ABSTRACT

Object.

To investigate the prevalence and implications of severe abnormalities of the rhomboid lip (RL) that make auditory brainstem implantation (ABI) surgery in children with cochlear nerve deficiency (CND) a difficult and risky procedure.

Methods.

In a cohort of 78 consecutive children with CND implanted with ABIs surgical video recordings and a full sequence of snapshots of the area surrounding the foramen of Luschka (FL) undergoing surgery were available for the present study. The individual surgical videos of each child were reviewed and analytically examined by four independent individuals to define the types and frequency of abnormalities of the RL.

Results.

The membranous structure forming the RL varied greatly in size, thickness and transparency in children of the present cohort. In a large number (72%) of children the RL extended laterally over the lower cranial nerves, obstructing the view of the FL and necessitating section of the RL to access the lateral recess (LR) and implant the ABI on the cochlear nuclei.

Conclusions.

An abnormal large RLs adhering to the IX and X and obstructing the FL is described for the first time in children with CND during ABI surgery. A surgical dissection of the arachnoid membranes from the foramen jugularis down to cranial nerves VII, IX and X and delicate detachment of the RL from these nerves is necessary to visualize the FL, access the LR and locate the ABI array on the cochlear nuclei.

INTRODUCTION

Pediatric auditory brain stem implantation (Pedi ABI) recently emerged for deaf children unsuitable to cochlear implant (CI). For fitting the ABI in children we have been using and recommending the modified retrosigmoid (RS) approach, technically similar to the lateral infrafloccular approach.

After exposing the cerebellopontine angle the arachnoid membranes around the jugular foramen are resected down to the level of the root of cranial nerves (cns) VII, IX and X . To avoid stretching these cns and visualize the foramen of Luschka (FL) the flocculus (F) is gently retracted in a caudo-rostral direction (6)

Occasionally during this step, a thick and large membranous structure, distinct from the arachnoid membrane may be encountered. When this membrane is stuck to cns IX and X, the view as well as the entrance of the FL is inhibited. This large and thick membrane is indeed a rhomboid lip (RL) of abnormal size. The RL is defined as a sheet-like layer of neural tissue that forms the ventral wall of the lateral recess (LR) of the fourth ventricle and part of the FL. Few cases of large RLs adhering to the lower cns have been described in the neurotologic literature during microvascular decompression (MVD) surgery of lower cranial nerves in adults. In these patients, to avoiding unexpected complications, such as hearing loss, hoarseness or dysphagia after MVD procedures, it is recommended to identify the abnormal RL before exposure of the root exit zone of cns VII , IX. To the best of our knowledge this abnormality of the RL has never been described during ABI surgery in adults or children and the aim of the present paper is to illustrate the prevalence and implication of severe abnormalities of the RL that make ABI surgery a rather difficult procedure in the cohort of children with CND.

METHODS

In a cohort of 78 consecutive children with CND implanted with ABIs surgical video recordings and a full sequence of snapshots of the area surrounding the foramen of Luschka (FL) undergoing surgery were available for the present study. The individual surgical videos of each child were reviewed and analytically examined by four independent individuals to define the types and frequency of abnormalities of the RL.

RESULTS

The membranous structure forming the RL varied greatly in size, thickness and transparency in children of the present cohort. In a large number (72%) of children the RL extended laterally over the lower cranial nerves, obstructing the view of the FL and necessitating section of the RL to access the lateral recess (LR) and implant the ABI on the cochlear nuclei.

During ABI surgery in adults the RL can be easily recognized forming the ventral wall of the lateral recess and part of the FL. In children with CND the RL is highly variable in size, thickness and transparency. It is much thicker, of tougher consistency and less transparent compared to the arachnoid membranes normally covering the entrance of the FL. Often at exposure of the area of the FL, a tough protruding cyst of different size and consistency is frequently encountered that, obliterating the entrance of the FL, blocks the view of the CP. In few cases when the RL is very large and bulging it may be mistaken for an arachnoid cyst. In other cases the RL may reach the jugular foramen and simulate a normal arachnoid membrane. When these abnormalities of the RL are encountered in children the identification of the point where to start dissection, open the FL and insert the ABI array may became quite difficult.

CONCLUSIONS

The RL is indeed in an important component of the FL that extends laterally from the floor of the IV ventricle and, together with the tela choroidea, forms a pouch at the outer extremity of the LR. The present study demonstrates that, in children with CNS, the abnormalities of the RL are extremely frequent probably as a result of a more complex malformation of the area that involves cn VIII, typically aplastic, occasionally cn VII and more frequently then known the constituents of the FL.

An abnormal RL tightly adherent to the cns IX and X and obliterating the entrance to the FL is a new finding observed during ABI surgery in children with CND. The obliteration of the area of the FL presents the aspects of a cist of different morphology, size and structure making identification of the FL an arduous task. This anatomical malformation imposes a meticulous and delicate separation of the RL from the lower cns, followed by an incision of the membrane of the cist, identification of the CP inside the FL, visualization of the LR and gently insertion of the ABI array on the CN.

This report sheds light on the clinical importance of the RL during ABI surgery in children with CND. An advanced anatomical knowledge of the microanatomy RL and the adjacent area may facilitate safer exposure of the FL and improved access to the LR and CN. Further studies on the anatomical variations of the RL in its size, transparency, thickness and relationship to the cns IX,X and XI are needed to refine our knowledge on the importance of the RL for ABI.

Figure 1. The large and thick membrane is obscuring the view of the FL is indeed a rhomboid lip of abnormal size.

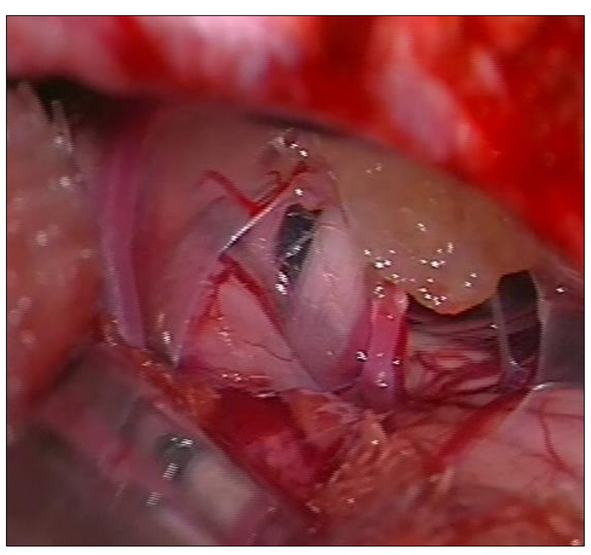

Figure 2. A large RL is adhering to the lower cns.

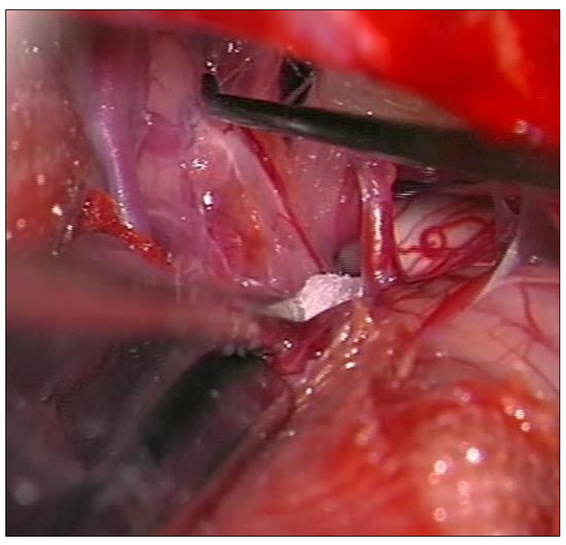

Figure 3. To identify the CP the membrane occluding the FL is sectioned.

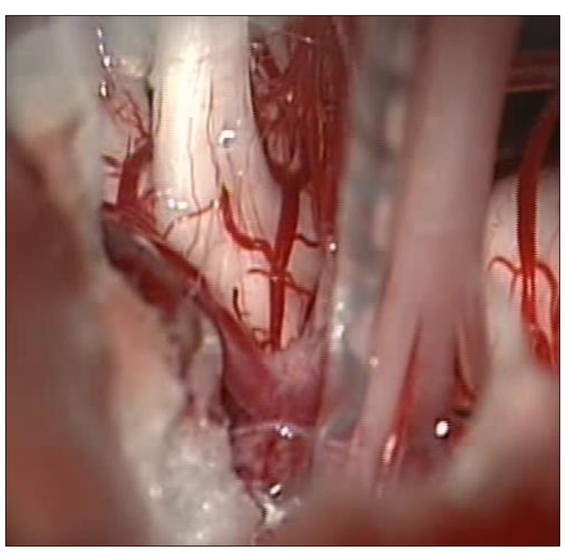

Figure 4. The concave space shown in the supraolivary fossette between the cns VII and IX mimics but is not the FL . The ABI array is inserted inside the LR on the cochlear nuclei complex.

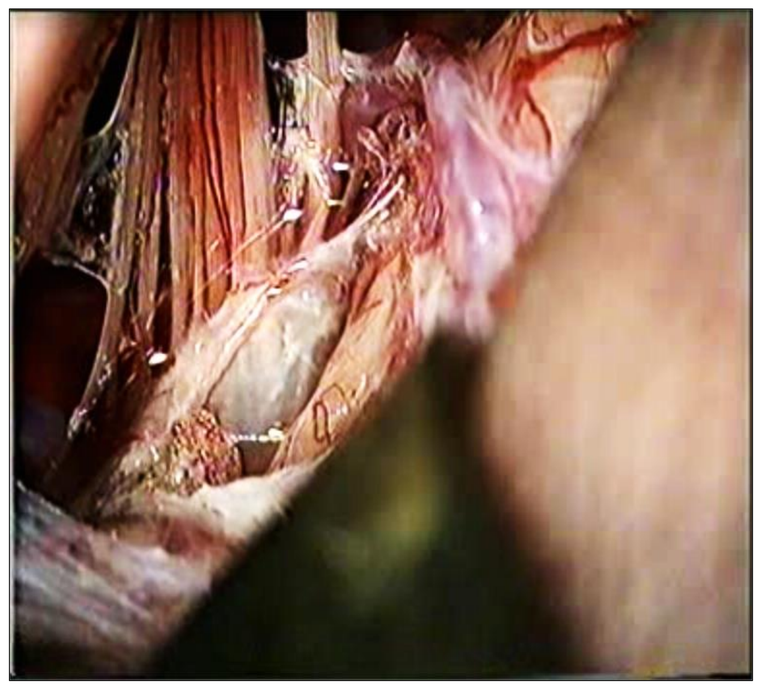

Figure 5. The RL in adults can be generally easily recognized forming the ventral wall of the lateral recess and part of the FL .

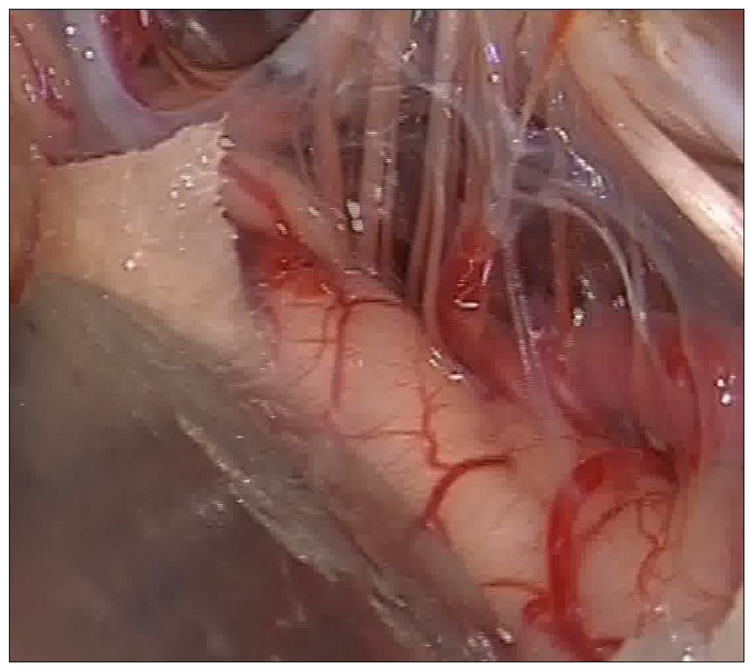

Figure 6: Often at exposure of the area of the FL, A tough protruding cyst large size and consistency is is shown to obliterate the entrance of the FL, blocking the view of the CP.

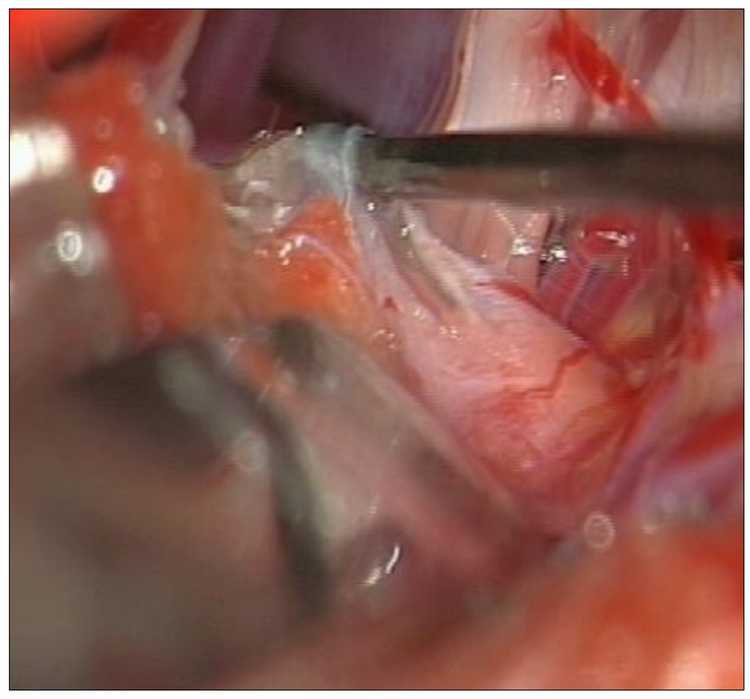

Figure 7: The abnormal RL shown in figure 6 is gently dissected parallel to the long axis of the RL allowing to proceed with the subsequent surgical steps.

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