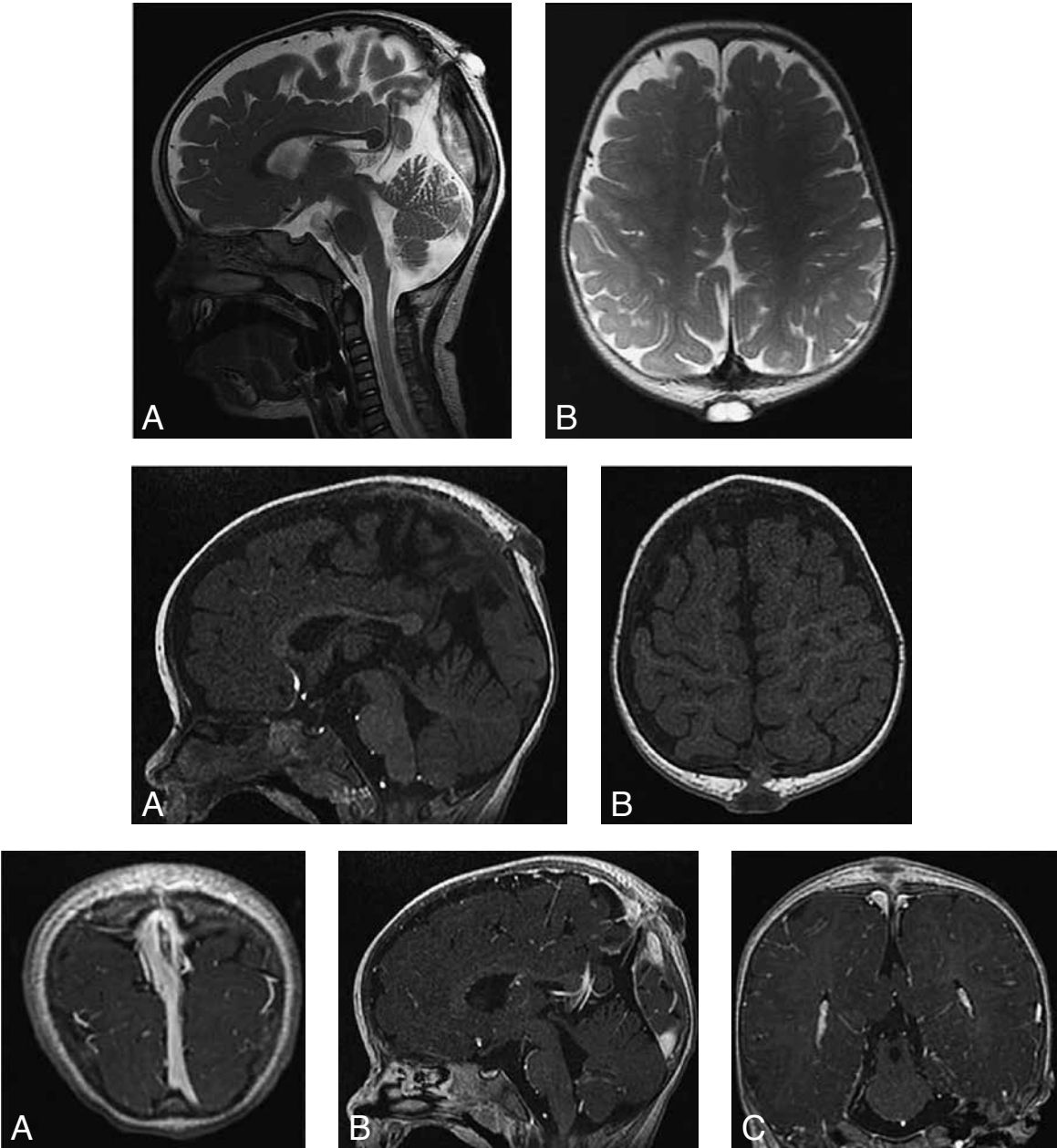


ATRETIC CEPHALOCELE

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Key-word: Brain, hernia

Background: A 1-year-old girl was referred to the neurosurgery department. The patient presented with a swelling on the vertex of the head which was present at birth but increased gradually over time. There were no perinatal problems and the child developed normally. The swelling produced no discomfort nor pain, and measured 2 cm × 3 cm. The child was followed during 6 months because there was a chance of involution. The patient was reevaluated 6 months later.



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Fig.

1A	1B
2A	2B
3A	3B 3C

Work-up

MRI of the skull, T2-weighted images (Fig. 1) shows on sagittal section (A) and axial section (b) a fluid filled parietal subgaleal lesion in connection with the intracerebral structures is seen. A lifted tentorium was noticed.

On unenhanced 3D T1-weighted images (Fig. 2, A: sagittal section, B: axial section), the lesion described in Fig. 1A has a low signal intensity.

On contrast-enhanced T1-weighted images (Fig. 3: A sagittal section, B: axial section, C: coronal section) a vertical position of the internal cerebral vein and close connection with the venous structures are shown. There is a clear defect through the superior sagittal sinus.

Radiological diagnosis

Based on these imaging findings and the pathological confirmation, the presence of *atretic cephalocele* was diagnosed.

Surgical excision with coagulation of the subcutaneous lesion was performed. The bony defect on the midline of the scalp was closed. During surgery no CSF leakage was noticed.

Histological examination showed fibrous tissue with a cavity without cyst wall. The tissue contained little but prominent vascular structures and some degree of inflammation.

Discussion

Atretic cephalocele was first described by James and Lassmann in 1972, and is an abortive form of a cephalocele. These defects are caused by failure of the neural tube to close completely during fetal development. Sometimes it is associated with an intracranial venous anomaly. The atretic cephalocele is a parenchymal or cystic mass attached to the dura mater by a connective tissue strand. Mostly it

is composed of fibrous tissue with sporadic presence of meninges, glia or neurons. The location of the atretic cephalocele is related to additional intracranial anomalies and is therefore also related to the clinical outcome.

Occipital cephaloceles have lower prevalence of associated anomalies and these patients have a better chance for normal development. Parietal cephaloceles comprise 10% of all cephaloceles. They are associated with callosal agenesis, prosencephaly, Chiari type II malformation, Walker-Warburg syndrome and Dandy-Walker malformation. In parietal cephaloceles, there is sometimes marked cerebral dysplasia with dorsal cyst formation.

Surgical excision is advisable, though surgery for intracranial venous anomalies is not indicated because these veins participate in normal venous drainage.

MR imaging is very helpful in the diagnosis, because it can reveal many associated underlying malformations. As the prognosis is related with the associated anomalies, it is very important to conduct a thorough MRI investigation.

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