Klinikum rechts der Isar

Technische Universität München

Intracranial neurenteric cysts – rare incidental findings with typical MRI characteristics

S. Prothmann*, R. Trampel, D. Fritzsch***, J.P. Schneider******

*University hospital rechts der Isar, Department of Neuroradiology, Munich
**Max Planck Institute for Human Cognitive and Brain Sciences, Department of Neurophysics, Leipzig
***Leipzig university hospital, Department of Neuroradiology, Leipzig
****Radiology group practice am Diakonissenhaus, Leipzig

Background

Neurenteric cysts (NECs) are manifestations of rare congenital dysplasia probably resulting from a separation failure of the notochord and the upper gastrointestinal tract and are therefore occasionally found as space-occupying lesions in the posterior mediastinum [1-4]. Most likely, they are of endodermal origin and are lined with ciliated to nonciliated, low cuboidal to columnar epithelium and contain a few mucin-secreting goblet cells. The cysts mostly content a clear fluid of varying viscosity or protein content [5].

Most frequently they are found in the lower cervical and upper thoracic spinal canal. Intracranial occurrences are very rare. Previously reported intracranial NECs were mostly located near the midline in the posterior fossa and, due to their size, almost always required therapy. In a few cases, supratentorial occurrences have also been reported [5,6]. We report on three patients with intracranial neurenteric cysts, two of which were incidental findings and in one case presented with only very mild symptoms.

Case 1

A 24-year-old female patient presented with a left-sided hemifacial spasm, which had first occurred one year earlier and had been progressing slowly over the last three months. An MRI study revealed a 1.2 x 0.9 cm cystic space-occupying lesion in the left pontocerebellar angle with contact to the facial and vestibulocochlear nerve. The cyst was smoothly delineated. T1-weighted imaging (T1-WI) showed a moderately hyperintense signal compared to CSF, in the T2-weighted imaging (T2-WI) and the FLAIR sequence, the lesion was distinctly hyperintense as compared to CSF. In the CISS sequence the cyst was slightly hypointense to CSF, but still hyperintense in comparison to brain tissue. No enhancement was found. No restriction was seen in diffusion imaging. The patient declined surgical therapy. A check-up performed after one year revealed no change in the findings (Fig 1-a, 1-b, 1-c, 1-e).

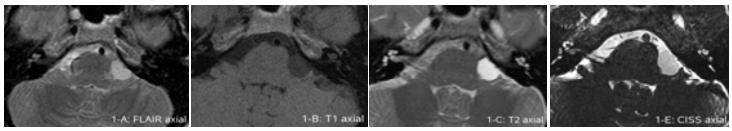

Fig 1:
Approximately 1.2 x 0.9 cm extra-axial cystic lesion in the left pontocerebellar angle. Distinctly hyperintense signal compared to CSF and brain tissue in the FLAIR (A, axial) and in T2WI (C, axial), slightly hyperintense to CSF in T1WI (B, axial). In the CISS sequence (E, axial) in comparison to CSF slightly hypointense, but still hyperintense signal compared to the surrounding brain tissue.

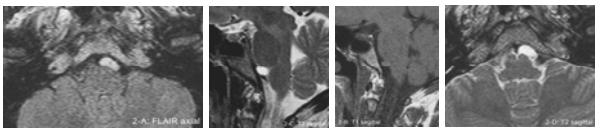

Fig 2:
Cystic, extra-axial, approximately 1.2 x 0.5 cm ovoid lesion at the pontomedullary junction, to the left of the midline. Distinctly hyperintense signal in T2WI (C, sagittal; D, axial) and the FLAIR sequence (A, axial). Moderately hyperintense in T1WI (B, sagittal) compared to CSF.

Case 2

A 46-year-old woman presented with a condition following left-sided retrobulbar neuritis, which had regressed completely under therapy with methylprednisolone. Neither CSF analysis nor brain MR imaging provided any indication of chronic inflammatory CNS disease. As an incidental finding, however, anterior to the brain stem left of the midline at the pontomedullary junction, we discovered an extra-axial, smoothly delineated cystoid structure with an ovoid shape measuring 1.2 x 0.5 cm. In the T2-weighted sequences (including FLAIR), the cyst showed a distinctly hyperintense signal in comparison to CSF. In T1-WI, a moderately hyperintense signal was found.
We found no restriction in DWI and no enhancement after administration of gadolinium DTPA. Since no symptoms attributable to the cyst were observed, there were no therapeutic consequences. A check-up after 13 months showed no changes in the findings (Fig 2-a – 2-d).

Case 3

A 14-year-old boy suffered from chronic tension-type headache and attention deficit disorder. MR imaging showed a sharply demarcated extra-axial cystic lesion measuring 1.6 x 1.9 x 1.2 cm posterior of the medulla to the right of the midline. Compared to CSF, the cyst was hyperintense in T2-, T1-WI and FLAIR. In CISS sequence, the structure was hypointense to CSF, but hyperintense in comparison to brain tissue. Since the boy's complaints were not attributable to the cyst and a subsequent check-up showed no change; no histological verification was indicated (Fig 3-a – 3-d).

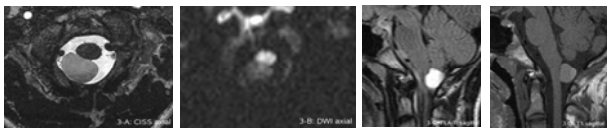

Fig. 3:
Approximately 1.6 x 1.9 x 1.2 cm, extra-axial, cystic lesion posterior to the medulla right of the midline. Hyperintense signal in the FLAIR sequence (C, sagittal) compared to CSF and brain tissue. Relatively rich signal in T1WI (D, sagittal). In the CISS sequence (A, axial) slightly hypointense compared to CSF, but hyperintense to the brain tissue. No restriction in DWI (B, axial).

Discussion

Intracranial NECs are mostly located infratentorial, intradural, extra-axial, near the midline, anterior to the brain stem and show typical MR signal characteristics: sharply demarcated and slightly to distinctly hyperintense compared to CSF in both T1-WI and T2-WI. Slight rim enhancement of the cyst wall after administration of contrast media and mild restriction in DWI are rare findings [5]. In the FLAIR sequence, the cysts show a hyperintense signal compared to CSF [7-10] because of the signal drop of low-protein fluids such as CSF in this sequence [11,12]. On the other hand, CSF shows a high signal in the CISS sequence, which is caused by the very similar T2 and T1 times and the characteristic T2/T1 weighting of all balanced steady-state free-precession sequences [13,14]. Therefore, in the CISS sequence the cysts are slightly hypointense compared to CSF but still hyperintense in comparison to the brain tissue. Due to the characteristic appearance at two different time points, such lesions can be classified with very high certainty as NECs.

Differential diagnoses for NECs are arachnoid cysts, which follow CSF in all sequences and can therefore be clearly distinguished, and epidermoid cysts, which can reliably be determined through the pathognomonic signal increase in DWI [5,7,15]. NECs can occur as small incidental findings in imaging or as large space-occupying lesions with corresponding clinical symptoms depending on their location. Symptomatic cysts require an evacuation of the cyst content and, if possible, complete marsupialization of the cyst wall. Recurrence or even metastatic dissemination has been reported in a few cases [16-18]. In the case of slight symptoms or incidentally discovered cysts, a conservative approach with check-ups at longer intervals is recommended as many cysts have no or only a slight tendency to grow [5].

The intracranial structures we detected incidentally displayed the typical MRI characteristics of NECs with typical infratentorial location on the ventral side of the brain stem and correspond very well to the histologically confirmed cases reported in the literature, as well as in their location as in their signal characteristics.

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Corresponding author:
Dr. med. Sascha Prothmann
University hospital rechts der Isar
Department of Neuroradiology
Ismaninger Str. 22
81675 Munich
Germany
phone: +49 89 4140 4651
fax: +49 89 4140 4887
e-mail: sascha.prothmann@roe.med.tu-muenchen.de