Short Illustrated Review

Enterogenous cysts of the cerebellopontine angle: short review illustrated by two new patients

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Summary

Intracranial enterogenous cysts are an uncommon entity rarely found in the midline within the posterior cranial fossa. The occurrence of an enterogenous cyst in the cerebellopontine angle is exceptional. We present two new cases of cerebellopontine angle (CPA) enterogenous cysts and review the literature to clarify the diagnosis and the management of these lesions.

Eighteen cases of CPA intradural enterogenous cysts have been reported to date, including the two cases presented in this article. All of them were symptomatic and underwent surgical treatment. After surgery, the symptomatic recurrence occurred in 31% of the patients, most of which had partial excision. Considering our patients and the published cases in the literature we suggest that the aim of surgery should be total removal of cyst and its content whenever possible. When partial resection of the cyst is performed, we recommend long-term clinical and neuroradiological follow-up.

Keywords: Cerebellopontine angle; enterogenous cyst; epithelial cyst.

Introduction

Enterogenous cysts are rare developmental cysts, lined by a mucous-secreting epithelium of a presumed endodermal origin [5]. They are most frequently reported in the lower cervical and upper thoracic spine, usually located ventral to the spinal cord [28]. Intracranial enterogenous cysts are quite rare and mostly observed along the midline in the posterior cranial fossa, anterior to the brainstem or in the fourth ventricle [3]. On rare occasions, an enterogenous cyst occurs in the cerebellopontine angle (CPA), which can lead to a misdiagnosis of this type of cyst with the more common arachnoid and epidermoid cysts [2, 20, 33].

We report two patients harbouring an enterogenous cyst at the CPA and we review the published cases of other such cysts [2, 4, 7, 10, 14, 20, 24–26, 29, 30, 32, 33, 35] to clarify the clinical, radiological, pathological and surgical aspects of these unusual lesions.

Literature review

The PubMed search engine of the national library of medicine and national institutes of health (www.pubmed.gov) was utilized to conduct an internet medline literature research based on the following search terms: “endodermal cyst”, “enterogenous cyst”, “neurenteric cyst”, “epithelial cyst”, “posterior cranial fossa” and “cerebellopontine angle”. Only reports in English were used and no date limitations were imposed in the search criteria. Solely pathologically proven intradural enterogenous cysts were considered, while midline posterior cranial fossa cysts extending to CPA were excluded.
Cases from bibliographies of retrieved references were included. In total, 71 cases of posterior cranial fossa enterogenous cysts were found and 16 of these were located in the CPA. Fourteen relevant reports dealing with the topic of enterogenous cyst of the CPA have been analyzed in our review. Thirteen authors reported on a single case [2, 4, 7, 10, 14, 20, 24–26, 29, 32, 33, 35] and one reported three cases [30]. Published data were reviewed according to clinical and demographic features, radiological appearance, surgical treatment, histological results and postoperative outcome.

Analysis of literature review

The results of our analysis are summarized in Table 1. Eighteen cases of intradural enterogenous cysts of CPA have been reported in the literature, including the two cases described in this article [2, 4, 7, 10, 14, 20, 24–26, 29, 30, 32, 33, 35]. The patient’s sex ratio showed a clear predominance of women (72%) to men (28%). Cysts localization was 61% on the left side and 39% on the right side. No cases of bilateral localization have been reported. The patient’s age at first presentation ranged from 14 to 67 years (average age 40 years).

Clinical features

The most frequent symptoms were cochleovestibular defined as follows: vertigo or balance disorders, 50%; hearing loss, 44%; and tinnitus, 11%. No patient was asymptomatic. Headache was reported in as many as 33% of the cases. Other clinical findings included facial nerve lesion symptoms (two cases) [35], transient blurred vision (two cases) [25, 32] and lower cranial nerve deficits (one case) [24]. Trigeminal signs, such hemifacial sensory loss and trigeminal neuralgia, occurred in three cases [10, 30]. One case presented with three bouts of chemical meningitis [29]. Diplopia caused by VI cranial nerve palsy occurred in one patient [26].

Radiological features

All patients but four underwent CT scanning. CT showed a low-density cyst in 71% of the cases (10 patients) [2, 7, 14, 20, 25, 26, 32, 35] with no contrast enhancement in 8 cases [2, 7, 14, 20, 25, 26] and a thin contrast enhancement around the lesion in two [32, 35]. In one case CT scanning revealed a slightly hyperattenuating mass [33] and in another the cyst was isodense compared to brain [30]. In two cases CT failed to reveal intracranial lesion [24, 29]. Bone windows showed erosion of the posterior surface of the petrous bone in two cases [32, 35]. The enterogenous cysts were avascular in all cases in which the angiography findings were described (6 cases). MRI was performed in 11 cases [2, 4, 7, 10, 30, 32, 33] and revealed a round or lobulated nonenhancing mass isointense or slightly hyperintense relative to CSF on T1-weighted MR images and isointense relative to CSF on T2-weighted MR images in all cases but one [4] in which the cyst was heterogenous on T1-weighted MR images. A solid nodule with strong enhancement from xanthogranulomatous reaction was detected in one case [33] and a posterior rim enhancement at the interface between the cyst and the adjacent nervous parenchyma was detected in another case [30]. Hydrocephalus was found in 4 patients [14, 24, 26, 32] one of which required a ventriculoperitoneal shunt insertion before cyst removal [24].

Surgical data

The lateral suboccipital approach was the most frequently used surgical procedure (13 patients). In one patient [33] the far-lateral approach was performed and in four cases the surgical approach was unknown [4, 30]. Tumor resection was assessed in 14 patients. Total cyst removal was achieved in 10 patients (71%). In 2 cases (14%) the cyst resection was incomplete due to adherent portions of the cyst to the pons in one case [26] and to the lower cranial nerves in the other [20]. Two patients initially underwent cyst biopsy and fenestration [10].

Postoperative outcomes

Postoperative outcomes were available in 13 of 18 patients. Four patients (31%) experienced recurrences. One patient had a recurrence 2 months after biopsy and fenestration of a multiloculated cyst [10]. In another patient, cyst recurred at 2 months and at 1 year after macroscopically complete cyst removal [7]. One patient had a recurrence at 9 years and at 10 years after the first surgery during which partial cyst resection was performed [26]. Finally the second patient described in this report had a recurrence 14 years after drainage and fenestration of the cyst. All recurrences were symptomatic and required an additional surgical procedure. Seven of the remaining 9 patients improved after surgery and 2 had new postoperative deficits. One of these patients experienced mild hoarseness [29] and the other complained facial palsy and mild extremity weakness [32].