Cerebrospinal fluid leak with recurrent meningitis following tonsillectomy

Case report

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The authors report an unusual case of bilateral large petrous apex cephaloceles in a 14-year-old boy with a history of recurrent meningitis. Although these lesions are rare and usually asymptomatic, surgical correction is recommended if they are associated with a persistent CSF leak. In this patient, the extensive bilateral cranial defects were not adequately treated by an intracranial approach alone. Repair of a defect in the posterior pharyngeal wall, the site of a prior tonsillectomy, ultimately resulted in repair of the CSF fistula. (DOI: 10.3171/2009.10.PEDS09336)

KEY WORDS • cerebrospinal fluid leak • petrous apex cephalocele • tonsillectomy

Bacterial meningitis is a rare and potentially life-threatening condition. Among other causes, anatomical, immunological, congenital, and oncological conditions can result in a CSF fistula that links the intradural compartment to the external environment, thus predisposing the patient to recurrent bouts of meningitis. Petrous apex cephaloceles represent a protrusion of meninges and CSF from the Meckel cave into the petrous apex.6 These lesions are often asymptomatic incidental findings discovered on imaging performed for a variety of clinical concerns. In children, however, there are reports of PACs associated with spontaneous CSF leaks resulting in recurrent bacterial meningitis.5,6,11 We report on a patient with large bilateral PACs, which were asymptomatic until a routine tonsillectomy resulted in a CSF fistula, causing recurrent episodes of bacterial meningitis. The relationship between the tonsillectomy and the CSF leak was not appreciated until the second attempt at repair. Preoperative imaging, intraoperative findings, and technical aspects of the repair are discussed.

Abbreviation used in this paper: PAC = petrous apex cephalocele.

Case Report

History. This 14-year-old boy, previously in excellent health, presented to another hospital for routine tonsillectomy and adenoidectomy. This procedure was performed without apparent complication, and the patient was discharged from the care of his treating otolaryngologist. Fifteen months after the tonsillectomy, the patient was hospitalized with bacterial meningitis. Over the next 18 months, the patient experienced 2 other episodes of meningitis. Each episode was associated with a different bacterial species; the first was caused by Streptococcus Group A, the second by Streptococcus Group B, and the third by Staphylococcus species. Intracranial hypotension was not suspected. The patient had no headaches between meningitic episodes. Similarly, he denied any abnormal taste sensations. Because of the recurrent episodes of meningitis, the patient underwent intracranial imaging. Head MR imaging studies (Fig. 1) revealed skull base lesions centered in the petrous apex portions of the temporal bones bilaterally, with a larger lesion identified on the left than on the right. There was no evidence of intracranial hypotension on imaging; no pathological pachymeningeal enhancement or “brain sag” was pres-
Recurrent meningitis with CSF leak after tonsillectomy

Examination and First Operation. The patient was referred to our medical center and underwent CT cisternography with iodinated contrast material administered via lumbar puncture (Fig. 2), which showed complex defects involving the skull base, fovea ethmoidalis, basi- sphenoid, and the medial aspects of both temporal bones, with bilateral meningoceles (larger on the left) arising in the petrous apex regions. Frank leakage of contrast into the retropharyngeal space was also identified, but no site of egress of spinal fluid into the pharynx was seen. Given the imaging findings, there was suspicion that CSF was communicating with the nasopharynx through the large left PAC. The patient was subsequently taken to the operating room for a left subtemporal craniotomy for exploration of these skull defects, pericranium patch and fat graft, duroplasty, and temporary lumbar drain placement. The left-sided combined intradural and extradural approach was chosen because that side was thought to have a more significant leak. At the time of that operation, the large cranial defects were easily identified and an attempt was made to fill them with fat, a pericranium patch, and fibrin glue. The lumbar drain was left in place for 4 days postoperatively. A second, right-sided approach was considered as a second stage only if the left side was repaired successfully and the right-sided leak persisted.

Three months postoperatively, however, CT cisternography showed a persistent leak through the extensive skull base defects bilaterally. The decision was made to direct our attention to potential distal sites of CSF egress.

Second Operation. The patient was returned to the operating room for a second attempt at surgical repair, with special attention to any potential site of distal CSF egress. A lumbar drain was placed. Fluorescein dye was injected through the lumbar drain, and the patient was placed in the Trendelenburg position for 10 minutes. Direct laryngoscopy was then performed to visualize the upper airway from the oral cavity to the larynx. With intraoperative CT-based navigation, a wide maxillary antrostomy was then performed, followed by evaluation and removal of the anterior and posterior ethmoid air cells. The sphenoid sinus was then identified and widely exposed. There was no evidence of CSF leakage or bone defect at these sites. Endoscopic ethmoidectomy and sphenoidotomy were performed to visualize the left ethmoid roof and sphenoid walls. No bone of the anterior or middle fossae was removed. No bone of the anterior or middle fossae was removed. Attention was then directed to the nasopharynx. A CSF leak was appreciated in the adenoid pad region overlying the clivus, the site of a prior adenoidectomy. The palate was then split in the midline, veering just lateral to the uvula, resulting in excellent exposure of the adenoid pad CSF leak. Sutures were placed, pulling the tissue medially to close the leak. The surface overlying the sutures was cauterized and coated with fibrin glue. The lumbar drain was left in place, with continuous drainage for 4 days postoperatively. The patient made an uneventful recovery from his procedure.
**Third Operation and Postoperative Course.** Twelve months after the initial repair, the patient was taken back to the operating room for lumbar puncture, intrathecal injection of fluorescein dye, direct laryngoscopy, and examination of the nasopharynx under anesthesia. At that time, the patient was asymptomatic. This final procedure was conducted to confirm the durability of the repair. In contrast to his preoperative examination, there was no longer any fluorescein dye leak or evidence of a CSF leak into the nasopharyngeal or oropharyngeal regions. The patient has had no additional episodes of meningitis over a 2-year follow-up interval.

**Discussion**

A PAC is a protrusion of the contents of the Meckel cave into the petrous portion of the temporal bone. These lesions are usually unilateral and asymptomatic. Although they are usually detected incidentally, PACs are occasionally associated with CSF rhinorrhea, otorrhea, or meningitis. Twenty-five PACs have been reported in the literature, with only 3 patients demonstrating bilateral lesions. Because they are usually asymptomatic, the incidence of PACs is not well established, but has been estimated to be between 1/10,000 and 1/100,000. A PAC may be either congenital or acquired. Congenital cases are thought to originate during embryogenesis in which mesenchymal clefts fail to close, leading to the development of encephaloceles in the skull base. Acquired PACs may also be due to intracranial hypertension, and have been associated with pseudotumor cerebri and empty sella syndrome.

Accurate diagnosis and identification of the site of fistula formation is critical for surgical planning. A PAC is distinguished from other conditions based on its location within, and contiguity with, the Meckel cave, and also by its well-delineated smooth bone margins. Both CT and MR imaging have been used to characterize PACs and to localize the site of a fistula. In retrospective analyses, high-resolution CT cisternography yielded positive results in 85–89% of cases when an active leak was present. Magnetic resonance cisternography has a sensitivity of >90% for detecting a CSF fistula. In this case, we found that thin-cut CT cisternography following intrathecal contrast administration was more helpful for illustrating the necessary anatomical details. The technique of intrathecal injection of fluorescein dye was described by Bateman et al., and was used when CT and MR imaging were unable to delineate the fistula. Although there are no studies reviewing the sensitivity of this technique, it proved to be useful in our patient. Intrathecal fluorescein injection may occasionally result in serious complications, including status epilepticus or spinal cord injury. The incidence of complications with this technique appears to be dose dependent; side effects are seen less frequently at lower doses and concentrations.

There are only 2 previously reported cases of CSF fistulas extending through a PAC that resulted in bacterial meningitis. Our patient’s PACs were bilateral and quite extensive, and most likely resulted in an initially asymptomatic CSF leak into the retropharyngeal space. A properly performed tonsillectomy then allowed the retropharyngeal CSF collection to communicate with the pharynx, resulting in repeated episodes of bacterial meningitis. This rare complication of recurrent bacterial meningitis after tonsil and adenoid surgery has not been previously reported.

Based on our experience with this patient, we believe that the optimal surgical management for large bilateral PACs resulting in a CSF leak should be repair of the distal site when possible. Repair of the distal site was straightforward and effective in our case. On the other hand, the attempted intracranial repair was ineffective in our case, and we will not attempt such a maneuver again in any similar case if a distal repair is possible. In some cases, as in this patient, the site of distal CSF egress is not readily apparent on imaging studies. A history of any trauma or surgical procedure to the head or neck should raise clinical suspicion and prompt directed radiological investigation or even direct exploration for a potential site of CSF egress.

**Conclusions**

A PAC is a rare skull base lesion that may be associated with spontaneous CSF fistulas and recurrent bacterial meningitis in children. Our case illustrates surgical repair of a previously unreported complication of tonsillectomy that resulted in a CSF fistula extending from a PAC into the nasopharynx through the adenoid fat pad.

**Disclosure**

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper. Author contributions to the study and manuscript preparation include the following. Conception and design: CO Maher, SL Hervey-Jumper. Manuscript writing: CO Maher, SL Hervey-Jumper, AK Ghori, LJ Marentette, DJ Quint. Data extraction: SL Hervey-Jumper, AK Ghori, CO Maher. Final approval: CO Maher, SL Hervey-Jumper, AK Ghori, LJ Marentette, DJ Quint.

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Recurrent meningitis with CSF leak after tonsillectomy