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73.1 Epidemiology and Clinical Presentation

- Cerebrospinal fluid (CSF) rhinorrhea may occur when a communication (fistula) develops between the nasal cavity and subarachnoid space. For this to occur, the bone, dura, and arachnoid membrane must all be breached by the offending process. CSF rhinorrhea may develop spontaneously or as a result of a variety of inciting factors, including craniofacial trauma, surgery, radiation, or medical treatment.
- If left untreated, CSF rhinorrhea may result in meningitis, pneumocephalus, and intracranial hypotension.

73.1.1 Spontaneous CSF Rhinorrhea

- Spontaneous CSF rhinorrhea classically develops in obese patients with idiopathic intracranial hypertension, also known as pseudotumor cerebri. The condition is more common in middle-aged women, especially those with obesity and/or multiparity [1–3].

- The mean age of patients with spontaneous CSF rhinorrhea is 50–53 years [3, 4].
- Concomitant findings often associated with spontaneous CSF rhinorrhea include elevated intracranial pressure, meningoencephaloceles, visual loss, an empty sella, and skull base erosion [5].
- The most common locations of CSF fistulas causing rhinorrhea are the lateral lamina of the ethmoid and the fovea ethmoidalis [6].
- Spontaneous CSF rhinorrhea may also develop in the setting of a pituitary adenoma or other skull base tumor, most commonly following successful medical treatment of large, invasive prolactinomas. CSF leaks following radiation and radiosurgery have also been reported [7, 8].

73.1.2 Traumatic CSF Rhinorrhea

- The incidence of CSF rhinorrhea following traumatic brain injury is 4.6 % [9].

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- CSF rhinorrhea following traumatic brain injury may be associated with skull base fractures, visual loss, anosmia, hypopituitarism, vascular injuries, and meningoencephaloceles. (See the previous section on Traumatic Injury of the Sellar Region.)

73.1.3 Postoperative CSF Rhinorrhea

- Following standard transsphenoidal operations for pituitary tumors, the incidence of CSF rhinorrhea is as high as 10 %. In recent series, however, the usual incidence is 1–4 % [10].
- In extended endonasal endoscopic surgeries, the incidence of postoperative CSF rhinorrhea was initially as high as 58 %. The more frequent use of lumbar drainage and pedicled nasoseptal flaps, however, has greatly reduced this incidence to approximately 5 % [11, 12].

73.2 Imaging Features

- Plain axial CT imaging will often show pneumocephalus but is not highly sensitive in identifying the fistula site (Fig. 73.3).
- Thin-slice CT imaging with sagittal and coronal reconstruction will often show the site of the bony defect (>90 % sensitivity) and sometimes an encephalocele (Fig. 73.2) [13].
- CT cisternography is a highly effective modality for identifying the site of CSF fistula (Fig. 73.1). It often shows contrast extravasation through a bony defect and pooling in a paranasal sinus. The technique is accurate in over 90 % of patients, but only 30–40 % in the absence of an active CSF leak [14, 15].
- MR cisternography is also a highly effective test for identifying a specific CSF leak site, with a sensitivity of 92 % and specificity of 100 %. It often shows a T2-hyperintense column of CSF communicating between the subarachnoid space and sinuses (Fig. 73.4). An encephalocele may be seen, often with corresponding encephalomalacia, edema, and downward traction of the lobe of origin [14–17].
- Radionuclide cisternography can be used to confirm the presence of a skull base CSF leak and to aid in its localization [18].

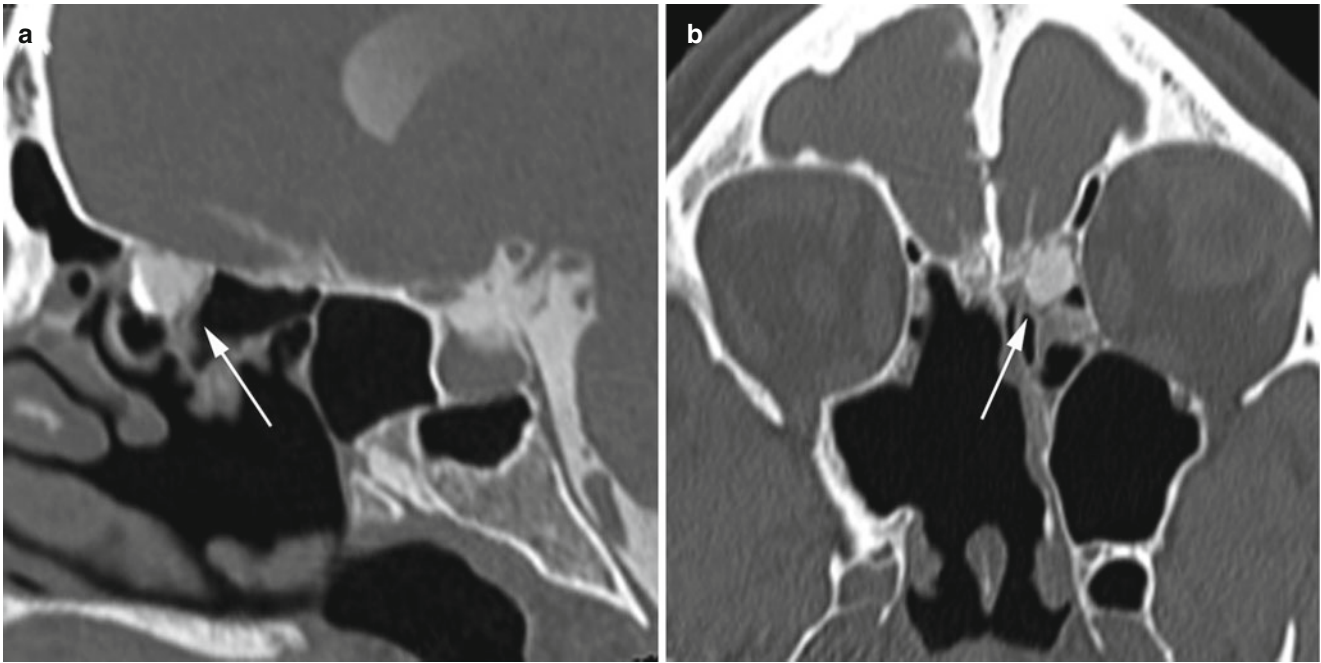


Fig. 73.1 CSF leak. (a) Sagittal CT cisternogram image. (b) Coronal CT cisternogram image. Postoperative changes from prior right ethmoid resection and medial maxillary antrostomy are present. In both

images, contrast pooling is detected in one of the ethmoid air cells on the left, immediately below the roof (*arrows*), consistent with leakage of CSF. A bony defect is not directly visualized

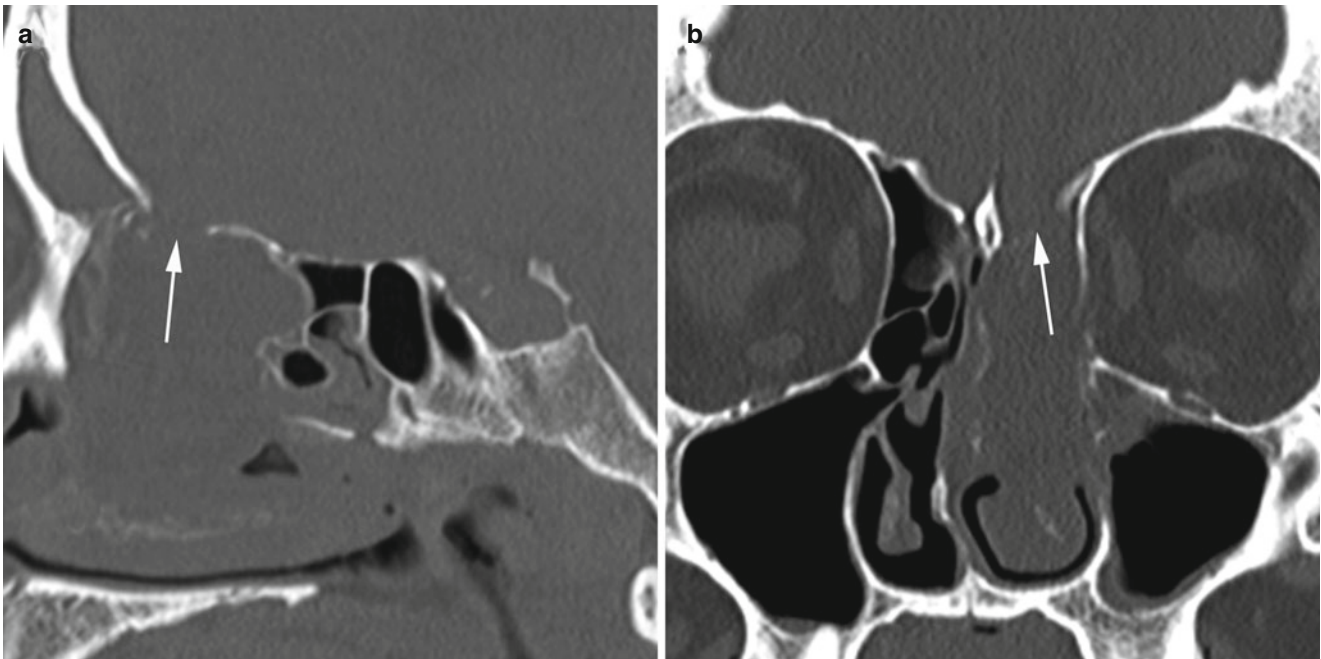


Fig. 73.2 CSF leak. (a) Sagittal CT image without contrast. (b) Coronal CT image without contrast. In both images, a bony defect is seen in the roof of the left ethmoid (*arrows*), with complete opacification of the left ethmoid air cells and part of the left nasal cavity

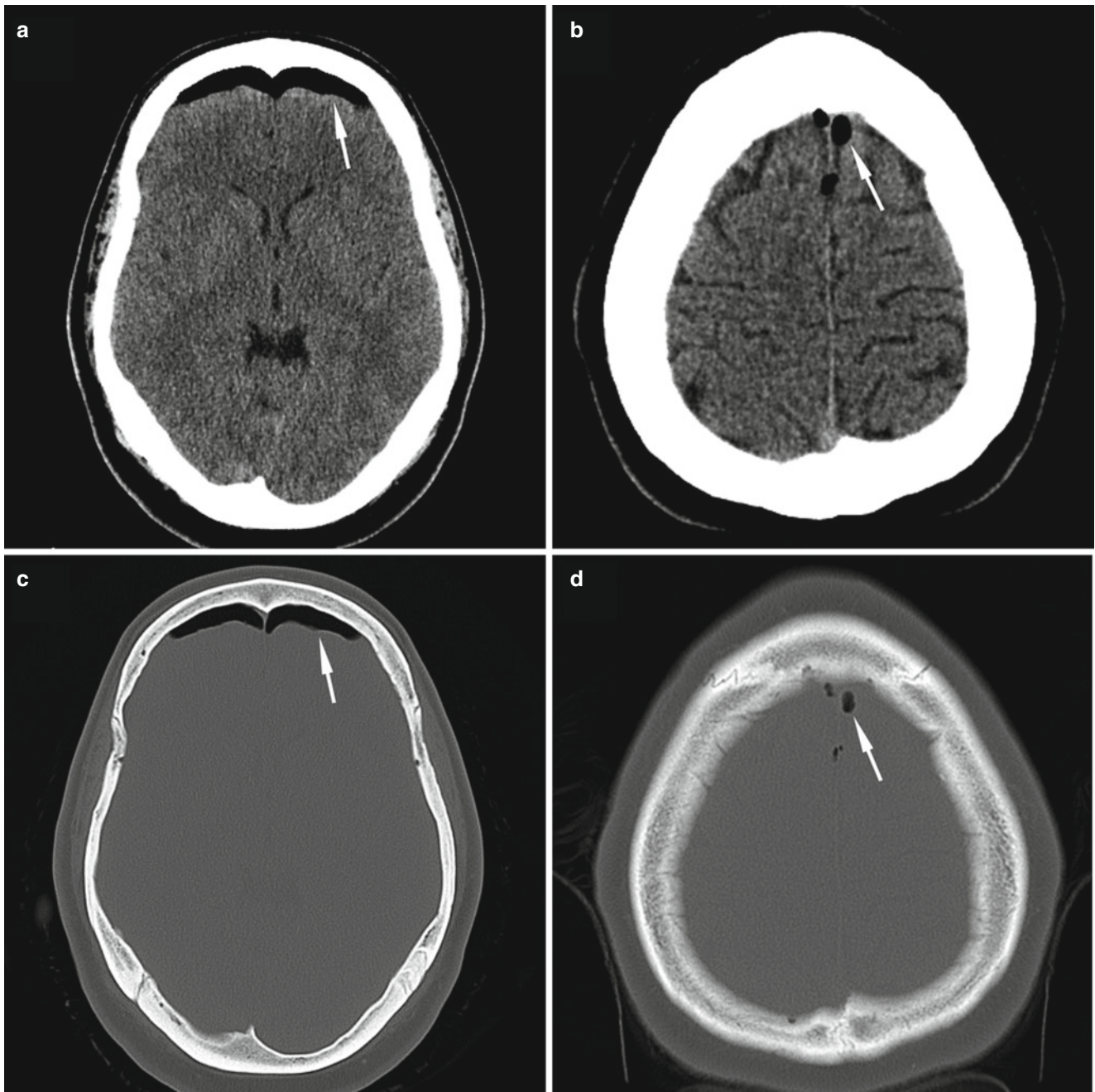


Fig. 73.3 CSF leak. Noncontrast axial CT images in a patient with a postoperative CSF leak following pituitary surgery. Images (a, b) are windowed for brain soft tissue, and images (c, d) are windowed for

bone imaging. Pneumocephalus (arrows) can be seen in the frontal region in (a, c) and near the superior sagittal sinus, along the convexity, in (b, d)

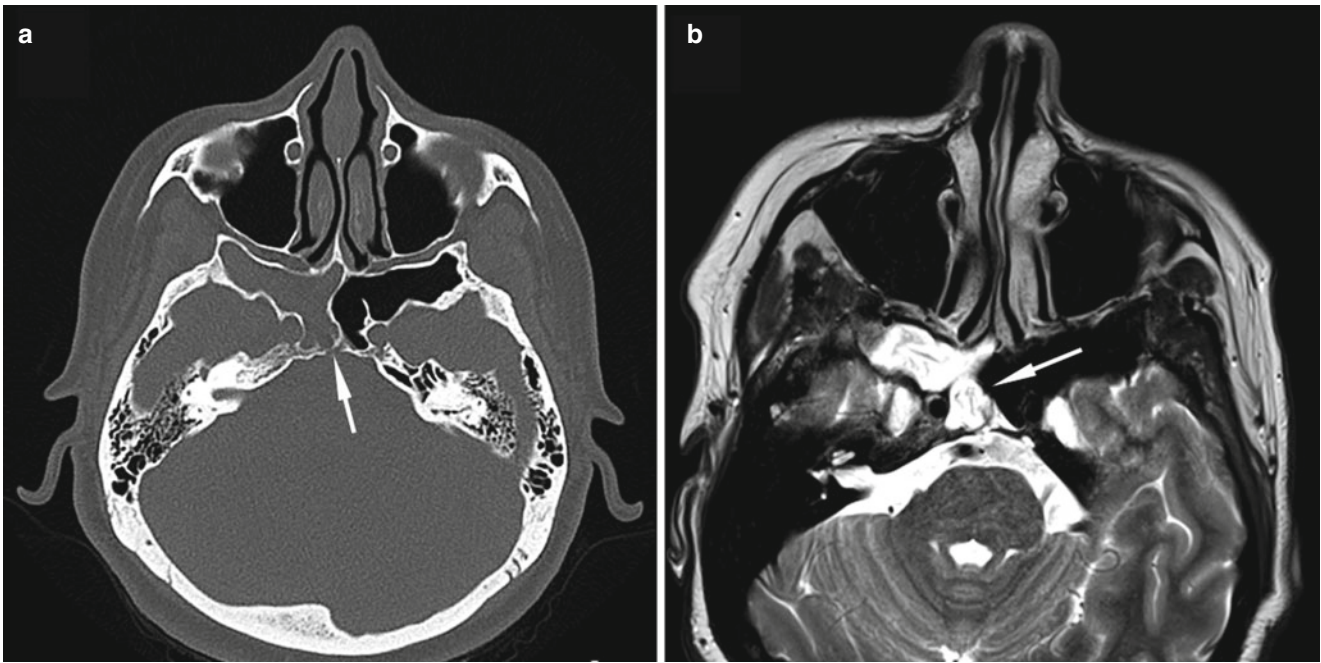


Fig. 73.4 CSF leak. Axial noncontrast CT image (a) and noncontrast T2-weighted MR image (b) in a patient with a CSF leak arising from the clivus and presenting with right-sided rhinorrhea. CT imaging

shows the area of bony dehiscence (*arrow*) and opacification of the right sphenoid sinus (a). MR imaging shows T2-hyperintense CSF pooling (*arrow*) in the right sphenoid sinus (b)

73.3 Clinical and Surgical Management

73.3.1 Diagnosis of CSF Leaks

- In most patients, the diagnosis of CSF rhinorrhea is clinical. The rhinorrhea may be dependent on exertion or on position, worsening when the patient leans forward (“tilt test”). CSF leaks often appear as clear or blood-tinged fluid that looks like a drip from a faucet as a patient tilts forward. Also associated with CSF leaks is the “halo” sign, in which a yellowish ring forms on paper or gauze when CSF is present.
- Confirmation of CSF may be done via laboratory analysis for beta-2-transferrin, which is highly specific in differentiating CSF from other clear fluids such as nasal secretions or tears.
- A CT scan of the head may show pneumocephalus, which is highly suggestive of a CSF fistula.
- In some patients with CSF rhinorrhea, the exact fistula point cannot be identified with standard imaging. In these patients, tests such as CT cisternography or radionuclide cisternography studies can be performed to help identify the site of leakage [18].
- Intraoperatively, during a transsphenoidal operation, a Valsalva maneuver should be performed to assess for a CSF leak. Many postoperative CSF leaks develop because the leak was not identified during surgery, rather than representing a failed sellar repair [19].
- In other patients, the intraoperative use of intrathecal fluorescein can help to identify and confirm the fistula site [19, 20].

73.3.2 Management of CSF Rhinorrhea

- CSF leaks resolve spontaneously in some patients, depending on the etiology.
- Medical management with acetazolamide, which reduces the production of CSF and therefore reduces intracranial pressure, can be used as a temporizing measure. It may cause severe nausea and vomiting, however, which can exacerbate the CSF leak, so acetazolamide should be used only after weighing the risks and benefits.
- Lumbar punctures and temporary lumbar drains are often successful treatments for CSF leaks causing rhinorrhea. These maneuvers usually work for smaller leaks and those where there is sufficient tissue in place to form a permanent scar, or seal, at the fistula point.
- Many surgical options are available for repair of skull base CSF leaks, including autologous fat and fascia, dural substitutes, collagen sponge, fibrin glue, and dural sealants. Multilayer closures tend to offer the best chance of achieving a permanent repair.

- For larger leaks with wide bony deficits, the use of a rigid buttress may be beneficial. This can be crafted from bone (the vomer or turbinate), cartilage, titanium, or an absorbable synthetic plate.
- The use of pedicled nasal or temporalis flaps can be highly successful in closing larger skull base defects, greatly adding to the armamentarium of endoscopic skull base surgeons. The most commonly used flap is the pedicled nasoseptal flap [21–23].
- Patients with refractory CSF leaks despite multiple attempts at surgical repair often have underlying intracranial hypertension, and the definitive management option may be insertion of a CSF-diverting shunt, such as a permanent ventriculoperitoneal or lumbar-peritoneal shunt.
- Intracranial pressure may increase following successful repair of a CSF leak, causing new symptoms or leaks in other locations [4].
- Outcomes following CSF leak repair are generally excellent, with the leak being repaired on the first attempt in over 90 % of patients [4, 24].

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