LYMPHANGIOMA comprises a benign proliferation of lymph vessels and is a congenital tumor. Roughly two-thirds of all lymphangiomas are identified shortly after birth, with most occurring in the soft tissues of the neck, axilla, and mediastinum. Bone is a rare location. Lymphangioma of the bone was first described in 1947 by Bickel and Broders. Several cases of lymphangioma in the skull base have been reported, but most have involved lesions extending or disseminating into the skull base from a primary neck lesion. We describe herein a case of lymphangioma localized in the skull base and leading to CSF rhinorrhea with meningitis.

**Case Report**

**History and Examination.** This 5-year-old boy had recurrent meningitis 4 times since he was 3 years old, presenting with CSF rhinorrhea on each occasion. Blood and CSF cultures detected *Streptococcus pneumoniae* on the second and third presentations with meningitis. Antibiotics were administered for the meningitis, with complete recovery each time.

Lymphangioma localized to the bones of the skull base is rare. The authors report herein the case of a 5-year-old boy who presented with lymphangioma of the bone, localized to the skull base and leading to cerebrospinal fluid (CSF) rhinorrhea with meningitis. Neuroimaging demonstrated lytic destruction with a cyst in the right middle skull base. The patient was successfully treated with resection of the tumor and prevention of CSF leakage. Histopathological examination revealed a lymphangioma. An enlarging lymphangioma can lead to bone destruction. A differential diagnosis of a lytic lesion for a cyst at the skull base is important for proper case management.

(DOI: 10.3171.PED.2008.2.10.276)

**KEY WORDS** • cerebrospinal fluid leakage • lymphangioma • skull base

When the boy was 1 year old, MR imaging revealed a cervical cystic lesion extending to the mediastinum without connection to the intracranial space (Fig. 1A and B). He underwent partial resection of a cervical lymphangioma, followed by the injection of sclerosing agents. The cervical lesion gradually shrank, and the mediastinal lesion was residual (Fig. 1C).

Magnetic resonance imaging (Fig. 2) demonstrated a localized cystic lesion in the right middle skull base. This lesion was not connected to any extracranial tissues. Computed tomography revealed a lytic lesion with bone destruction in the right middle skull base (Fig. 3A and B). Three-dimensional CT showed the osseous defect on the lateral wall of the sphenoid bone (Fig. 3C). Radioisotope cisternography showed a hot spot in the cystic lesion, indicating communication of this lesion with the subarachnoid space.

Based on these findings, the lesion was diagnosed as the leak point of CSF into the sphenoid sinus and the cause of subsequent meningitis. Surgery was performed to prevent CSF leakage and determine the histopathological features of the cystic lesion.

**Operation.** With the patient in a state of general anesthesia, the otolaryngologist performed sphenoidotomy to eliminate infection in the sphenoid sinus and enlarge the drain-
age route from the sinus. No active infection was present and no lesion was apparent in the sphenoid sinus. After making a right temporal skin incision, the plastic surgeon created a temporoparietal muscle-galeal flap. A right temporal craniotomy was then made. The middle cranial base was epidurally exposed. A thin-walled cyst was identified between the foramen rotundum and foramen ovale (Fig. 4A). Bone around the cyst was drilled to fully expose the cyst, and the lesion was removed to the fullest extent possible. The cyst wall was inspected using an endoscope with a 70° angle, but a connection between the defect and sphenoid sinus was difficult to identify because of the blind angle. A dural hole connecting to the intradural space was eventually located medial to the second division of the trigeminal nerve (Fig. 4B). The hole was covered with a piece of temporal fascia, and the osseous defect was packed with the temporoparietal muscle-galeal flap.

Postoperative Course. The patient’s postoperative course was uneventful, and he was discharged without neurological deficit or CSF leakage. No recurrence of CSF leakage or meningitis had occurred as of 16 months after surgery.

Photomicrography of the surgical specimen revealed thick endothelial cells lining the cystic wall with invasion of lymphocytes (Fig. 5A). Immunohistochemical staining demonstrated positive expression for D2-40 in cells lining the cystic wall (Fig. 5B). The histopathological diagnosis was lymphangioma.

Discussion

Lymphangioma of the skull base bone is rare. To the best of our knowledge, Kaya et al.11 provided the first report of primary lymphangioma localized in the clivus. The most frequent sites of bone lesion are the tibia, humerus, ilium, cranium, mandible, and vertebrae.12 Such lesions can be associated with substantial bone loss, such as in Gorham disease or vanishing bone disease.7,9 Several diseases involving lytic lesions at the skull base have been described, including chordoma, chondroid chordoma, chondrosarcoma, plasma-cytoma, fibrous dysplasia, giant cell tumor, Paget disease, lymphoma, eosinophilic granuloma, meningocele, and nasopharyngeal carcinoma.1,4,5,10,13,20,21 Lymphangioma should be considered among the differential diagnoses of a lytic lesion at the skull base.

Most cases of lymphangioma in the skull base have involved lesions extending or disseminating into the skull base from a primary neck lesion.16,18 In the present case, lymphangioma of the skull base was completely separate from the cervical lesion. Lymphangioma is congenital and can be multifocal.14,22 The skull base lesion in the patient in the present case therefore may have been occult until the meningitis developed.

In lymphangioma of the bone, symptoms are related to pressure and/or erosion of bone with enlargement of the lymphatics.6,7 Patients may present with local pain and swelling or pathological fracture,12 or the disease may be diagnosed incidentally when radiography is performed for other purposes. Patients with lymphangioma of the clivus can present with diplopia and headache.11 The patient in the present case showed bone destruction leading to CSF leakage with meningitis. Lymphangioma of the bone can also cause lytic destruction at the skull base. Close follow-up for bone lesions is thus recommended even in asymptomatic patients.

The preferred treatment for lymphangioma is complete resection. Although laser treatment or injection of sclerosing agents represents alternate therapies for lymphangioma of the soft tissue, such options should be not used for intracranial lesions given the central nervous system’s lack of tolerance. In the present case, surgical treatment was selected because of the complication of CSF leakage and to obtain a histopathological diagnosis. Lymphangioma has a strong tendency to local recurrence if excision is incomplete. Recurrence rates are reportedly 15–50%.17 In addition, mortality rates are 3–7% and related to complications such as respiratory compromise, aspiration, and infection in crit-
ical areas such as the neck. In fact, total resection is difficult to achieve, as vital or functionally important structures cannot be sacrificed.

Early recognition of a recurrence or regrowth is necessary for tumor control in the natural history. Several surgical approaches to prevent CSF rhinorrhea have been reported. A neurosurgical approach offers direct access to and wide exposure of the lesion, albeit with the morbidity associated with craniotomy. A rhinological approach, such as transseptal or transethmoidal approaches, would not have permitted access to this lesion as it was not located in the sphenoid sinus. An infratemporal fossa approach also could have been used to access this area, but the procedure carries a considerable risk of morbidity for such a benign disease. The patient in the present case was successfully treated via a combination of the transcranial and transnasal approaches. We performed sphenoidotomy via a transnasal approach to eliminate the source of infection. The dural hole was patched using a piece of fascia, and

![Image](image_url)

**Fig. 3.** Axial (A) and coronal (B) CT scans revealing a lytic lesion in the right middle skull base. Three-dimensional CT scan (C) showing an osseous defect (arrow) in the lateral wall of the sphenoid bone.

![Image](image_url)

**Fig. 4.** A: Intraoperative photograph showing a thin-walled cyst (arrow) in the bone between the foramen rotundum and foramen ovale. The far lateral triangle had been drilled away. S = suction tube; V2 = second division of the trigeminal nerve; V3 = third division of the trigeminal nerve. B: Endoscopic photograph with a 70° angle at the superior wall of the cyst showing a dural hole medial to V2.
the osseous defect was packed with a vascularized flap via a transcranial epidural approach to ensure the seal of the skull base without dead space.

Conclusions

Lymphangioma in bones of the skull base can cause lytic destruction with lesion enlargement. The differential diagnosis of lytic lesions for a cyst at the skull base is important for proper disease management.

Disclaimer

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

References


Accepted July 23, 2008.
Address correspondence to: Kiyoshi Saito, M.D., Department of Neurosurgery, Nagoya University Graduate School of Medicine, 65 Tsurumai, Showa-ku, Nagoya 466-8560, Japan. email: kiyoshis@med.nagoya-u.ac.jp.