CASE REPORT

Nasopharyngeal glial heterotopia with delayed postoperative meningitis

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SUMMARY

A male infant, who underwent radical resection of a large glial heterotopia at the nasopharynx at 8 days, developed delayed postoperative bacterial meningitis at 9 months. Neuroradiological examination clearly demonstrated that meningitis had occurred because of the intracranial and extracranial connections, which were scarcely seen in the perioperative period. A transsphenoidal extension of hypothalamic hamartoma is possible because the connection started from the right optic nerve, running through the transsphenoidal canal in the sphenoid bone and terminating at the recurrent mass in the nasopharyngeal region.

BACKGROUND

Nasal glial heterotopia is a midline nasofrontal mass composed of mature brain tissue isolated from the cranial cavity by closure of the cranial sutures during embryonic development.¹ Thirty per cent of glial heterotopia may manifest as an intranasal lesion. This nasal mass could cause neonatal airway obstruction because infants are nasal breathers for the first few months of life. Some patients require intubation during the neonatal period.^{2,3} Meningitis and/or cerebrospinal fluid (CSF) rhinorrhoea could not be complicated either before or after lesion resection because nasal glial heterotopia is separate from the intracranial contents,⁴ and no case of heterotopia causing postoperative meningitis has been reported. However, we encountered a case of a patient with glial heterotopia in the nasopharyngeal region, who developed meningitis 9 months postoperatively. Possible pathophysiological mechanisms of the delayed onset of postoperative meningitis will be discussed.

CASE PRESENTATION

A male neonate was delivered vaginally, without respiratory distress. However, a large cystic lesion in his right nasal cavity was noted at birth. Viscous fluid exuded after puncturing the cyst, and he became slightly dyspneic. After being referred to our hospital, rhinoscopy showed that the nasopharynx was filled with smooth and whitish mass (figure 1A). There was very little possibility that the fluid content was CSF because the protein level was more than 250 mg/dL, and glucose level was not elevated.

T1-weighted MRI (T1WI) with gadolinium (Gd) enhancement demonstrated a large multicystic lesion in the nasopharyngeal region (figure 1B). No

direct communication was found to the cerebrum and CSF space. The sagittal view of the T1-weighted image (T2WI) equivocally showed a tiny strand of high intensity in the sphenoid bone between the intracranial CSF space and cystic lesion (figure 1C).

After admission, we performed transnasal intubation to secure the airway. Transnasal surgical excision of the lesion was performed 8 days after birth. Under endoscopic guidance, the lesion was dissected from its base in the mucous membrane of the nasopharyngeal space, and the mass was totally removed. Neither CSF leakage nor bony defect was noted intraoperatively.

Microscopic examination of the lesion documented a cystic mass, with its wall mostly covered by the respiratory epithelium, consisted of variable sized islands of glial tissue separated with fibrovascular tissue (figure 1D). Rosette formation of ependymal cells were noted (figure 1E). The histopathological findings were consistent with those of nasal glial heterotopia.

The postoperative course was uneventful, and the patient was extubated on the sixth postoperative day. Follow-up MRI showed complete removal of the lesion but failed to reveal the connection (figure 1F).

At the age of 9 months, the patient was readmitted for respiratory syncytial viral pneumonia. However, he became lethargic on the third hospital day. Neck stiffness was also noted.

The anterior fontanelle was not distended. CSF examination showed leucocytosis ($0.527 \times 10^9/L$, 64% polymorphonuclear leucocytes), decreased glucose level (9 mg/dL), and increased protein level (426 mg/dL). Culture of the CSF, sputum and venous blood samples showed positive results for penicillin-resistant *Streptococcus pneumoniae*.

MRI with fluid-attenuated inversion recovery sequence with Gd administration revealed meningeal enhancement (figure 2A). The sagittal and coronal views of the conventional MRI and bone target CT scan clearly demonstrated the isointense stalk, running through the transsphenoidal canal in the sphenoid bone, between the right optic nerve in the intracranial CSF space and the recurrent mass in the nasopharyngeal space (figure 2B–E). With Gd administration, the stalk and recurrent tumour is enhanced (figure 2G).

TREATMENT

With the diagnosis of delayed postoperative meningitis via the connection between the CSF and nasopharyngeal spaces in the sphenoid bone,

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Figure 1 (A) Rhinoscopic examination reveals a cystic mass filling the left nasal cavity. (B) Sagittal view of the T1-weighted MRI (T1WI) with gadolinium (Gd) enhancement demonstrates the large multicystic lesion (*) in the nasopharyngeal region, not at the frontonasal region. No direct communication with the intracranial space was found. The white arrow indicates the foramen caecum. (C) Sagittal view of the T2WI equivocally shows a tiny strand of high intensity (white arrow) in the sphenoid bone between the intracranial cerebrospinal fluid space and cystic lesion. (B) and (C) are not exactly the same section. (D) Histological examination demonstrates that a cystic mass, with its wall mostly covered by the respiratory epithelium, consisted of variable sized islands of glial tissue (G, black arrows) separated with fibrovascular tissue. (E) Rosette formation of ependymal cells (black arrows) was noted. (H&E staining) (F) Sagittal view of T2WI on postoperative seventh day shows complete removal of the lesion (white arrow) but fails to reveal the connection.

panipenem/betamipron (160 mg/kg/day) was initiated for 19 days. The patient had good recovery from meningitis and was discharged without sequela.

OUTCOME AND FOLLOW-UP

A conservative follow-up was indicated after discussion of the multidisciplinary team, since the additional (or multiple) and radical surgery was thought to be too invasive for less than 1-year-old baby. Surgical excision of the recurrent mass and dural plasty will be scheduled in case of repeated meningitis. Evidence of CSF leak or meningitis was not observed after 22 months of follow-up.

DISCUSSION

At the development of delayed postoperative meningitis in this case, the stalk between the intracranial and extracranial spaces in the sphenoid bone was enhanced with Gd administration, indicating that meningitis had occurred through this connection. Although nasal glial heterotopia can be thought as cephaloceles that have lost their intracranial connection, a fibrous

stalk representing a remnant of the intracranial connection can be found in 15% of cases.^{5 6}

During the perioperative period, we could not be convinced that the connection with intracranial structures was present. Although preoperative T2WI equivocally shows a small strand in the sphenoid bone, the postoperative MRI failed to reveal the connection. Furthermore, neither CSF leakage nor bony defect was noted intraoperatively. One of the possible reasons is that the connection size was not adequate to be demonstrated on MRI.

Another possible pathomechanism is the unusual localisation of the connection in this case. Nasal glial heterotopia is speculated to result from brain herniation through the foramen caecum. Because the foramen caecum lies posterosuperior to the pneumatizing nasal processes of the frontal bone, being positioned between the frontal bone and crista galli, the most common site of nasal glial heterotopia and associated connection is the frontonasal (or glabellar) region.^{5 6} However, glial heterotopia was localised in the nasopharyngeal region in this case and was not attached in the frontonasal region. Furthermore, the connection was present in the transsphenoidal canal,

Figure 2 (A) Axial view of MRI with fluid-attenuated inversion recovery sequence with Gd administration reveals the meningeal enhancement of the convexity and interhemispheric surface of the bilateral frontal lobes. (B) Sagittal views of T1WI and (C) T2WI and (D) caudorostral serial coronal sections of T2WI clearly show the isointense stalk (white arrows) between the intracranial cerebrospinal fluid space and recurrent mass in the nasopharyngeal space (white dotted arrows). The stalk starts from the medial wall of the right optic nerve (the second nerve, II), runs through the transsphenoidal canal in the sphenoid bone, and terminates at the recurrent tumour. (E) Sagittal and (F) coronal views of the bone target CT scans, which correspond to the levels of (B,C) and (D), respectively, demonstrate the bony defect (transsphenoidal canal) in the sphenoid bone. (G) On coronal view of T1WI, with Gd administration, the stalk (white arrows) and recurrent tumour (white dotted arrows) are enhanced.

which is persistent craniopharyngeal canal.⁷ The nasopharynx (endodermal) and nasal cavity (ectodermal) are quite different areas embryologically, and that developmental abnormalities are likely to have different mechanistic details.⁸

Furthermore, nasopharyngeal glial heterotopia has fibrous cords extending to the dura similar to some nasal glial heterotopia.^{8,9} Extensive research of the literature reveals that only three similar cases presenting as transsphenoidal extension have been reported, none with complication of meningitis.^{3,9} Husein *et al* reported a case of a patient with a large glial heterotopia in the nasopharynx with similar bony defect in the sphenoid bone.³ The bony defect was covered with a mucosal flap during the surgery performed on the ninth day of life, and postoperative meningitis was not seen. Kau *et al* reported two cases of neuroglial heterotopia in the nasopharynx presenting as transsphenoidal extension of hypothalamic hamartoma through the sphenoid bone.¹⁰ Although the intracranial mass lesion was not demonstrated in our case, the neuroradiological features of the connection in this case were quite similar to those of the cases reported by Kau *et al*.¹⁰ Intracranial origin of the connection was in the right optic nerve, which was adjacent to the hypothalamic area. These findings suggest that the histology of the connection

is not a fibrous stalk, which is seen in the frontonasal region of nasal glial heterotopia, but heterotopia itself or hamartoma. The heterotopia could grow along with the child's growth,^{3,6,8} as in this case, although heterotopia is not a neoplasm. This idea could explain why the connection, which was barely recognised in the neonatal period, became obvious on conventional MRI and CT scan at the age of 9 months. This would also explain the delayed occurrence of meningitis.

Basal encephalocele, especially transsphenoidal encephalocele in this case, could not be completely differentiated from the nasopharyngeal heterotopia only from the histological findings.¹¹ However, transsphenoidal encephalocele is generally a serious malformation, and pituitary gland and hypothalamus might herniate through the large bony defect of the sphenoid bone.¹² It can be complicated with serious condition, such as facial abnormalities and optic nerve hypoplasia. In our case, no other malformation was complicated. Second, encephalocele cannot grow along with the child's growth. Third, the bony defect is not cranium bifidum, but the craniopharyngeal canal remnant.⁷ Thus, we prefer that the diagnosis of our case be termed as heterotopia rather than encephalocele.

In conclusion, the pathophysiology of nasopharyngeal glial heterotopia may be different from that of nasal glial heterotopia. The occurrence of delayed postoperative meningitis should be carefully observed with the combined use of MRI and CT scan,¹¹ even when the initial neuroradiological examination does not reveal the connection with the CSF space.

Learning points

- ▶ Glial heterotopia is an intranasal mass composed of mature brain tissue isolated from the cranial cavity, with frontonasal region as the most common site of development.
- ▶ Although some cases show fibrous stalks through bony defects, they lack communication to the CSF space.
- ▶ When the glial heterotopia is located in the nasopharyngeal (instead of nasofrontal) area, the occurrence of delayed postoperative meningitis, through intracranial and extracranial connection in the sphenoid bone, should be carefully observed, even in cases with the initial neuroradiological examination showing negative connection with the CSF space.

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