Ectopic Brain Tissue in a Child: A Case Report
A Case of Ectopic Brain Tissue in the Nasophaynx in Thailand

Vannipa Vathanophas, M.D., Archwin Tanphaichitr, M.D., Kitirat Ungkanont, M.D.

Department of Otorhinolaryngology, Faculty of Medicine Siriraj Hospital, Mahidol University, Bangkok 10700, Thailand

ABSTRACT

Brain heterotopia is a benign tumor composed of differentiated neural tissue that is located outside the cranial vault. This condition is uncommon and presents as a congenital pharyngeal mass. Here, we report a case of neuroepithelial heterotopia in the nasopharyngeal area of a six-month-old boy who presented with cleft palate and stridor. The tumor demonstrated aggressive growth with oropharyngeal involvement. Radiologic finding revealed a large heterogeneous enhancement on the left side of the nasopharynx, involving the uvula, left lateral pharyngeal wall, and left tonsil. No connection to the brain or spinal cord was apparent on imaging. Histologic features included presence of neuroglial heterotopias, composed predominately of glial cells in a surrounding neurofibrillary matrix. Surgery was the selected intervention, with wide excision performed via cleft palate. Previously published literature relevant to this case were reviewed and discussed. Recurrence is common in incomplete resection, although there was no evidence of recurrence at the two-year follow-up in this patient.

Keywords: Brain heterotopia; ectopic brain tissue; neuroglial heterotopias (Siriraj Med J 2017;69: 94-96)

INTRODUCTION

Brain heterotopia or differentiated neural tissue outside the cranial vault is an uncommon congenital tumor. This anomaly most commonly occurs in the nasal cavity and, erroneously, it is often identified as being a nasal glioma.[1] Heterotopic brain tissue is less commonly reported in other locations, including the pharynx, lung, orbits, palate, tongue, cheek, lip, and neck area.[2] Less than 30 cases of ectopic neuroglial tissue in head and neck locations have been reported in the English language literature. Here, we report a case of a six-month-old boy with heterotopic brain tissue in the pharyngeal area which was protruding through the cleft palate and which had no identifiable connection to the brain or spinal cord.

CASE REPORT

A six-month-old boy was admitted to the pediatric otorhinolaryngology ward of Siriraj Hospital with a large mass in his mouth. His mother reported progressive snoring and mild feeding difficulty. The patient had no history of facial or neck trauma. No neck stiffness or upper respiratory tract infection was observed in the days prior to presentation. His prenatal history was unremarkable. Physical examination revealed a mass that occupied the entire oropharynx and which obscured the view of the posterior pharyngeal wall. He also had cleft palate. The lesion was not seen through anterior rhinoscopy. Endoscopy was not done at the outpatient department. The mass was rubbery (not tender and fixed) and extended into part of the hypopharynx. Our

Correspondence to: Vannipa Vathanophas
E-mail: vannipa.vat@mahidol.ac.th
Received 10 June 2016 Revised 26 August 2016 Accepted 19 September 2016
doi:10.14456/smj.2017.18

patient also had occasional stridor and decreased left nasal airflow when he awoke. All other aspects of the physical examination were normal. Plain chest radiograph showed normal lung and normal cardiothoracic ratio. CT scan of the paranasal sinus and neck showed mixed solid and cystic soft tissue mass with heterogeneous enhancement on the left side of the nasopharynx that involved the uvula, left lateral pharyngeal wall, and left tonsil (Fig 1). The soft tissue mass, which measured approximately 1.4*3.6*3.8 cm (W*I*H), was suspected of being a soft tissue tumor. The tumor was further suspected of being either a venolymphatic malformation or a rhabdomyoma.

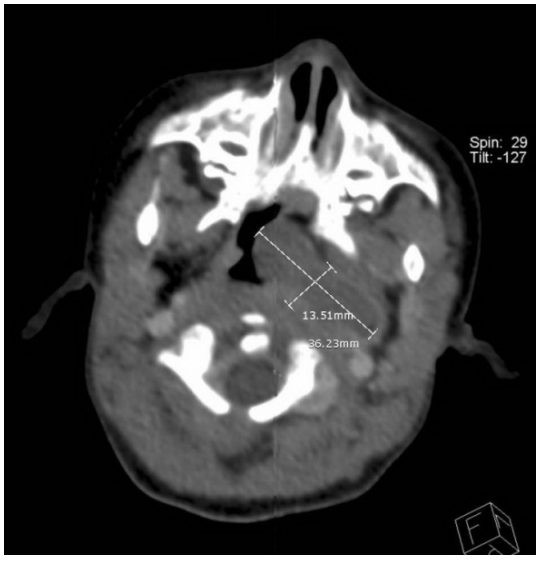

Fig 1. Axial CT scan showed a soft tissue mass in the left nasopharnx.

Transoral incisional biopsy was performed with prevention of airway obstruction by tracheotomy. During biopsy, a large mass filled with clear fluid was found attached to the surrounding tissue. Biopsy was done using No.15 surgical blade. Gross appearance showed a grayish mass with white cross-sectional surface. Histological examination showed a neuroglial heterotopia, composed predominantly of glial cells in a surrounding neurofibrillary matrix. The cleft in the lesion was lined with ependymal-like columnar cells and was surrounded by meninges, but with no malignancy. MRI was performed from the level of the brain to the oropharynx to evaluate intracranial extension and to plan for surgery. MRI imaging indicated that the mass was not in contact with the base of the skull. The patient then underwent total excision of the tumor via a transoral approach. The tumor was approximately 2*3.5*3.5 cm in size (Fig 2). It invaded the nasopharyngeal mucosa and adhered firmly to the left nasopharyngeal area. During the surgery, tissue surrounding the tumor was removed to ensure that no residual tumor cells remained. A McIvor mouth

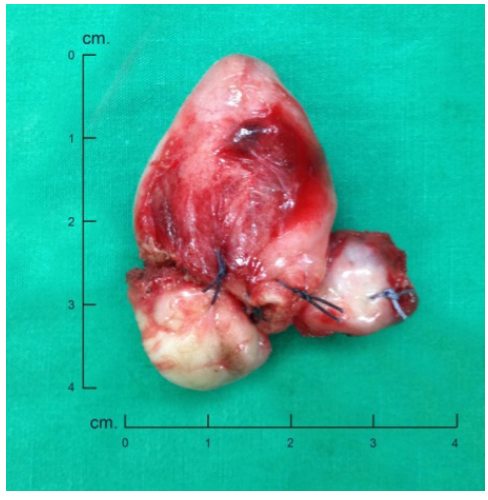

Fig 2. Gross finding of the whole tumor.

gag retractor was applied to open the mouth and to retract the base of the tongue. The soft palate was retracted by two malleable retractors for good exposure. A monopolar electrode with ultra-sharp tungsten tip was used for precise and bloodless tumor dissection. The residual tumor was evaluated by a telescope via nose and mouth. There were no post-operative complications and the patient was discharged in good health on the fifth day. The final pathology of the excised tumor was the same as that of the biopsy. At the one-year follow-up, a fibrous mass of 1 cm in diameter was found in the same area. An additional surgery was performed, with no finding of neuroglial heterotopia in the pathology report. The patient underwent physical examination again at one year after the last operation, with no recurrence or complication being observed. Palatoplasty and decannulation were then performed, respectively.

DISCUSSION

Heterotopic neuroglial tissue (HNT) was first described by Reid in 1852.[3] Although our patient was a boy, neuroglial heterotopia has been shown to be more prevalent in females.[4,5] Prominent symptoms described in the literature are airway obstruction, feeding difficulty, and/or neck mass6, and these symptoms are usually initially seen in the newborn period. Heterotopic brain tissue is mostly seen in the nasal cavity, being only infrequently found in the nasopharyngeal area. The majority of patients with HNT are products of uncomplicated pregnancies. Other associated developmental anomalies have been reported, including cleft palate, micrognathia, choanal atresia, and congenital heart disease.[5,7] Our patient presented with cleft palate.

In 1946, Bratton reported the first case of HNT in the nasopharyngeal area. Uemura, *et al.*, reported a case of heterotopic nasopharyngeal brain tissue associated

with cleft palate and reviewed 17 published cases, which revealed a 44% occurrence rate of heterotopic nasopharyngeal brain tissue found in patients with cleft palate.[8] From this study, they hypothesized that the presence of HNT directly influences the nonaligning of the palate during prenatal development. In 2006, Al-Ammar reported on a preterm baby with midline nasopharyngeal HNT, and this was the last case reported in the literature before this case report.[1] Our patient is the first case of ectopic brain tissue in the nasophaynx to be reported in Thailand. The pathogenesis of HNT is unclear. One commonly accepted theory is that HNT is a variant of encephalocele, in which the central nervous system connection has either been absorbed or has become vestigial.[1] Radiologic imaging is necessary in preoperative planning to determine the extent and location of the mass and to determine if there is any connection to the brain. In addition to plain radiography, our patient was also evaluated by CT and MRI imaging. The mass was seen to be predominantly solid with a small cystic component, which was similar to other previous reports.[8]

Histologically, the gross appearance of brain heterotopia is firm and being dark brown or red in color. The tissue may also have cystic components containing clear cerebrospinal fluid4. In our patient, microscopic evaluation found lobulated glial tissue surrounded by benign non-keratinizing squamous mucosa. The glial tissue had a fine capillary network and contained a large number of neural/glial cells. Choroid plexus structures and ependymal cells and clefts were occasionally seen.[5] There was no evidence of immature tissue or components to indicate the presence of a teratoma.[3]

Differential diagnosis of brain heterotopia consists of mature brain tissue, including encephalocele, gliomas, and teratoma. Encephalocele has the exact same microscopic profile as brain heterotopia, but encephalocele has a connection with the cranial cavity or spinal canal and heterotopic brain tissue does not. Glioma is a congenital extracervical and extracranial tumor made of glial and fibrous tissue. However, gliomas do not contain choroid plexus structures, and this is the main histological difference that differentiates them from heterotopic neuroglial tissue.[2] Teratoma, however, consists of all three germ layers, while heterotopic brain tissue is composed solely of ectodermal elements.[4]

Surgical resection is the treatment of choice in ectopic brain tumor. Considering the aggressive growth of this tumor and the potential for encroachment to vital structures – especially the airway, adequate resection was recommended. Surgical excision can be performed via many approaches, depending on the location of the mass. The aim of treatment is early identification and preservation of vital neurovascular structures. In this case, our patient's airway was controlled by tracheostomy to relieve failure to thrive and to prevent airway obstruction. Excision of the tumor was performed via cleft palate when his condition improved.

Recurrence of brain heterotopia as a result of incomplete excision has been reported.[9] Malignant transformation is rare, although close observation and frequent follow-up is strongly recommended. No recurrence of brain heterotopia was observed in our patient at the 2-year follow-up.

CONCLUSION

This case report of ectopic brain tissue in the nasopharyngeal area profiles a six-month-old boy who presented with cleft palate and partial airway obstruction. Wide surgical resection yielded a satisfactory result. Imaging studies and histologic examination were the necessary investigations for diagnosis. Although this tumor was benign in nature, it could cause airway obstruction in a newborn. Focal recurrence of the residual lesion was previously reported, but no recurrence was observed in this patient at 2-year follow-up. Postoperative care and long-term follow-up with focused attention on the airway are essential.

REFERENCES

1. Al-Ammar AY, Al Noumas HS, Alqahtani M. A midline nasopharyngeal heterotopic neuroglial tissue. J Laryngol Otol 2006;120:E25.
2. Hendrickson M, Faye-Petersen O, Johnson DG. Cystic and solid heterotopic brain in the face and neck: a review and report of an unusual case. J Pediatr Surg 1990;25:766-8.
3. Reid F. Uber angeborene Hirnbrucke in den Stirn und Nasengegend. Illustrierte Med Ztg 1852;1:133-41.
4. Broniatowski M, Witt WJ, Shah AC, Galloway PG, Abramowsky CR. Glial tissue in the parapharyngeal space. Arch Otolaryngol 1981;107:638-41.
5. Madjidi A, Couly G. Heterotopic neuroglial tissue of the face. Report of six cases and review of the literature. Oral Surg Oral Med Oral Pathol 1993;76:284-8.
6. Aboud MJ. Respiratory difficulty caused by an ectopic brain tissue mass in the neck of a two-month-old baby: a case report. J Med Case Rep 2011;5:220.
7. Forte V, Friedberg J, Thorner P, Park A. Heterotopic brain in the parapharyngeal space. Int J Pediatr Otorhinolaryngol 1996;37:253-60.
8. Uemura T, Yoshikawa A, Onizuka T, Hayashi T. Heterotopic nasopharyngeal brain tissue associated with cleft palate. Cleft Palate Craniofac J 1999;36:248-51.
9. Behar PM, Muller S, Gerber ME, Todd NW. Heterotopic neuroglial tissue causing airway obstruction in the newborn. Arch Otolaryngol Head Neck Surg 2001;127:997-1002.