AJNR

Heterotopic brain in the pterygopalatine fossa.

J E Kallman, L A Loevner, D M Yousem, A A Chalian, D C Lanza, L Jin and R E Hayden

AJNR Am J Neuroradiol 1997, 18 (1) 176-179
http://www.ajnr.org/content/18/1/176

Heterotopic Brain in the Pterygopalatine Fossa

James E. Kallman, Laurie A. Loevner, David M. Yousem, Ara A. Chalian, Donald C. Lanza, Li Jin, and Richard E. Hayden

Summary: Heterotopic brain outside the cranial vault is uncommon. It occurs most frequently in the nasal region, although rests elsewhere in the aerodigestive tract have been reported. We describe a case of heterotopic brain in the pterygopalatine fossa.

Index terms: Brain, abnormalities and anomalies; Pterygopalatine fossa

Heterotopic brain, or differentiated neural tissue outside the cranial vault, is uncommon, occuring most commonly about the nose. Other reported sites of heterotopic brain include the scalp (1), orbit (2), lip (3), tongue (4), palate (5, 6), pharynx (7–9), and lung (10). Heterotopic brain involving the nose, face, or scalp is frequently detected early in life at physical examination; however, heterotopic brain located deep within the head and neck is often asymptomatic and, hence, when detected, is usually an incidental finding on radiologic examinations performed for unrelated reasons. We describe a patient with heterotopic brain in the pterygopalatine fossa. Computed tomography (CT) and magnetic resonance (MR) imaging were useful not only in identifying the heterotopic brain but also in excluding associated cranial defects and direct communication with the brain, thereby distinguishing it from an encephalocele.

Case Report

A 47-year-old woman had a 3-month history of sensory symptoms in the distribution of the third division of the left fifth cranial nerve (V_3), including left-sided facial numbness involving the skin overlying the mandible, the corner of the mouth, the lower lip, and the undersurface of the tongue. Her medical history was otherwise unremarkable. She was treated with a 2-week trial of oral steroids, which resulted in temporary relief of her symptoms, although they ultimately recurred. The patient was referred to our institution for further evaluation.

Physical examination was remarkable only for reduced sensation to pinprick and light touch over the left side of the face, including the skin overlying the mandible, tongue, floor of mouth, and hard palate. There were no other focal neurologic abnormalities. Findings at nasal endoscopy were normal.

Radiologic examination included CT and MR imaging. While no abnormalities were noted to explain the patient's symptoms in the left V_3 distribution, a soft-tissue mass was incidentally detected in an expanded right pterygopalatine fossa. There was also osseous remodeling of the undersurface of the inferomedial sphenoid bone as well as the foramen rotundum, suggesting a long-standing process. No communication with the brain and no osseous defect of the skull base were present (Fig 1A–G). The lesion enhanced slightly on postcontrast MR images and had signal intensity characteristics consistent with neuronal tissue. A schwannoma of the fifth cranial nerve in the pterygopalatine fossa was suspected.

Fine-needle aspiration of the mass using CT guidance suggested neuronal tissue, but was nondiagnostic. An open biopsy through a transmaxillary approach showed glial tissue. Final histologic analysis, which included stains for neurofilament and glial fibrillary acidic protein, confirmed the presence of neurons, reactive glial cells, and fibrovascular tissue without malignant features, consistent with the diagnosis of heterotopic brain tissue (Fig 1H).

Discussion

Heterotopic brain is a rare clinical entity that was reported by Reid in 1852 (11). It has been described in various sites of the head and neck, most commonly in the nasal region, where it has been referred to as a "nasal glioma." It may occur within or outside the nasal cavity, the latter being more common. The term *glioma* was used by Schmidt in 1900 (12), but it is a misnomer, as these lesions are not neoplastic

Received October 25, 1995; accepted after revision March 5, 1996.

From the Departments of Radiology (L.A.L., D.M.Y.), Otorhinolaryngology, Head and Neck Surgery (A.A.C., D.C.L., R.E.H., D.M.Y.), Pathology and Laboratory Medicine (L.J.), and the University of Pennsylvania School of Medicine (J.E.K.), Hospital of the University of Pennsylvania, Philadelphia.

Address reprint requests to L. A. Loevner, MD, Department of Radiology, Neuroradiology Section, Hospital of the University of Pennsylvania, 3400 Spruce St, Philadelphia, PA 19104.

AJNR 18:176–179, Jan 1997 0195-6108/97/1801–0176

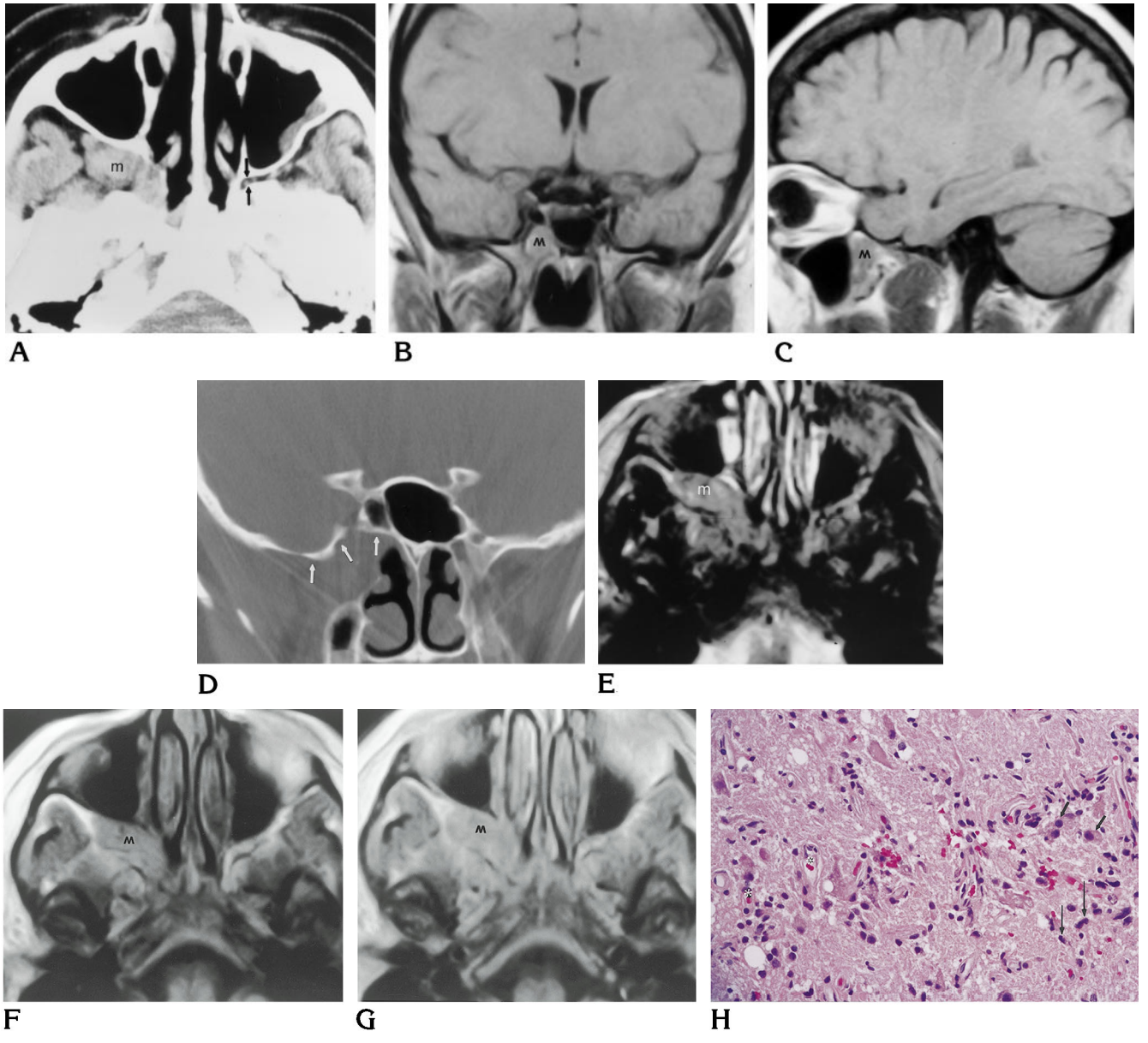

Fig 1. Forty-seven-year-old woman with heterotopic brain tissue in the pterygopalatine fossa.

A, Axial CT scan shows a well-demarcated soft-tissue mass (*m*) in the right pterygopalatine fossa with secondary expansion of the fossa as compared with the normal left side (*arrows*).

B and *C*, Coronal (*B*) and sagittal (*C*) unenhanced T1-weighted MR images show the mass (*M*) in the pterygopalatine fossa with maintenance of the integrity of the floor of the middle cranial fossa both inferiorly and medially. The mass is not in contiguity with the temporal lobe. The foramen rotundum is secondarily expanded.

D, Coronal CT scan photographed for bone detail shows the mass in the enlarged right pterygopalatine fossa with osseous remodeling of the undersurface of the inferomedial sphenoid bone and foramen rotundum (*arrows*). Serial thin-section coronal images (not shown) revealed no osseous defect of the skull base, including Meckel's cave and the cavernous sinus, and no communication with the brain.

E, Axial T2-weighted MR image shows that the mass (*m*) is essentially isointense with the cerebellum.

F and *G*, Unenhanced and enhanced T1-weighted images, respectively, with similar photography show mild homogeneous enhancement of the pterygopalatine fossa mass (*M*).

H, Histologic section after open biopsy of the pterygopalatine fossa mass shows an admixture of neurons (*short arrows*), reactive glial cells (*long arrows*), and fibrovascular tissue (*asterisks*). Neural and glial elements stained positively with immunohistochemical stains for neurofilament and glial fibrillary acidic protein, respectively. These findings are diagnostic of heterotopic central nervous system tissue.

(13). Over 140 cases of nasal heterotopic brain have been described in the literature (7, 13, 14). Less common sites for heterotopic brain include the scalp (1), orbit (2), lip (3), tongue (4), soft and hard palates (5, 6), nasopharynx and oropharynx (7–9), and lung (10).

Heterotopic brain may present at any age, but it is frequently diagnosed in infancy, particularly when it involves superficial facial tissues such as the nose, eye, or lip, allowing early clinical detection. Heterotopic brain usually consists of neuroglial elements, but it may contain elements of choroid plexus (14). As a rule, growth of heterotopic brain parallels growth of normal tissue, except in cases in which the lesion is cystic owing to the presence of functional choroid plexus (14). Patients with cystic lesions may have gross facial deformities (14, 15) or, when the airway is involved, acute respiratory distress (6, 9, 10). Associated developmental anomalies have been reported to include bifid nose, cleft lip, and cleft palate (8–10). Frequently, heterotopic brain is an incidental finding, discovered on radiologic examinations performed for unrelated reasons, such as in our case.

Three theories have been advanced regarding the pathogenesis of heterotopic brain tissue. First, heterotopic brain may derive from an encephalocele that subsequently loses its communication with the brain (8, 9). Formation of the skull base begins in the embryo at approximately 6 weeks' gestational age, when the parachordal cartilages first appear along the cranial notochord. Subsequently, the hypophyseal cartilages grow around the pituitary gland and fuse laterally with the alisphenoid to form the sphenoid bone, while the prechordal cartilages (trabeculae cranii) give rise to the ethmoid bone. These cartilages usually fuse by the 12th gestational week to form the chondrocranium or early skull base (16). It has been hypothesized that tissue protruding through the basal skull sutures may become separated from the developing brain after closure of the sutures (16, 17). This theory explains nasal heterotopic brain tissue and is supported by the observation that 25% of lesions in the nasal region retain a fibrous, extradural connection to the central nervous system (9). The origin of heterotopic brain at sites other than the nasal region, however, is less clear. An alternative theory suggests that ectopic brain in remote locations may result from separation of extracranial embryonic neural tissue, independent of cranial suture closure (12). Finally, a third hypothesis suggests that heterotopic brain derives from isolated rests of displaced pluripotential neuroectodermal cells that subsequently differentiate into mature neural tissue (9, 12).

The pathogenesis of distal heterotopic brain in the lung is uncertain. Systemic embolization of brain tissue following intrauterine trauma or aspiration of brain tissue fragments from the amniotic fluid are two hypotheses proposed to explain these lesions (9, 10).

The differential diagnosis of a pterygopalatine fossa mass typically includes perineural extension of tumors along the second division of the trigeminal nerve, nerve sheath tumors (schwannomas, neurofibromas), angiofibromas, hemangiomas, and, rarely, ectopic lesions of minor salivary glands (7). CT and MR imaging are useful in the evaluation of masses involving the skull base. CT provides information about the adjacent osseous structures while MR imaging, because of its improved soft-tissue resolution, helps to characterize the soft-tissue abnormality. Angiofibromas and hemangiomas are vascular tumors that enhance avidly after contrast administration and may show an abundance of flow voids on MR images. Nasopharyngeal carcinoma involving the pterygopalatine fossa will show replacement of fat in this location and, if extensive, may result in expansion of the fossa. Schwannomas are circumscribed, enhancing tumors that, when large enough, exhibit remodeling and expansion of the adjacent osseous margins of the pterygopalatine fossa (18).

Both encephaloceles and heterotopic brain may have signal characteristics similar to brain on all MR pulse sequences (T1-, proton density–, and T2-weighted). The two entities may be distinguished from one another in that encephaloceles usually retain a visible connection with the brain. Heterotopic brain or neuronal tissue comprising an encephalocele may be hyperintense on T2-weighted images because of dysplastic neural tissue (5, 18). Heterotopic brain containing functioning choroid plexus elements may present as a cystic mass owing to production of cerebrospinal fluid (10, 14, 15). It may enhance depending on the vascularity or presence of choroid plexus. CT is useful for identifying small bone defects at the skull base, whereas MR imaging, with its multiplanar capabilities, is excellent for identifying communication with the adjacent brain.

Pathologically, heterotopic brain displays a

variety of central nervous system elements. Commonly identified components include astrocytes, oligodendroglia, and neurons. Ependyma, retinal components, and choroid plexus elements are seen less frequently (9). The neural elements are typically embedded in fibrous mesenchymal tissue and may exhibit calcification. Rare instances of neoplasm occurring within excised lesions have been reported, including oligodendroglioma (19) and neuroectodermal tumor (20). The absence of many components—particularly neurons, choroid plexus, and ependyma—from nasal heterotopic lesions has led some authors to view them as separate entities (19). However, others argue that because neuronal precursors do not appear in the developing brain until the 10th gestational week, embryonic brain tissue that separates before that time may be relatively ischemic, giving rise to neuron-poor lesions (17).

Because heterotopic brain is often asymptomatic, treatment is usually conservative. Surgical excision may be performed for symptomatic lesions, such as those obstructing the airway, or for lesions resulting in cosmetic deformities, such as those involving the midline facial region. Excision is curative and recurrence is rare (2).

References

1. Zook EG, Nickey WM, Pribaz JJ. Heterotopic brain tissue in the scalp. *Plast Reconstr Surg* 1984;73:660–663
2. Elder JE, Chow CW, Holmes AD. Heterotopic brain tissue in the orbit: case report. *Br J Ophthalmol* 1989;73:928–931
3. Pasyk KA, Agenta LC, Marks MW, Friedman RJ. Heterotopic brain presenting as a lip lesion. *Cleft Palate J* 1988;25:48–52
4. Knox R, Pratt M, Garvin AJ, White B. Heterotopic lingual brain in the newborn. *Arch Otolaryngol Head Neck Surg* 1989;115:630–632
5. Fuse T, Aoyagi M, Ota N, Koike Y, Yuda F. Heterotopic brain tissue of the soft palate. *ORL J Otorhinolaryngol Relat Spec* 1992;54:54–56
6. Al-Nafussi A, Hancock K, Sommerlad B, Carder PJ. Heterotopic brain presenting as a cystic mass of the palate. *Histopathology* 1990;17:81–84
7. Feldman BA, Schwartz RH, Chandra R, Anderson K. Heterotopic brain tissue simulating a neonatal tonsil. *Clin Pediatr* 1982;7:428–430
8. Okulski EG, Biemer JJ, Alonso WA. Heterotopic pharyngeal brain. *Arch Otolaryngol* 1981;107:385–386
9. Cohen AH, Abt AB. An unusual case of neonatal respiratory obstruction: heterotopic pharyngeal brain tissue. *J Pediatr* 1970;76:119–122
10. Fuller C, Gibbs A. Heterotopic brain tissue in the lung causing acute respiratory distress in an infant. *Thorax* 1989;44:1045–1046
11. Reid F. Über angeborene Hirnbrucke in den Stirn und Nasengegend. *Illustr Med Ztg* 1852;1:133–141
12. Schmidt MB. Über seltene spaltbildungen in Bereiche des mittleren Stirnforsatzes. *Arch Pathol Anat* 1900;162:340
13. Azumi N, Matsuno T, Tateyama M, Inoue K. So-called nasal glioma. *Acta Pathol Jpn* 1984;34:215–220
14. 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;25:766–768
15. Wismer G, Wilkinson AH, Goldstein JD. Cystic temporofacial brain heterotopia. *AJNR Am J Neuroradiol* 1989;10:S32–S33
16. Larsen WJ. *Human Embryology*. New York, NY: Churchill-Livingstone; 1993
17. Genut AA, Miranda FG, Garcia HJ. Organized heterotopia in the ethmoid sinus. *J Neurosci* 1976;28:339–344
18. Grossman RI, Yousem DM. *Neuroradiology: The Requisites*. St Louis, Mo: Mosby; 1994
19. Bossen EH, Hudson WR. Oligodendroglioma arising in heterotopic brain tissue of the soft palate and nasopharynx. *Am J Surg Pathol* 1987;11:571–574
20. Lee SC, Henry MM, Gonzalez-Crussi F. Simultaneous occurrence of melanotic neuroectodermal tumor and brain heterotopic in the oropharynx. *Cancer* 1967;38:249–253