AJNR

Extracerebral intracranial glioneural hamartoma with extension into the parapharyngeal space.

J D Moritz, D Emons, O D Wiestler, L Solymosi, S Kowalewski and M Reiser

AJNR Am J Neuroradiol 1995, 16 (6) 1279-1281
http://www.ajnr.org/content/16/6/1279

Extracerebral Intracranial Glioneural Hamartoma with Extension into the Parapharyngeal Space

Jörg D. Moritz, Dieter Emons, Otmar D. Wiestler, László Solymosi, Sabina Kowalewski, and Maximilian Reiser

Summary: A newborn had an extracerebral, intracranial mass extending from the right middle cranial fossa through the base of the skull to the parapharyngeal space. The mass was predominantly composed of immature brain tissue. It was enclosed by its own leptomeninges and dura and was classified as a glioneural hamartoma.

Index terms: Hamartoma; Infants, neoplasms

In an extracerebral location, hamartomas of central nervous system (CNS) tissue are very uncommon (1, 2). Intracerebral hamartomas are found in the cortex and white matter (3, 4). Extracerebral CNS hamartomas are located intracranially within leptomeninges and dura (5–8). Occasionally, hamartomas are found in extracranial locations, such as in the nasal fossae, soft palate, pharynx, and the oral cavity (9, 10), or in the leptomeninges of the spine (11). We describe an extremely rare form of an extracerebral intracranial neuroglial hamartoma that extended into the extracranial space.

Case Report

In this case, polyhydramnious was recognized in the 32nd week of gestation, and an intracranial mass was depicted by fetal ultrasound. After delivery, bulging of the skull in the right temporal area and of the right side of the neck was evident. A bone defect was palpable in the area of the right temporal squama, and the right external ear was displaced caudally. There were signs of a right seventh, sixth, and third nerve palsy. The neurologic and physical findings were otherwise normal. Plain film radiography of the skull disclosed deformation of the middle cranial fossa, bulging of the right parietotemporal skull, and considerable diastasis of the right parietotemporal suture. Ultrasound of the brain (Fig 1A) and computed tomography (CT) (Fig 1B) revealed an intracranial tissue mass in the right temporal region. The right temporal lobe was hypoplastic. The moderately enlarged ventricles were not displaced. No enhancement within the mass was found on CT. On magnetic resonance (MR) (Fig 1C and D) the mass was clearly separated from the brain. Angiography demonstrated that the mass was supplied by branches of the external carotid artery and by dural branches of the internal carotid artery. No hypervascularity or pathologic vessels were seen. CT and ultrasound findings led us to consider a glial neoplasm and manifestations of phacomatosis as differential diagnoses. However, other stigmata of phacomatosis and a family history were missing. The MR studies made the diagnosis of a glial neoplasm less likely. Various features of this lesion were considered consistent with a teratoma; however, the extension and shape of the mass made a teratoma unlikely.

When the infant was 22 days of age, surgery was performed for increasing stridor and dyspnea. Neural tissue enclosed by dura was found within the mass. At the base of the skull, the mass extended through a bone defect to the parapharyngeal space, where the mass was no longer surrounded by dural tissue, and a clear demarcation was not possible. The intracranial portion of the mass was completely removed, whereas in the parapharyngeal region radical resection was not possible.

Histopathologic examination (Fig 1E) revealed a mass of neuroectodermal tissue covered by a fibrous, presumably leptomeningeal, membrane. Immunohistochemical reactions with antibodies to synaptophysin and neurofilament protein demonstrated advanced neuronal differentiation of these structures. Astrocytes were readily detectable with an antibody to glial fibrillary acidic protein. The neuropathologic findings prompted the diagnosis of a glioneural hamartoma.

Discussion

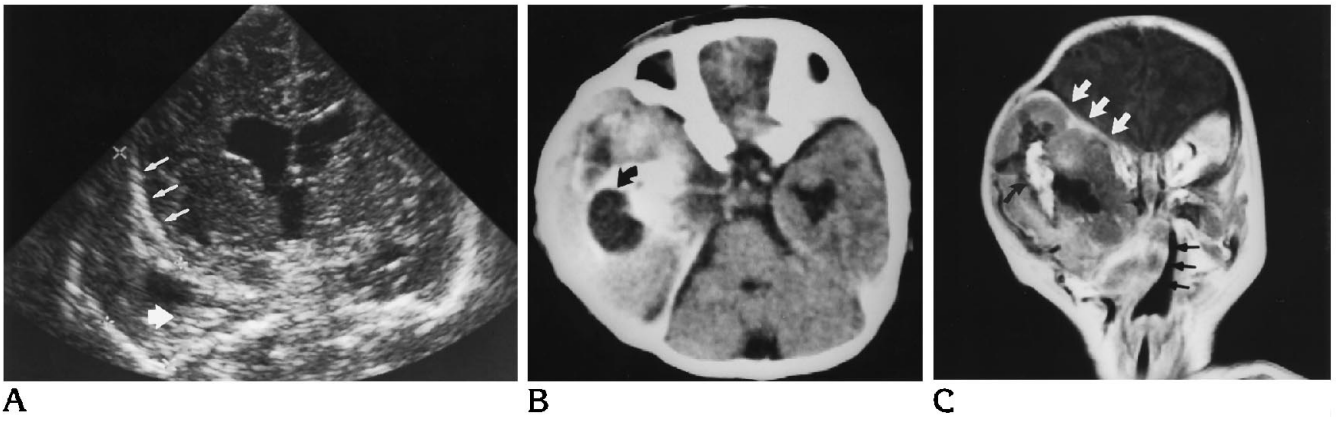
We found five cases of extracerebral glioneural hamartomas in the literature (5–8, 12). The term *glioneural hamartoma* designates tumorlike but nonneoplastic malformed mature tissue of neuroglial origin. The extent of differentiation

Received June 16, 1993; accepted after revision September 8, 1994.

From the Radiologische Universitätsklinik (J.D.M., L.S., M.R.); Universitätskinderklinik (D.E., S.K.); and Institut für Neuropathologie, Universitätskliniken Bonn (O.D.W.); Bonn, Germany.

Address reprint requests to Dr Jörg D. Moritz, Zentrum Radiologie, Abteilung Röntgendiagnostik I, Georg-August-Universität Göttingen, Robert-Koch-Strasse 40, 37075 Göttingen, Germany.

AJNR 16:1279–1281, Jun 1995 0195-6108/95/1606–1279



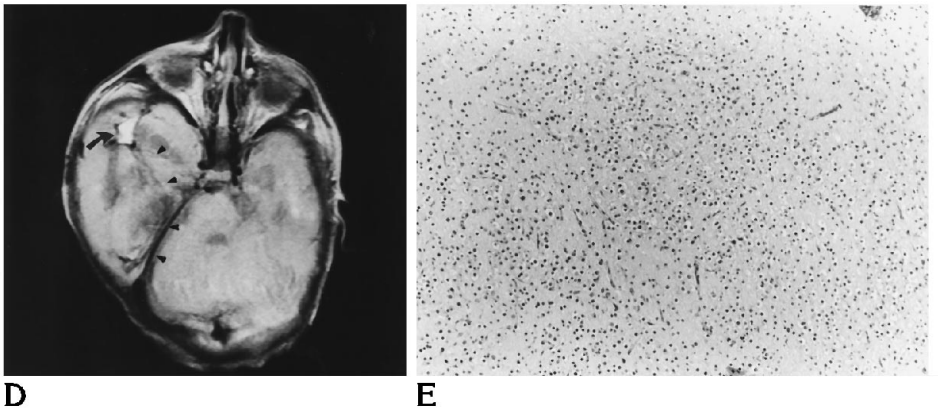


Fig 1. *A*, Coronal ultrasound of the brain shows an intracranial extracerebral mass in the right temporal region, marked by crosses (*x*). Separation of the mass from the cerebrum is outlined by *thin arrows*. The echogenicity of the mass is similar to normal cerebral tissue. An inhomogenous echogenic and cystic structure (*thick arrow*) can be delineated at the floor of the middle cranial fossa. Medial and cranial displacement of the temporal lobe by the mass can be seen. A clear-cut differentiation of the mass from the surrounding cerebral tissue is possible.

B, The contrast-enhanced axial CT scan demonstrates the mass within a considerably enlarged and deformed right middle cranial fossa. It shows a calcified area adjacent to a cyst (*curved arrow*).

C, T1-weighted coronal MR image (1.5 T, spin-echo sequence, 587/15/2 [repetition time/echo time/excitations]) and *D*, proton density–weighted axial MR image (1.5 T, spin-echo sequence, 3000/20) show the mass located in the right temporal region. It extends through a defect at the base of the skull to the oropharynx, which is compressed on the right side (*black arrows*). Signal intensities indicative of high fat content are detected on T1- and proton density–weighted images (*curved arrow*). There was no compression or infiltration of the right subarachnoidal space. Clear separation of the mass from the surrounding brain tissue was possible (*white arrows*).

E, Histopathologic appearance of the hamartoma. The lesion is composed of neural tissue with properties of an immature brain. Signs of neoplastic growth such as increased mitotic activity, vascular endothelial proliferation, diffuse infiltration, or a neuroblastic cellular component are absent.

of the cellular elements is variable. Significant mitotic activity and atypical tumorlike vasculature have never been identified in these structures, which are generally well separated from the surrounding tissue by a thin fibrous capsule (7), leptomeninges, or dura (6, 8) (Fig 1A, C, and D).

Among these reported cases there was only one of extracerebral, intracranial hamartoma with extension to the oropharynx (8). In contrast to our report, the pharyngeal portion showed a clear demarcation from the surrounding tissues. Others have found heterotopic brain tissue in the pharynx without communication to the intracranial space (9, 10); however, extension of an intracranial mass to the parapharyngeal space is not uncommon in encephalomeningoceles (13, 14). In our case, the clear-cut demarcation of the lesion from the adjacent temporal lobe and the solid nature of the tissue mass virtually excluded a temporal encephalocele. Nasal glioma was excluded because of the prominent neuronal and neuropil component of the tissue.

The pathogenesis of extracerebral glioneural hamartomas has not been resolved (1, 2). The two mechanisms to be considered are: (*a*) formation of an aberrant temporal lobe precursor in the early embryonic brain and (*b*) separation from the temporal brain primordium and independent development of a neural tube fragment during an early stage of neurogenesis. Protrusions of "mature" brain through preexisting pial defects have been postulated (2, 16). Another

possibility is an aberrant migration of embryonic neuroepithelial tissue (7, 15, 17), as it is discussed for the development of intracerebral heterotopias (3, 4). This theory involves the concept of a choristoma (18). Aberrant migration and heterotopia were reported after methylmercury poisoning during fetal life (19).

In conclusion, we have presented the case of a large glioneural hamartoma of the middle cranial fossa with marked extension into the parapharyngeal compartment. This lesion should be added to the differential diagnosis of intracranial tumors in newborn infants.

References

1. Brun A. Marginal glioneural heterotopias of the central nervous system. *Acta Pathol Mikrobiol Immunol Scand* 1965;65:221–233
2. Cooper IS, Kernohan JW. Heterotopic glial nests in the subarachnoid space: histopathologic characteristics, mode of origin and relation to meningeal gliomas. *J Neuropathol Exp Neurol* 1951; 10:16–29
3. Dunn V, Mock T, Bell WE, Smith W. Detection of heterotopic gray matter in children by magnetic resonance imaging. *Magn Reson Imaging* 1986;4:33–39
4. Takashima S, Chan F, Becker LE, Kuruta H. Aberrant neuronal development in hemimegalencephaly: immunohistochemical and Golgi studies. *Pediatr Neurol* 1991;7:275–280
5. Ball RY, Treip CS. Intracranial extracerebral neuroglial hamartoma. *Acta Neuropathol (Berl)* 1974;65:172–179
6. Farhat SM, Hudson JS. Extracerebral brain heterotopia: case report. *J Neurosurg* 1969;30:190–194
7. Nishio S, Mizuno J, Barrow DL, Takei Y, O'Brien MS. Intracranial extracerebral glioneural heterotopia. *Childs Nerv Syst* 1988; 4:244–248
8. Wakai S, Nakamura K, Arai T, Nagai M. Extracerebral neural tissue mass in the middle cranial fossa extending into the oropharynx in a neonate: case report. *J Neurosurg* 1983;59:692–696
9. Broniatowski M, Witt WJ, Shah AC, Galloway PG, Abramowsky CR. Glial tissue in the parapharyngeal space. *Arch Otolaryngol* 1981;107:638–641
10. Low NL, Scheinberg L, Andersen DH. Brain tissue in the nose and throat. *Pediatrics* 1956;18:254–259
11. Popoff N, Feigin I. Heterotopic central nervous tissue in subarachnoid space. *Arch Pathol* 1964;78:533–539
12. Marubayashi T, Matsukado Y. Intracranial extracerebral brain heterotopia: case report. *J Neurosurg* 1978;48:470–474
13. Suwanwela C, Suwanwela N. A morphological classification of sincipital encephalomeningoceles. *J Neurosurg* 1972;36: 201–211
14. Tranmer BI, Singh S, Ketch L. An unusual case of temporal encephalocele. *Childs Nerv Syst* 1989;5:371–373
15. Bailey OT. Relation of glioma of the leptomeninges to neuroglia nests: report of a case of astrocytoma of the leptomeninges. *Arch Path* 1936;21:584–601
16. Work WP, Hybels RL. A study of tumors of the parapharyngeal space. *Laryngoscope* 1974;84:1748–1755
17. Baily P, Bucy PC. Origin and nature of meningeal tumors. *Am J Cancer* 1931;15:15–35
18. Cohen AH, Abt AB. An unusual cause of neonatal respiratory obstruction: heterotopic pharyngeal brain tissue. *J Pediatr* 1970; 76:119–122
19. Choi BH, Lapham LW, Amin-zaki L, Saleem T. Abnormal neuronal migration, deranged cerebral cortical organization, and diffuse white matter astrocytosis of human fetal brain: a major effect of methylmercury poisoning in utero. *J Neuropathol Exp Neurol* 1978;37:719–733