Cerebral heterotopia of the temporofacial region

Case report

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The authors report a case of cerebral heterotopia in the right temporofacial region of a neonate. The lesion presented as a massive cystic swelling containing cerebrospinal fluid. It produced calvarial and facial bone deformities. After the mass was excised, histological examination revealed glial tissue and choroid plexus.

Key Words • heterotopia • cystic brain tumor • temporofacial tumor

EXTRACRANIAL subcutaneous glial heterotopias are rare ectopic rests of neural origin and are found at the root of the nose. They are similar in origin to nasofrontal encephaloceles but differ from them in lacking any patent connection with the central nervous system (CNS). Likewise, glial heterotopia in the temporofacial region and temporal encephalocele are similar in origin, but the former differs from the latter in lacking any patent CNS communication. The prognosis for these patients is excellent. Therefore, once the lesion is recognized, every effort should be made to remove it. To our knowledge, cerebral heterotopia with cyst formation has been described in only five previous reports. In only one report was the lesion wholly extracranial and situated in the temporofacial region. We report another such case and emphasize its pathogenesis.

Case Report

This baby girl was delivered at full term by lower-segment cesarean section following a failed vaginal extraction. She had a massive swelling on the right side of her head and face for which she was referred to us 4 days after her birth. Her Apgar score was 8 at 1 minute and 10 at 5 minutes after birth.

Examination. Examination revealed a well-developed and healthy neonate with a body weight of 4.7 kg. There was an 11 × 11-cm mass on the right side of her head and face, extending downward to the base of the neck. The mass caused outward traction on the right eyelids. The angle of her mouth on the right was drawn downward and outward. The right ear was displaced posteriorly. The lesion was soft, fluctuant, translucent, and did not adhere to the overlying skin. There was no pulsation and there was no change in size when the child cried. The mass could not be moved freely over the skull and the facial bones. The anterior fontanelle was flat and soft. The general physical and neurological examination revealed no other abnormality. Routine hematomatological and biochemical studies were normal. The x-ray films of the skull showed normal sutures, right-sided calvarial and facial deformities, and an extensive extracranial soft-tissue shadow.

Operation. The mass was exposed through a vertical incision. The sac could be dissected free from the skin. It was opened at its fundus and 700 ml of straw-colored liquid was evacuated. On opening the sac further, loculated cysts of varying sizes were seen in the infratemporal fossa. These were evacuated. The sac was then dissected down to its base at the cranial bone to which it was adherent. It was excised, keeping the adherent base in situ. There was no intracranial communication.

Histological Studies. The surgical specimen was soft and grayish. It measured 10 × 6 × 1 cm. Histological analysis showed brain tissue with meningeal structure lined by choroid plexus and surrounded by a vascularized fibrous tissue (Fig. 2).

Biochemical Examination. The cyst fluid was composed of 290 μg/dl protein (normal range 15–45 μg/dl), 76 μg/
Cerebral heterotopia

Fig. 1. *Left:* Computerized tomography scan revealing a large right-sided temporofacial swelling, compression of the frontotemporal bones and the lateral wall of the orbit, absence of zygoma, and probable site of origin of heterotopia. *Right:* Computerized tomography scan at a lower level demonstrating erosion and splaying of the right maxilla and the mandibular ramus.

dl glucose (normal range 50–85 μg/dl), and 114 mmol/L chloride (normal range 122–132 mmol/L).

Postoperative Course. Postoperatively, the baby had an uneventful recovery. A CT scan of the head was obtained 4 weeks after surgery, revealing normal growth of the brain and marked restoration of the calvarial deformity (Fig. 3). There was no change in the deformity of the facial bones. She was discharged home under the care of her family.

The patient was followed at regular intervals. At 1 year of age she was growing well. The right eyelids and the ear were normally located. The right-sided facial deformity persisted. At 3 years of age she underwent a facial reconstructive procedure. At 15 years of age the patient had grown normally, and her cranial and facial contours were normal.

Discussion

Glioneural heterotopias are widely believed to be malformations of the CNS. They are seen in only 1% of control volunteers but in 25% of patients with malformation of the CNS. These heterotopias are closely related topographically to the malformation. Any part of the neuraxis may be affected by these rests, but the common sites are at the level of the medulla oblongata and the lumbar-sacral area. Intracranial extracerebral heterotopias have also been reported in the middle cranial fossa. Extracranial subcutaneous lesions are rare. They are seen at the root of the nose. One case each has been reported from the temporofacial, occipital, and nape of the neck area. Cooper and Kernohan suggested that glioneural islands originate as protrusions of the brain or spinal cord. They are “pinched off” the neuraxis and come to lie in the subarachnoid space or, rarely, in the extracranial or spinal tissue. During embryonic formation the frontal, parietal, and greater wing of the sphenoid bones do not interdigitate in sutures but are bridged by a fibrous mem-

Fig. 2. *Upper:* Photomicrograph showing spaces lined by ependymal cells. Papillae suggestive of choroid plexus are present in the largest cavity. *Lower:* Photomicrograph showing glial tissue (pale area) alternating with collagen. H & E, original magnifications × 100.
brane. This separates the dura from the developing skin. Protrusion of the dura and the brain through this membrane take place during Week 3 of gestation. Complete obliteration and destruction of the connecting stalk by the growing frontal, parietal, and sphenoid bones could result in the formation of a subcutaneous temporal glioneural heterotopia, as it did in our patient.

The ectopic nodule grew and increased in size. It contained cerebrospinal fluid (CSF) secreted by the choroid plexus. As the secretion continued it caused further swelling and formation of a cyst. It produced increasing pressure on the underlying bones, resulting in deformity of the calvaria and destruction, deformity, thinning, and splaying of the facial bones.

The calvarial deformity was markedly restored within 1 month after surgery as a result of normal growth of the brain. These changes were shown in the 4-week postoperative CT scan and support our hypothesis that the pressure effect caused the deformity in our patient. It seems likely that the absence of the zygoma in our patient might be the result of its total destruction. The maxilla and the mandible are formed from the mandibular arch at approximately Week 6 of gestation. At the end of Week 8 the zygoma begins to form when three centers take shape in the maxillary process of the mandibular arch. They unite at midterm. The considerable size of the temporofacial mass in our case might have disturbed and destroyed the developing process of the zygoma before Week 8 of gestation.

The biochemical composition of the cyst fluid was similar to that of CSF and was considered to be the secretion of ectopic choroid plexus. The liquid was straw colored and contained a high level of protein. This was considered to be the result of traumatic bleeding into the cyst that occurred during the failed vaginal delivery. The neural tissue lining a meningoencephalocele sac, in general, consists of glia and neurons, and it is often lined by ependyma. It is not a heterotopia because of the presence of neural tissue resulting from herniation of a part of the brain through a bone defect in the skull. In our patient the brain was of normal anatomical and morphological composition. The glioneural tissue in the sac was therefore considered to be heterotopic. These lateral heterotopias are not associated with any cerebral defect, in contrast to midline heterotopias, which are associated with various congenital defects of the brain.

References


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