Mature teratoma associated with an interparietal encephalocele

Case report

MEHMET TURGUT, M.D., PH.D.,1 AND İBRAHİM METEOĞLU, M.D.2

Departments of 1Neurosurgery and 2Pathology, Adnan Menderes University Hospital, Aydın, Turkey

Teratomas and encephaloceles are both uncommon congenital malformations that originate near the midline in the head and neck. An encephalocele is an extracranial extension of intracranial structures through a defect in the skull, whereas a teratoma consists of tissue from all three embryonic germ layers and is a true congenital tumor.

In the present study the authors describe the first reported case of a mature teratoma associated with an interparietal encephalocele presenting as a large midline subcutaneous mass in a newborn child. The appearance of these two pathological entities together suggests a common origin. The embryological significance of this finding is discussed, and the pertinent literature is reviewed.

KEY WORDS • interparietal encephalocele • mature teratoma • bifid sagittal sinus • magnetic resonance imaging • cranium bifidum • pediatric neurosurgery

Case Report

History and Examination. This 2-month-old girl, born by uneventful vaginal delivery after an uncomplicated pregnancy, was referred to our neurosurgery clinic because of a mass in her parietal region that was present since birth. No enlargement in the lesion was noticed when the infant cried. On physical examination, an alopecic and nonpulsatile subscalp lesion surrounded by an area of hypertrichosis was observed in the posterior interparietal area (Fig. 1A). Results of a neurological examination were normal. Computed tomography scanning of her head revealed a bone defect, and on MR imaging a midline subscalp mass in the parietal region was demonstrated with intracranial extension and a persistent primitive falcine vein (Fig. 2A and B). In addition, a 2D time-of-flight MR venography study revealed the presence of a fenestration at the posterior portion of the SSS, known as a bifid sinus (Fig. 2C and D).

Operation, Pathological Findings, and Postoperative Course. The patient underwent resection of the mass and the associated fibrous tract (Fig. 1B). There were no connections with any major vascular structures or the subarachnoid space, and the lesion was excised en bloc (Fig. 1C). Gross pathological examination revealed a light-brown polypoid cystic mass. Histopathological examination showed that the mass was covered with skin on all sides, and the fibrous stroma within the tumor contained mature adipose tissue, mature cartilage, respiratory epithelium, smooth muscle fibers, and nerve bundles, in addition to normal ependymal cells on the fibrous tract. Neuroglial remnants and lipo-matous tissue were observed within the mass (Fig. 3); there was no evidence of malignancy. A histopathological diag-
nosis of a mature teratoma was made. The child's early postoperative course was uneventful. Nearly 1 month postoperatively, follow-up MR imaging confirmed that the mass had been completely removed and that there was no macroscopic abnormality of the brain.

Discussion

Teratomas are primary congenital tumors made up of tissue that arises from all three embryonic germ cell layers: the ectoderm, endoderm, and mesoderm. The term teratoma originates from the Greek word "teraton," which means "monster." These lesions occur in approximately one of every 4000 live births and are more common in girls. Embryologically, teratomas are believed to arise from germ cells, during their course of migration during embryogenesis, and from nongerm embryonic cells that have escaped their regulatory influences. On the other hand, it has been suggested that parietal encephaloceles result from a developmental error occurring sometime during the first 7 to 10 weeks of fetal life, although the exact pathogenesis remains obscure. 

Teratomas are typically sporadic, but genetic factors have also been implicated. Some teratogens such as x-ray or valproic acid exposure, folate deficiency, and high doses of vitamin A have been implicated as a causative agent for the development of encephalocele. In the embryo, the left and right primordia of the SSS are seen on embryonic Day 35 and fuse on embryonic Day 50. Therefore, any interference by an encephalocele could cause venous channel anomalies because neurulation is completed by the time of SSS fusion in the midline.

Clinically, a hairless round scalp lesion with clear margins that is located at the midline of the parietal region between the lambda and bregma is referred to as an atretic parietal encephalocele or an IPE. Other authors have reported that, in some cases, the subscalp mass enlarges with crying because of the presence of a communication with the supracerebellar, suprapineal, or quadrigeminal cistern, or with the lateral ventricles. In our patient, however, there was only a fibrous tract terminating in the falx or tentorium, as seen on gross pathological and microscopic examination. Associated congenital anomalies including hydrocephalus, Fallot tetralogy, interhemispheric cyst, Dandy–Walker malformation, holoprosencephaly, Chiari malformation Type II, macrocephaly, microphthalmia, agenesis of the vermis or corpus callosum, and hypointensity of the white matter have been previously described. Microscopically, meningeal and vestibial neural tissue, ectopic glial tissue, and abnormal vascular tissue have been observed within the mass. As far as we know, there are only a few reports in the literature regarding an associated encephalocele and teratoma in a frontal or occipital location. Until now, however, the occurrence of mature teratoma elements within the mass in a case of IPE has not been reported in the English-language literature.

Fig. 1. Preoperative photograph of the midline subscalp mass in the skin of the parietal region (A), and intraoperative views of the surgical area before (B) and after (C) resection of the fibrous tract that connected the mass to the falx.

Fig. 2. Contrast-enhanced T1-weighted coronal (A) and sagittal (B) MR images demonstrating intracranial extension of the IPE and a huge heterogeneous extracranial mass with a hyperintense lipomatous tissue component. Magnetic resonance venograms (C and D) showing the defect in the bifid SSS through which the encephalocele protruded. Note the ascending primitive falicne vein associated with the subscalp mass.
Since the advent of neuroimaging techniques, the morphological features of this uncommon embryological lesion are being described more frequently in clinical practice. The differential diagnoses include lipoma, epidermoid and dermoid cysts, sinus pericranii, proliferating hemangioma, and respiratory epithelium (D). Inset: Immunopositive reaction for GFAP. H & E (B–D) and anti-GFAP staining (inset). Original magnifications × 20 (B), × 100 (C), × 40 (D), and × 200 (inset).

Patient outcome is excellent when the mass is completely excised, as in our case.

References


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Address reprint requests to: Mehmet Turgut, M.D., Ph.D., Cumhuriyet Mahallesi, Cumhuriyet Caddesi, No: 6 Daire: 7, TR-09020 Aydin, Turkey. email: drmturgut@yahoo.com.