Bilateral temporal bone encephaloceles after cranial irradiation

Case report

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Irradiation of the central nervous system may cause significant morbidity, including endocrine dysfunction and intellectual impairment. The authors report a case of bilateral temporal bone encephaloceles in a 21-year-old man who had received prophylactic central nervous system irradiation for acute lymphocytic leukemia in early childhood. Endaural encephaloceles are uncommon, and most occur as a complication of mastoid surgery. The etiology, clinical features, radiological diagnosis, and surgical treatment of temporal bone encephaloceles are discussed.

KEY WORDS • encephalocele • central nervous system • radiation-induced lesion

IRRADICATION of the central nervous system (CNS) may induce a variety of complications, including necrosis of bone and brain, intellectual impairment, visual loss, cranial neuropathies, hypopituitarism, and malignancies. We report a case of bilateral temporal bone encephaloceles in a man who had received external beam irradiation to the CNS in early childhood.

Case Report

This 21-year-old man was referred to an otologist for evaluation of a middle ear mass that caused bilateral intermittent tonal tinnitus and a vague “echo” in the right ear. He had experienced recurrent otitis media during childhood, but had no history of hearing loss, otalgia, otorrhea, otological surgery, meningitis, or significant head trauma. At 2 years of age, he was diagnosed as having acute lymphocytic leukemia. He was treated with vincristine, prednisone, and intrathecal methotrexate, and received 24 Gy of whole-brain radiation therapy to opposing 15 × 16-cm fields with a linear accelerator. A maintenance regimen of 6-mercaptopurine, methotrexate, prednisone, and vincristine was continued for 3 years. At 7 years of age, the patient suffered a testicular relapse of the acute lymphocytic leukemia, which was treated with an additional 18 Gy of radiation to the whole brain and 28 Gy to the scrotum, as well as with vincristine, prednisone, and L-asparagi-

nase. A maintenance drug regimen of 6-mercaptopurine and methotrexate was continued for 4 years.

At his current presentation, the patient’s acute lymphocytic leukemia was in remission. He was receiving replacement testosterone and growth hormone to compensate for pituitary insufficiency. His intellectual development, which previously had been delayed, was normal.

Examination. Otoscopy of the right ear revealed a posterior mesotympanic mass with a pial vascular pattern. The anterior tympanic membrane was normal.
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The left tympanic membrane was minimally retracted. Weber's and Rinne's auditory tests showed no abnormalities. A 10- to 20-dB low-frequency conductive hearing loss and normal sensorineural function were revealed on an audiogram. The speech reception threshold was 5 dB, and the speech discrimination score was 100%.

Computerized tomography (CT) demonstrated a grossly dystrophic skull with widened internal auditory canals bilaterally. There was a soft-tissue mass in the right mesotympanum, epitympanum, and mastoid cavity; on the left side, the soft tissue was confined to the mastoid antrum (Fig. 1). Magnetic resonance (MR) imaging demonstrated bilateral endaural encephaloceles, larger on the left than on the right, that were isointense with brain on T₁-weighted images and showed minimal enhancement after administration of gadolinium (Fig. 2).

Operations. The right endaural encephalocele was repaired through a combined right transmastoid and middle fossa craniotomy procedure. The basal temporal lobe had herniated through a 2-cm defect, which represented diastasis of the petrosquamous suture line, on the floor of the middle fossa. The encephalocele was not covered with dura and occupied the mastoid cavity. The arachnoid covering of the encephalocele was only mildly hyperemic. Herniated brain enveloped the ossicles, necessitating a tympanotomy to remove the encephalocele completely without causing ossicular injury. To minimize transmitted vibratory trauma, the KTP laser* (2 W, 0.1 second) was used to vaporize the brain from the ossicular chain. The herniating temporal lobe appeared nonviable and was divided. The defect was repaired in three layers: a free temporalis fascia graft was placed extradurally, the inner table of the middle fossa craniotomy bone flap placed extradurally along the middle fossa floor, and an inferiorly based temporalis muscle flap placed between the dura and the bone graft (Fig. 3).

* KTP laser manufactured by Laserscope, San Jose, California.
Six months after the operation, the left mastoid encephalocele was repaired through a middle fossa craniotomy and mastoidectomy. The defect in the petro-squamous suture line extended from the malleal head anteriorly to the junction of the transverse and sigmoid sinuses posteriorly. Nonviable herniating temporal lobe was divided, and the defect was repaired in three layers with intradural temporalis fascia, temporalis muscle between the dura and the temporal floor, and cortical bone bridging the bone defect.

**Postoperative Course.** After both operations, recovery was rapid and uncomplicated. An MR image obtained after the first operation showed a successful repair of the right temporal lobe encephalocele and the un repaired mastoid encephalocele on the left temporal lobe (Fig. 4). After the second operation, an audiogram showed resolution of the conductive hearing loss bilaterally.

**Pathological Examination.** Histological examination of both encephaloceles showed gray matter with extensive gliosis, degenerative changes, and chronic inflammation consisting principally of macrophages.

**Discussion**

**Clinical Presentation**

Cranial irradiation for acute lymphocytic leukemia in children has long been associated with adverse physiological effects.4 Endocrine dysfunction after radiation therapy is well documented. Irradiation disturbs the hypothalamic pituitary axis and causes dysfunction in the secretion of growth hormones; it has also been reported that the basal frequency and amplitude of growth hormone secretory pulses are decreased.4 Growth hormone deficiency may retard growth and in females disturbs the timing of puberty.2,20 Several investigators have shown a direct correlation between the onset of menarche and the patient’s age at the time of irradiation.2,21 Central nervous system malignancies, including sarcomas, meningiomas, and gliomas, are virulent and often lethal complications of radiation therapy.6

The extent of neuropsychological impairment resulting from cranial irradiation is controversial. Radiation therapy decreases head and skeletal growth.3,8 Children with acute lymphocytic leukemia who are treated with cranial irradiation are three to four times more likely to have learning problems than those who are not irradiated.12 Children who undergo repeat radiation therapy for CNS relapse of acute lymphocytic leukemia are at greater risk for a decline in intellectual function than those who remain in remission.10,12,13 In one study, all children treated with 24 Gy and those under the age of 3 years treated with 18 Gy had reduced head growth, decreased concentration, impaired short-term memory, and learning difficulties in the classroom.1

Endaural encephaloceles are uncommon; fewer than 150 cases have been reported in the last 40 years.3 Most temporal bone encephaloceles are due to complications of mastoid surgery; others are spontaneous or idiopathic, traumatic, or a complication of chronic otitis media and chronic mastoiditis. Common signs and symptoms at presentation include cerebrospinal fluid (CSF) otorrhea or rhinorrhea, “serous” otitis media due to CSF in the tympanic cavity, conductive hearing loss, and meningitis (often recurrent). Unusual presentations of patients with endaural encephaloceles include a mass behind the tympanic membrane (as in this patient), expressive aphasia, temporal lobe seizures, and facial nerve weakness. In patients with suspected CSF otorrhea or rhinorrhea, evaluation of the fluid for glucose, chloride, and β2-transferrin is useful in confirming the diagnosis of a CSF leak.15

Despite a diligent search of the literature, we found no reports of a temporal bone encephalocele after radiation therapy. Our patient received 24 Gy of radiation administered prophylactically to the CNS at the age of 2 years and 18 Gy at the age of 7 years. This early exposure to radiation probably retarded the growth of the floor of the middle cranial fossa. Other peculiarities in the patient’s cranial base anatomy support this theory. Normally, the caudal squamous part of the temporal bone extends medially, joining the superior surface of the petrous bone to form the roof of the middle ear and mastoid.19 The petro-squamous suture is formed by the dehiscence between the squamous and petrous bone prior to their fusion; it is usually obliterated by 1 year of age, but fusion can be delayed.17 In our patient, the petrous and squamous parts of the temporal bone apparently failed to fuse, resulting in a bone dehiscence along the temporal floor. We postulate that gravity and CSF pulsations gradually thinned the dura overlying the dehiscence, and brain protruded into the mastoid and tympanic cavities. Chemotherapy and hypopituitarism may have contributed to the disturbance of skull growth.
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Radiological Diagnosis

High-resolution CT in the coronal and axial planes are needed to define defects in the tegmen tympani or mastoid cavity and to identify soft tissue and fluid in the tympanic and mastoid cavities. Magnetic resonance imaging is required to differentiate herniated brain from fluid and other lesions.6 When CT scans suggest a defect of the tegmen, MR imaging is ideal for determining the integrity of the overlying dura. Distortion of the gyri resulting from herniating temporal lobe into the temporal bone ("tear-drop" sign) on coronal CT or MR images strongly suggests an encephalocele. Computerized tomography obtained after intrathecal administration of Omnipaque, a low-osmolarity, nonionic, watersoluble contrast agent that is useful for opacifying the CSF, can identify any fistulous tract responsible for the CSF leak.

Surgical Treatment

Endaural encephaloceles of the middle fossa floor may be repaired from below (mastoidectomy), from above (middle fossa craniotomy), or by a combination of both intra- and extradural exposures. When the transmastoid approach alone is used, the herniated gial tissue is amputated and the tegmen defect is repaired with a bone or composite cartilage-perichondrium graft. To reinforce this closure and to separate it from the mucosal envelope, the mastoid cavity may be obliterated with temporalis muscle or a Palva flap.14 Larger encephaloceles are best approached from the middle fossa. The dural defect can be repaired extradurally, intradurally, or both. Dura should be reconstituted with a thick connective-tissue graft, such as temporalis fascia, pericranium, and fascia lata. The tegmen defect is best repaired with a bone or cartilage graft, as soft-tissue repairs alone may not prove durable. A vascular covering for this free bone graft may be provided by rotating the superior portion of the temporalis muscle over the middle fossa floor. When the encephalocele impinges on the ossicular chain, a combined tympanomastoid-middle fossa approach,1 as in our case, provides the additional exposure needed to permit atraumatic dissection of the pia mater tissue from the conductive mechanism.

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References


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