Intradiploic cerebrospinal fluid fistula

Report of two cases

ANTONIO C. G. D'ALMEIDA, M.D., AND ROBERT B. KING, M.D.
Department of Neurosurgery, State University of New York,
Upstate Medical Center, Syracuse, New York

Two patients with asymptomatic osteolytic skull lesions were found to have cerebrospinal fluid diploic fistulas. The radiographic and operative findings have not been reported previously.

KEY WORDS • cerebrospinal fluid fistula • osteolytic skull lesion • intradiploic fistula

This report describes the clinical, radiographic, and operative findings in two patients with idiopathic intradiploic cerebrospinal fluid (CSF) fistulas. A review of the neurosurgical and radiological literature has failed to reveal a description of patients with similar findings.1-5,8

Case Reports

Case 1

This 61-year-old woman was well until 12 days before admission, when she suddenly became light-headed and noticed decreased hearing on the left side. There was no history of antecedent infection or trauma. She was thought to have Ménière's disease or labyrinthitis. Her presenting symptoms gradually subsided. Her past history was unremarkable except that she had had a slowly enlarging asymptomatic mass over the right parietal area for 5 years.

Examination. Her general physical examination was normal. A large mass (7 × 8 × 3 cm in size) that was bony, hard, nonpulsatile, and non-tender was visible in the right parietal bone. There was no scalp abnormality or bruit.

Skull x-ray film (Fig. 1a) demonstrated an osteolytic parietal lesion. The scalloped margins were distinct in some areas and appeared eroded in others. Tomograms of the lesion (Fig. 1b) showed widened diploic spaces, thinning or erosion of the flattened inner table, and elevation of the thinned outer table, which was otherwise intact. Computerized tomography (CT) of the skull (Fig. 1c) revealed no intracranial pathology and again demonstrated the intradiploic lesion. A technetium (99mTc) bone scan (Fig. 1d) demonstrated the defect in the skull, with a margin of increased activity. Selected internal and external carotid angiograms showed no blood supply to the lesion or other vascular abnormality. Routine laboratory studies and chest films were unremarkable. Serum protein electrophoresis was normal.

Operation. A craniectomy was performed. The peristium was intact over the surface of the lesion and appeared normal. The cortical bone at the vertex of the mass was extremely thin, mottled, and bluish in color (Fig. 2a). When this thin bone at the center of the mass was removed, there was a flush of crystal clear fluid which appeared to be normal CSF. It welled up from widely expanded diploic spaces that were filled with a latticework of fine bony trabeculae. When the trabeculae were removed and the inner table exposed, a small defect with a 2 × 2 mm tuft of opaque gray tissue protruded through the inner table, spurting small quantities of CSF into the field with each pulse (Fig. 2b). The inner table of the skull was removed, exposing thickened dura. In the middle of the exposed dura was found a defect measuring approximately 3 mm in diameter, through which a bleb of arachnoid protruded, still squirting CSF with each
Intradiploic cerebrospinal fluid fistula

Fig. 1. Case 1. a: Detail of a skull x-ray film demonstrating a radiolucent defect with irregular margins in the right parietal bone. The inner and outer table of the skull is thinned and expanded, and a hint of trabeculae can be seen within the lesion. b: Tomogram of the right parietal bone showing widening of the diploic space with erosion, thinning, and bowing of the inner and outer table, suggesting a polycystic lesion. c: Computerized tomography of the head, in conventional (left) and coronal (right) planes, showing an expanding lesion of the skull with bony trabeculae. The lesion is seen as a very low density area between the skull tables. d: Vertex view of the bone scan showing the ring enhancement at the margin of the lesion.

Fig. 2. Case 1. Operative photographs. a: Patchy thinning of the outer table with underlying cerebrospinal fluid at the vertex (arrows). b: Arachnoid bleb penetrating the inner table of the skull spurting cerebrospinal fluid into the diploic space (arrow).
A. C. G. D’Almeida and R. B. King

FIG. 3. Case 2. a and b: Skull films showing a lytic lesion of the skull with scalloped margins, suggesting multiple cystic components. c: Tomogram showing expanded cortical bone but no diploic trabeculae. The skull is thickened at the margins of the lesion (arrows). d: Technetium scan showing increased uptake at the margins of the lesion where the skull is thickened (arrow).

pulse. The margins of the dura were freed from the arachnoid which was lightly coagulated, sealing the CSF leak. A periosteal graft was placed on the inner and outer surfaces of the dura and sutured in place in a watertight closure. The skull defect was then filled with a methacrylate cranioplasty, and the incision was closed in the usual fashion. The patient has had an uneventful postoperative course.

Case 2

This 53-year-old man was hospitalized with an incidental finding on skull films (Fig. 3a and b) of an asymptomatic osteolytic lesion in the right frontal bone.

Examination. A CT scan (Fig. 3c) was thought to indicate a lesion arising within the diploic space, extending into the epidural space and expanding the outer table. There was some reaction about its margin on 99mTc bone scan (Fig. 3d). A brain scan was negative. The lesion was not evident on physical examination. He had bumped his head against a cupboard about 5 years earlier without sequelae, but gave no history of other head trauma or chronic disease.

Operation. At the time of craniectomy, the periosteum appeared normal. The outer table of the skull was thinned and appeared to have some clear or opalescent fluid beneath it. When the bone was chipped away, pulsating CSF welled up from the cavity beneath. Further craniectomy of the outer table exposed very thin inner cortical bone with two small holes. Apparently normal CSF poured into these two holes. With further removal of the inner table of the skull, a small area of arachnoid adhesions was identified but no arachnoid cyst or cicatrix was encountered. The underlying brain appeared to be “slightly atrophic.” The dura was closed with a periosteal patch graft and methacrylate cranioplasty was used to fill the cranial defect. The patient’s postoperative course has been uneventful.

Discussion

Solitary asymptomatic osteolytic skull defects have been described with a variety of lesions, including congenital malformations (meningocele, encephalocele, arachnoid cyst), infectious processes (tuberculosis, syphilis, osteomyelitis, hydatid cyst), granulomatoses

Intradiploic cerebrospinal fluid fistula

(eosinophilic granuloma, lipoid granulomatosis), benign tumors (epidermoid or dermoid cysts, lipoma, osteoma, fibroma, hemangioma, meningioma, fibrous dysplasia, giant-cell tumors, aneurysmal bone cysts), malignant tumors (metastatic tumors, osteosarcoma, fibrosarcoma, Ewing's sarcoma, reticulum cell sarcoma, solitary myeloma), and as local manifestations of systemic disease (sarcoidosis, von Recklinghausen's disease, and Paget's disease).

In these two cases, considering the late age of onset without evidence of systemic disease or significant trauma, our initial considerations included plasmacytoma, hemangioma, aneurysmal bone cyst, dermoid or epidermoid cyst, and eosinophilic granuloma. Plasmacytoma was thought to be unlikely because of the normal serum protein electrophoresis. Metastatic tumor was considered unlikely after a limited but unsuccessful search for a primary lesion or other metastatic foci and a 5-year clinical history in Case 1. Hemangioma, aneurysmal bone cyst, and meningioma were not seriously suspected on the basis of the angiography. The preoperative diagnoses still being considered in Case 1 included fibrous dysplasia, dermoid or epidermoid cyst, and eosinophilic granuloma, although one consultant still thought a sarcomatous change in a benign lesion was a possibility. In Case 2, the preoperative diagnoses included eosinophilic granuloma, reticulum-cell sarcoma, multiple myeloma, or metastatic neoplasm. None of these possibilities seemed well suited to the findings in either case. It was noted in Case 2 that the thinning of bone inferior to the lytic lesion might represent an extradural or atrophic lesion associated with a cystic lesion which in fact thinned the bone from within. Surgery was therefore indicated for both diagnostic and therapeutic purposes.

We were not familiar with the lesions identified at the time of the craniectomy. In both cases, there clearly was a CSF fistula through a punctate defect in the arachnoid and dura mater, extending into the medullary portion of the skull. This part of the skull must have expanded gradually over many years, elevating the outer table, flattening the inner table, and expanding the diploic spaces, to finally present as an asymptomatic skull mass in Case 1 and an incidental osteolytic lesion in Case 2.

In retrospect, there were unusual features of these lesions. We consider them, therefore, to represent idiopathic intradiploic cerebrospinal fluid fistulas.

Intradiploic cerebrospinal fluid fistula

(tions within the lesion, and x-ray absorption numbers in some areas approximated those of CSF; however, they were higher in other portions of the lesion, presumably due to the small bony trabeculations or artifact from nearby bone. The combination of these observations from tomography, CT scan, and bone scan with an asymptomatic chronic bone lesion may lead to a correct preoperative diagnosis in future cases.

In one of these cases, needle aspiration for diagnostic purposes was discussed but not undertaken. Unless the proper diagnosis was clearly established and alternative diagnoses in all probability were excluded, we would not feel comfortable with that simple confirmatory maneuver, although to be sure it might have avoided craniectomy and cranioplasty for these two patients. An alternative method of studying the lesion preoperatively would be to instill metrizamide into the spinal subarachnoid space and then examine the lesion with CT scan to demonstrate the accumulation of metrizamide within the cavity of the skull lesion.

It seems unlikely that a congenital defect of arachnoid, dura, or bone contributed to setting the stage for these lesions. Unlike the floor of the middle fossa or sella turcica, small congenital defects of the inner table are not encountered over the cranial vault in association with brain ex crescences or dural ectasia unless additional evidence of ectodermal or mesodermal abnormalities is evident.

We were unable to obtain a clear history of significant head trauma from either the patients or from their families. It is presumably possible, however, that, at some forgotten time in the past, they sustained a minor blunt head injury that caused a small linear fracture of the inner table of the skull with injury to the dura and the delayed onset of a minute arachnoid pouch extending through a small dural rent, which gradually eroded through the inner table of the skull and ruptured into a diploic space establishing the CSF fistula. A true leptomeningeal cyst did not develop. There was no arachnoid in the diploic spaces, which were filled with CSF.

We have no clear understanding of the etiology of these lesions. We consider them, therefore, to represent idiopathic intradiploic cerebrospinal fluid fistulas.

Acknowledgment

Dr. Donald H. Stewart, Jr. kindly permitted us to report the second case, and read the manuscript.

References


Address reprint requests to: Robert B. King, M.D., Department of Neurosurgery, State University of New York, Upstate Medical Center, Syracuse, New York 13210.