Intrinsic brainstem epidermoid cyst

Case report and review of the literature

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Brainstem epidermoid cysts are rare lesions, with only 18 reported cases in the literature and only five purely intrinsic epidermoid cysts within this group. The authors present the case of a 3-year-old girl with a history of chronic headaches, progressive diplopia, and relapsing and remitting mild right hemiparesis who was found to harbor an intrinsic brainstem epidermoid cyst at the pontomedullary junction. Initial working diagnoses included intrinsic brainstem astrocytoma and cavernoma. After tumor enlargement and progressive symptoms, a diffusion-weighted (DW) magnetic resonance (MR) imaging sequence was performed and a definitive diagnosis of an intrinsic brainstem epidermoid cyst was made in the patient. The patient underwent a suboccipital craniotomy and complete resection of the cyst with the aid of intraoperative neurophysiological monitoring. Three years after the operation, the patient is neurologically intact and no evidence of tumor recurrence has been found. The rarity of brainstem epidermoid cysts can make their diagnosis difficult; thus a DW MR imaging sequence of the brain is a useful diagnostic modality. Intrinsic brainstem epidermoid cysts can be removed safely, in a manner similar to that used for the surgical treatment of focal tumors.

KEY WORDS • epidermoid • brainstem • diffusion-weighted magnetic resonance imaging • pediatric neurosurgery

Brainstem epidermoid cysts are slow-growing congenital tumors that account for 0.2 to 1.8% of all primary brain tumors.7 Arising primarily in the basal subarachnoid spaces, these tumors are mainly intradural lesions, commonly found in the CPA and juxtasellar areas.1,5,20,22 Growth rates are similar to those of epidermal cells, and the lesions appear to proliferate along open cisternal areas rather than infiltrating tissue parenchyma.22 Although they are considered histologically benign, epidermoid cysts can cause significant neurological defects by compressing vital structures.9

Epidermoid cysts of the brainstem are rare, with only 18 reported cases in the literature3,4,6–18,23,25 and only five of them intrinsic to the brainstem.1,5,18,23,25 We describe an interesting case of an intrinsic brainstem epidermoid cyst in a 3-year-old girl. Because the cyst was purely intrinsic, the diagnosis was initially unclear until definitive neuroimaging was performed using DW MR imaging sequences. The presentation, differential diagnosis, radiological imaging, and surgical management are discussed, along with a review of the reported cases in the literature.

Case Report

Presentation and Examination. This 17-month-old girl initially presented to an outside institution after having experienced sudden-onset right hemiparesis, facial asymmetry, and loss of balance. Computerized tomography and MR imaging of the brain revealed a well-circumscribed lesion in the anterior pontomedullary junction. The lesion was predominantly hypointense on T1- and hyperintense on T2-weighted images. A positron emission tomography scan of the brain revealed hypometabolic activity in the major portion of the mass but not at the periphery. The results of the MR imaging and positron emission tomography studies suggested that the lesion represented an intrinsic brainstem tumor; however, the patient had experienced marked clinical improvement with steroid treatment only and a short rehabilitation stay. A subsequent MR imaging study obtained 3 months later showed slight reduction in the size of the lesion, minimal signal abnormality, and no evidence of extension into the pons. Some hyperintensity on T1-weighted images was observed, however, which was suggestive of intratumoral hemorrhage. The child never received oncological therapy, and was followed prospectively with sequential MR imaging. Images obtained subsequently showed a stable lesion at the pontomedullary region of the brainstem (Fig. 1). A presumptive diagnosis of brainstem cavernoma was then made. On initial examination, the patient exhibited a horizontal gaze paresis, but otherwise she had no significant neurological deficit and was quite active.

At 35 months of age (18 months after the initial exami-
nation), she suffered progressive recurrent headaches as well as fever. At that time, her white blood cell count was quite high, but no additional workup was performed. She was started on a course of steroid agents because MR images revealed significant enlargement of the lesion. The preoperative MR images (Fig. 2 upper left and right, lower left) showed a large intrinsic cystic lesion arising at the pontomedullary junction, which had a low T1 signal, a high T2 signal, and no contrast enhancement. She was referred to our institution, where a DW MR imaging sequence was obtained (Fig. 2 lower right), revealing a bright signal within the tumor suggestive of an epidermoid cyst rather than a cavernous malformation or brain tumor.

Operation. The patient underwent a suboccipital craniotomy and C-1 laminectomy for resection of the lesion. The floor of the fourth ventricle was exposed and found to be distended from the underlying cyst. Intraoperative monitoring was performed throughout the surgery. The facial colliculus and the hypoglossal nuclei were localized using intraoperative electrophysiological mapping of the floor. A myelotomy was performed through the thinned floor, revealing a viscous, pearly white substance that was easily aspirated. No significant change in her neurophysiological monitoring was noted.

Postoperative Course. Postoperatively, the patient experienced mild, transient sixth and seventh cranial nerve pal-
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Epidermoid cysts form between the 3rd and 5th week of embryonic development from displaced epithelial remnants that remain after neural tube closure. They are composed of an outer capsule, an epithelial layer, and, in some cases, an inner cystic fluid. The outer layer is composed of connective tissue, which surrounds a layer of keratinized stratified squamous epithelium. As the epithelial layer desquamatates, the cells accumulate and form a cholesterol-rich inner layer that gives the cyst its characteristic pearly white appearance. The CPA and parasellar areas are the most common sites for the development of epidermoid cysts. Those located in the brainstem occur very rarely, and purely intrinsically lesions without exophytic extensions have been reported previously in five cases only. Epidermoid cysts develop slowly, and onset of neurological symptoms is usually gradual. Patients harboring these cysts typically become symptomatic between the ages of 20 and 40 years. Due to the insidious manner in which they grow, brainstem epidermoid cysts in children are extremely rare and have been reported in eight cases only.

Patients presenting with intrinsic epidermoid cysts commonly exhibit symptoms related to compression of associated brainstem structures. Among the 18 reported cases, the most common signs included hemiparesis (72%), seventh cranial nerve palsy (72%), sixth cranial nerve palsy (61%), and gait ataxia (61%). Other notable signs included aseptic meningitis (17%) and increased intracranial pressure (11%). In our patient, the initial presentation suggested lesion enlargement that resulted in seventh cranial nerve palsy, balance problems, and hemiparesis. Given the subsequent decrease in size of the cyst, it is likely that the fluid drained into the subarachnoid space. Although drainage of epidermoid cyst fluid into the subarachnoid space has been shown to produce aseptic meningitis, no sign of meningitis was noted in this case.

The mean age at presentation of purely intrinsic brainstem epidermoid cysts (21.6 ± 5 years) was not significantly different from the mean age at presentation of such cysts having an extrinsic component (22.1 ± 14 years); however, the median age at presentation of purely intrinsic epidermoid cysts (1.42 years) was different from the median age of presentation of intrinsic epidermoid cysts with an extrinsic component (25 years). Although the mean age of presentation was not significantly different between the two groups, the difference in their median age of presentation suggests that purely intrinsic epidermoid cysts preferentially afflict infants, whereas intrinsic epidermoid cysts containing an extrinsic component preferentially present in adults. The other possible reason for the younger age may pertain to the rapid growth that the purely intrinsic tumors often demonstrate. There is no preoperative information concerning the previous reports, but on the basis of the rapid growth of the tumor in our case, we postulate that these lesions may grow quicker when in the intrinsic brain.

Epidermoid cysts, often found in the CPA and juxtasellar areas, have characteristic appearances on conventional MR imaging sequences. The cyst often exhibits a slightly higher signal than cerebrospinal fluid on T1- and T2-weighted images. One distinguishing feature of epidermoid cysts is the lack of edema surrounding the lesion, which is responsible for their strongly hyperintense appearance on DW imaging. In extrinsic locations, DW imaging has emerged as an important and noninvasive diagnostic modality. Epidermoid cysts have a very high signal intensity related to the restricted diffusion of tumoral molecules, which is responsible for their strongly hyperintense appearance on DW imaging. In extrinsic locations, DW imaging has been shown to distinguish epidermoid cysts from mildly hyperintense or hypointense lesions such as abscesses or other tumors and to delineate their borders from cerebrospinal fluid. This case represents the first use of DW imaging in the literature on intrinsic brainstem epidermoid cysts, and it played a central role in diagnosis and preoperative planning. Postoperatively, it was used to assess the extent of resection and to monitor for recurrence. Given its dis-
Subtotal resection of epidermoid cysts is preferred if tumoral material can lead to aseptic meningitis. Great care must be taken to avoid spilling the capsular tissue is not easily resectable. Aggressive attempts to remove adherent epidermoid tissue are unsafe and can result in disastrous outcomes. Although evacuation of cystic fluid and complete resection of the surrounding capsule were achieved safely in this case, total resection of the epidermoid capsule is not always possible. In several reported cases, the cystic capsule was very adherent to neurovascular structures. Subtotal resection of epidermoid cysts is preferred if the capsular tissue is not easily resectable. Aggressive attempts to remove adherent epidermoid tissue are unsafe and can result in disastrous outcomes. Although epidermoid tissue that is left behind carries the risk of recurrence, good long-term outcomes with minor morbidity have been achieved with a more conservative approach to difficult cases.

In conclusion, this case represents one of the few reported instances of a purely intrinsic epidermoid cyst. The first use of DW imaging in the workup of a brainstem epidermoid cyst is reported and a central role for this modality in the diagnosis and postoperative follow up of brainstem epidermoid lesions is suggested. Although such a procedure is not always technically possible, this case represents a situation in which complete resection of a brainstem epidermoid cyst was both feasible and safe.

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