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Authors

Chung, Lawrance K
Lagman, Carlito
Duong, Courtney
et al.

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Dermoid Cyst of the Prepontine Cistern and Meckel's Cave: Illustrative Case and Systematic Review

Lawrance K. Chung[1] Carlito Lagman[1] Courtney Duong[1] Daniel T. Nagasawa[1] Alexander M. Tucker[1] William H. Yong[2] Isaac Yang[1,3,4]

[1] Department of Neurosurgery, David Geffen School of Medicine, University of California, Los Angeles, Los Angeles, California, United States
[2] Department of Pathology and Laboratory Medicine, David Geffen School of Medicine, University of California, Los Angeles, Los Angeles, California, United States
[3] Department of Radiation Oncology, David Geffen School of Medicine, University of California, Los Angeles, Los Angeles, California, United States
[4] Jonsson Comprehensive Cancer Center, David Geffen School of Medicine, University of California, Los Angeles, Los Angeles, California, United States

Address for correspondence Isaac Yang, MD, Department of Neurosurgery, University of California, Los Angeles, 300 Stein Plaza, Suite 562, 5th Floor Wasserman Building, Los Angeles, CA 90095-1761, United States (e-mail: iyang@mednet.ucla.edu).

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Abstract

Objective Dermoid cysts are benign, congenital malformations that account for ~0.5% of intracranial neoplasms. The authors describe a 42-year-old female with a prepontine dermoid cyst who underwent apparent gross total resection (GTR) but experienced cyst recurrence. To date, very few cases of prepontine dermoid cysts have been reported. The prevalent region where these cysts are located can be difficult to determine. In addition, the authors systematically review the literature to characterize the clinical presentation, anatomical distribution, and surgical outcomes of intracranial dermoid cysts.

Design Systematic review.

Setting/Participants PubMed, Web of Science, and Scopus databases.

Main Outcome Measures Extent of resection, symptom improvement, and recurrence rates.

Results A total of 69 patients with intracranial dermoid cysts were identified. Three (4.3%) intracranial dermoid cysts were located in the prepontine cistern. The average age of patients was 33.3 years. The most common presenting symptoms were headache (52.2%) and visual disturbances (33.3%). Intracranial dermoid cysts were distributed similarly throughout the anterior, middle, and posterior cranial fossae (29.0%, 36.2%, and 29.0%, respectively). GTR was achieved in 42.0% of cases. Thirty-four (49.3%) patients experienced symptom resolution. Recurrence rate was 5.8% at a mean follow-up of 2.1 years.

Conclusions Intracranial dermoid cysts most often present as headaches and visual disturbances. Intracranial dermoid cysts were found in the anterior, middle, and posterior cranial fossae at similar frequencies but with clear predilections for the Sylvian fissure, sellar region, and cerebellar vermis. Outcomes following surgical excision of intracranial dermoid cysts are generally favorable despite moderate rates of GTR.

Keywords
- dermoid cysts
- intracranial
- prepontine cistern
- systematic review

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Introduction

Dermoid cysts arise from ectopic epithelium sequestered within the neural tube during embryogenesis.[1–15] Intracranial dermoid cysts are uncommon and account for ~0.5% of brain tumors.[1,9,16,17] Intracranial dermoid cysts have been commonly reported near the sella and cerebellar vermis.[18] However, only three cases of prepontine dermoid cysts have been reported to date.[19–21] The authors herein describe an additional case of a prepontine dermoid cyst arising in a 42-year-old female who underwent apparent gross total resection (GTR) with subsequent cyst recurrence. The prevalent region where these deep-seated cysts are located may be difficult to determine; thus, the appropriate surgical approach is often unclear. Given the rarity of this location, a systematic review was performed to characterize the clinical presentation, anatomical distribution, and surgical outcomes of intracranial dermoid cysts.

Case Illustration

History and Physical

A 42-year-old female with no significant past medical history presented with 4 months of progressive dental paresthesias and lower jaw pain. On examination, she had left-sided facial pain in the distribution of V2 and V3. Her hearing was intact bilaterally with an excellent House–Brackman score.

Imaging

The patient underwent further evaluation with computed tomography (CT) and magnetic resonance imaging (MRI), including diffusion studies. CT revealed a hypodense mass, anterior to the brainstem and that extended into Meckel's cave on the left. T1- and T2-weighted sequences demonstrated a non-enhancing, hyperintense, 3 cm lobulated mass in the prepontine cistern, inferior to the left trigeminal nerve (►**Fig. 1**). Initial assessment suggested a dermoid cyst.

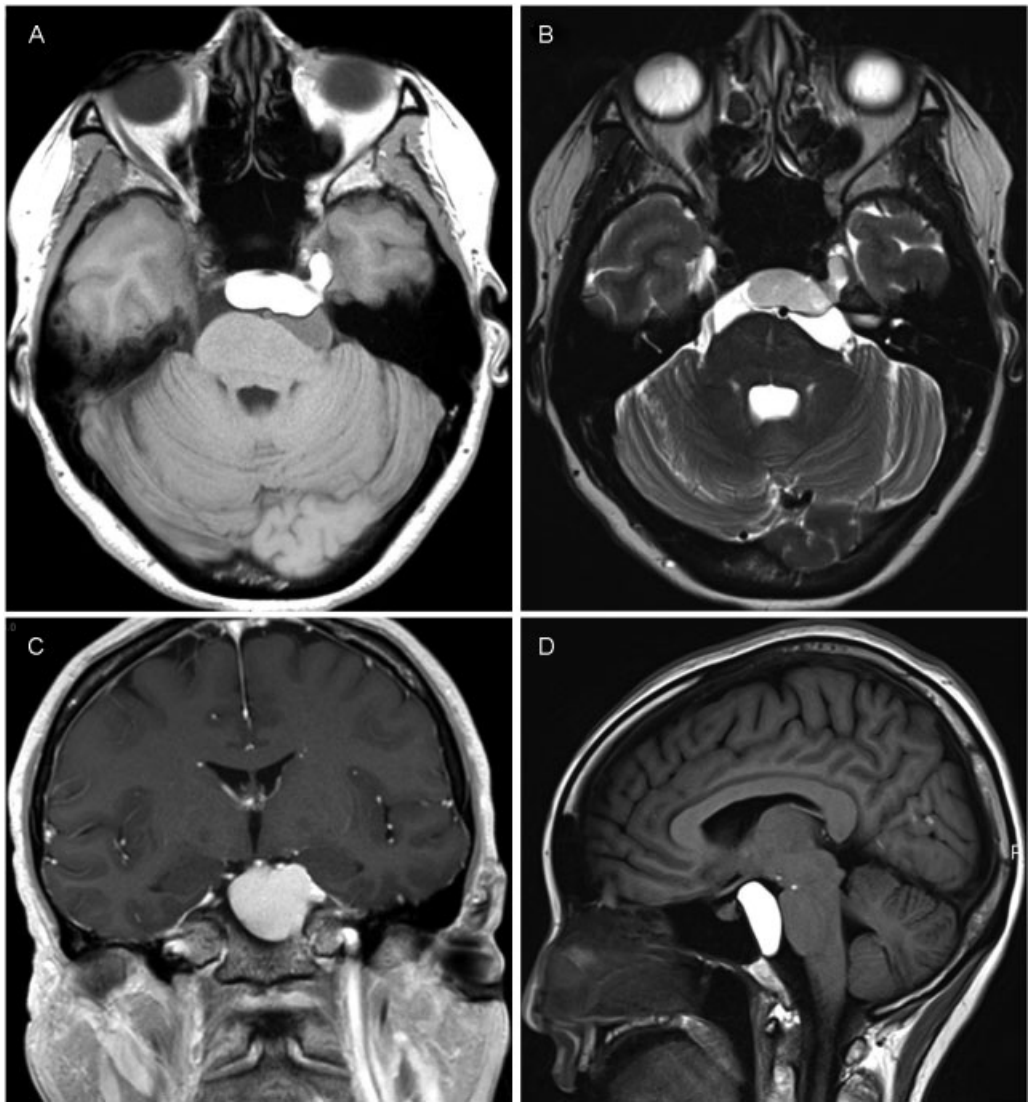

Fig. 1 Preoperative imaging for the illustrative case, (A) axial T1-weighted MRI showing a hyperintense mass in the prepontine cistern with extension into Meckel's cave, (B) axial T2-weighted MRI, (C) coronal T1-weighted MRI with contrast, and (D) sagittal T1-weighted MRI. MRI, magnetic resonance imaging.

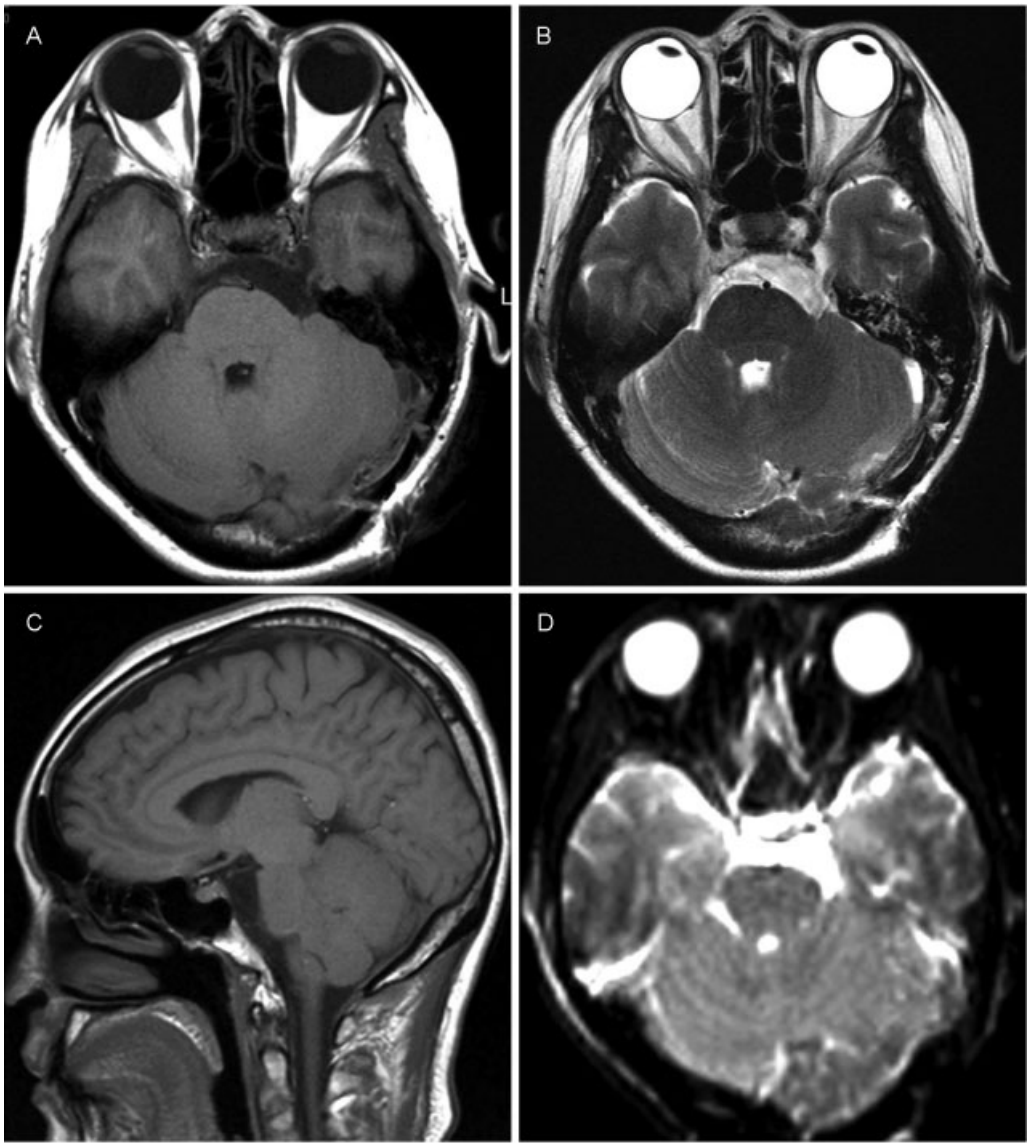

Fig. 2 Postoperative imaging for the illustrative case, (A) axial T1-weighted MRI demonstrating apparent gross total resection of the previously described prepontine mass, (B) axial T2-weighted MRI, (C) sagittal T1-weighted MRI, and (D) axial DWI. DWI, diffusion-weighted imaging; MRI, Magnetic resonance imaging.

Management and Pathology

Surgical resection was advised, and the patient was placed on gabapentin for symptomatic management. The patient underwent a suboccipital craniotomy (retrosigmoid approach) for resection of the mass. Dissection of several pontomedullary adhesions was performed under the operative microscope. Cerebrospinal fluid drainage and table rotation enabled visualization of the ventral brainstem. The capsule and its cystic contents were fully excised with no remnants visualized intraoperatively. The wound was irrigated with saline until clear, and the incision was closed in standard fashion. Immediate postoperative imaging indicated GTR (►**Fig. 2**). Final pathological examination showed hair fragments and scaly epithelium within the specimen, consistent with a dermoid cyst (►**Fig. 3**).

Follow-up

One-year follow-up imaging demonstrated a 9×3 mm recurrent dermoid cyst in the prepontine cistern, (►**Fig. 4**). At 4-year follow-up, the mass demonstrated no further growth and the patient reported complete symptom resolution. She continues to be followed with serial imaging to monitor cyst size. Informed consent to publish this case was obtained from the patient at the most recent follow-up visit.

Methods

Data Sources

A comprehensive search of the PubMed, Web of Science, and Scopus databases was independently performed through January 2017 by the first and second authors. Articles related to intracranial dermoid cysts were identified using search terms "dermoid cyst" AND "intracranial." A second independent query was performed in a similar fashion to identify cases of prepontine dermoid cysts using search terms "dermoid cyst" AND "prepontine." After removing duplicates, title and abstracts were screened for relevant articles.

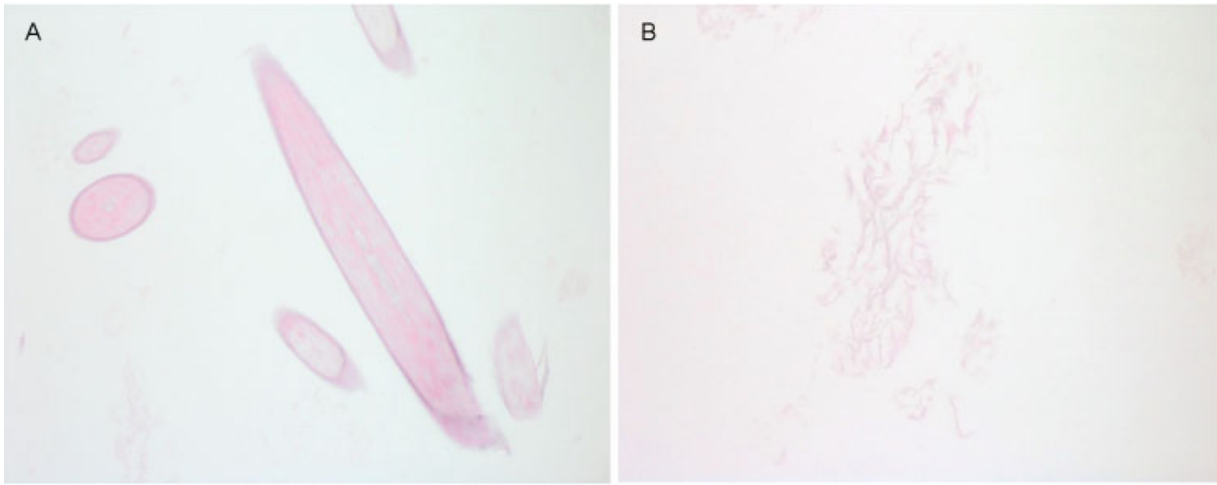

Fig. 3 Hematoxylin and eosin stain of the operative specimen showing the presence of (A) hair shafts and (B) keratin debris, which are consistent with a dermoid cyst.

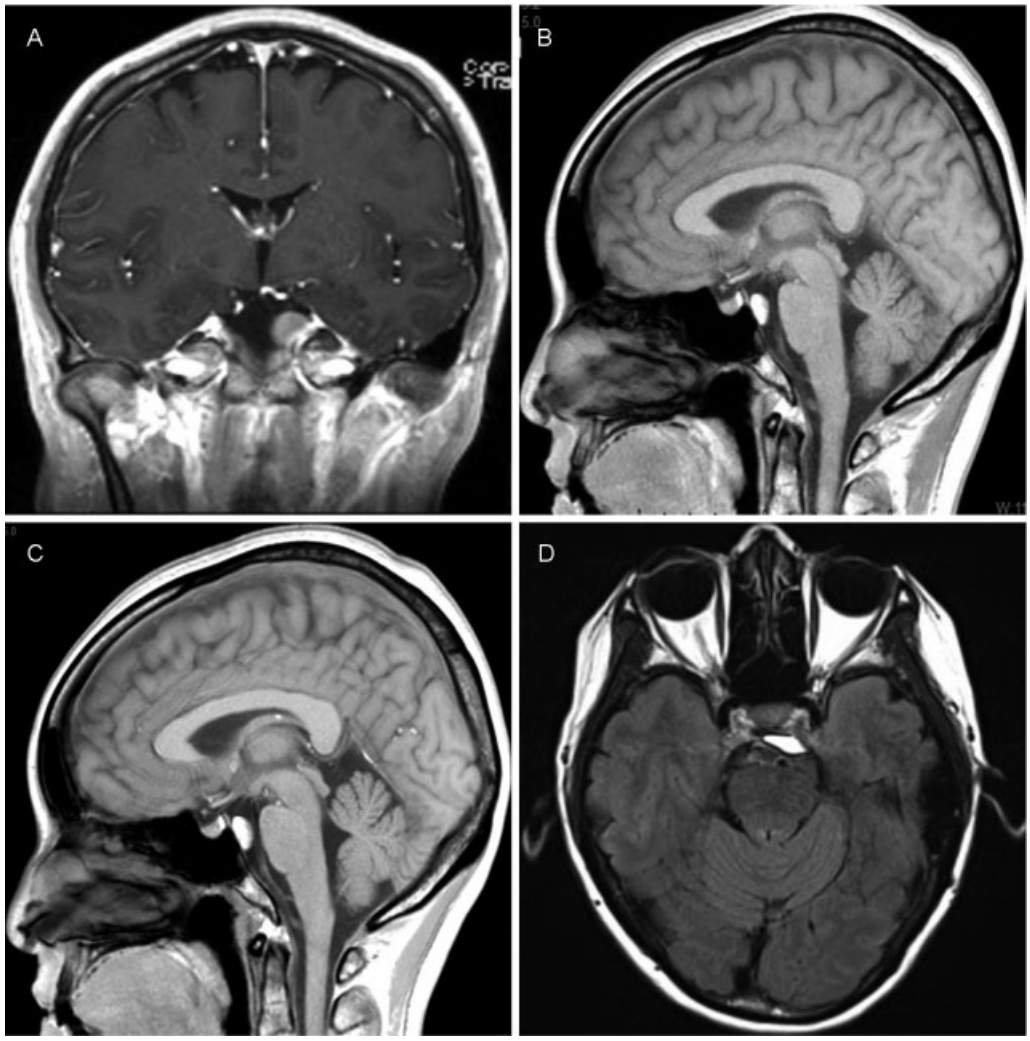

Fig. 4 Postoperative imaging for the illustrative case demonstrating what appears to be a recurrent dermoid cyst in the prepontine cistern, (A) coronal T1-weighted MRI, (B) and (C) sagittal T1-weighted MRI, and (D) axial T2-weighted MRI. MRI, Magnetic resonance imaging.

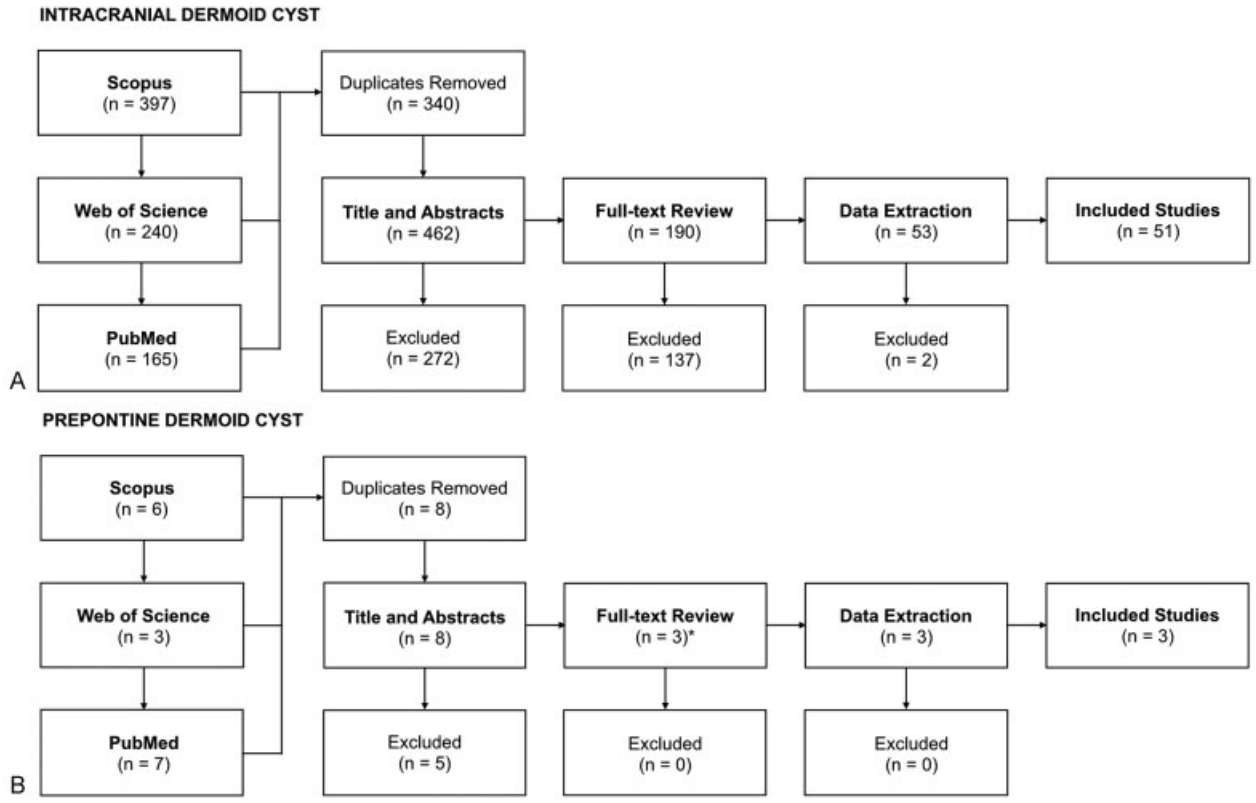

Fig. 5 Schematic diagrams of the search strategies for (A) intracranial dermoid cysts and (B) prepontine dermoid cysts. *All articles were reviewed regardless of exclusion criteria.

Schematic diagrams of the search strategies are provided in ►**Fig. 5**.

Study Selection

Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guidelines were adhered to throughout this study. English, full-text case reports and series were included. Non-English, abstract-only text, and review articles were excluded. Studies with insufficient outcomes data or those involving dermoid cysts with extradural components, malignant transformations, sinus tracts, and/or associated with clinical syndromes were eliminated. Studies without mention of histopathological or gross confirmation of the dermoid cyst were further excluded. However, due to the paucity of prepontine dermoid cyst cases, all relevant studies were reviewed independent of the above-mentioned exclusion criteria.

Data Extraction

Patient demographics, presentations, cyst locations, and surgical outcomes were extracted. Symptom presentation was categorized as cognitive/psychiatric, cranial nerve palsy, headache, hydrocephalus (on imaging), nausea/vomiting, motor (motor weakness or cerebellar signs), seizure, visual disturbances (diplopia, decreased acuity, or nystagmus), or other. The included studies are summarized in ►**Table 1**.

Results

A total of 69 patients were identified in 54 studies published between years 1977 and 2016.[1,4,11,12,18–67] The mean age was 33.3 years. Males comprised 50.7% of the cases. Headache was the most common presenting symptom (52.2%), followed by visual disturbances (33.3%). Thirty-four (49.3%) patients experienced cyst rupture prior to presentation. Intracranial dermoid cysts were equitably distributed throughout the cranial vault, with 29.0%, 36.2%, and 29.0% located in the anterior, middle, and posterior cranial fossae, respectively. Anterior cranial fossa dermoid cysts were predominately located in the Sylvian fissure, middle cranial fossa dermoid cysts in the sellar region, and posterior cranial fossa dermoid cysts in the cerebellar vermis. The interpeduncular fossa (2.9%) and other locations (2.9%) were also described.

All patients underwent surgical resection. GTR was achieved in 42.0% of cases. Thirty-four (49.3%) patients achieved symptom resolution, while 15 (21.7%) patients experienced only partial symptom improvement. Two (2.9%) patients died of either surgical or post-surgical complications. There were four (5.8%) recurrences at a mean follow-up of 2.1 years. Patient characteristics, management, and outcomes data are detailed in ►**Table 2**.

Discussion

Dermoid cysts are benign, congenital tumors primarily filled with lipid components, desquamated cells, and keratinaceous debris.[1,4,68,69] These cysts are diagnosed most frequently between the third and fifth decades of life, a finding supported by the cases we reviewed.[1,10,69,70] We found no sex predominance for intracranial dermoid cysts,

Table 1 Summary of studies included in review

Study and year	Age, years	Sex	Presentation	Rupture	Location	EOR	Outcome	Recurrence	FU, years
Ageshio 2013[22]	52	M	CNP	N	MF	GTR	NR	NR	NR
Aggarwal 2014[23]	8	F	C/P, HCP, visual	N	PF (fourth ventricle)	GTR	NR	NR	NR
Akdemir 2004[1]	18	M	CNP, HA, visual	N	MF (sellar)	GTR	Improved	N	0.01
Anand 2014[24]	30	F	HA, Sz	N	AF (Sylvian fissure)	NR	Improved	N	1.00
Berginer 1988[25]	26	M	HCP, Sz, other	N	MF (sellar)	STR	Improved	N	2.00
Bizzozero 1992[26]	20	F	HA, HCP, N/V	N	PF (fourth ventricle)	GTR	Improved	N	1.50
Brown 2001[27]	18	F	C/P, HA, HCP, visual, other	N	PF (cerebellar vermis)	GTR	Partial	N	0.67
Caldarelli 2001[28]	16	F	CNP	N	PF (brainstem)	STR	Improved	N	5.00
Chen 2005[29]	61	F	C/P, CNP, motor	N	AF (Sylvian fissure)	STR	Improved	N	1.00
Ciurea 2005[30]	4	M	CNP, HA, N/V, visual	N	PF (brainstem)	NR	Improved	N	3.00
Detweiler 2009[31]	58	M	C/P, visual	Y	AF	GTR	Partial	N	0.67
Ecker 2003[32]	23	F	HA, N/V, other	Y	AF (Sylvian fissure)	STR	Partial	N	0.58
Eren 2003[33]	3	M	HA	N	PF (cerebellar vermis)	GTR	NR	NR	NR
Ford 1981[34]	26	M	Motor, N/V, other	Y	AF	NR	Improved	N	0.17
Garces 2016[18]	61	F	Sz	N	AF (Sylvian fissure)	GTR	Improved	N	2.50
Geyik 2016[35]	12	M	HA, HCP, visual	N	PF (fourth ventricle)	GTR	Partial	N	NR
Hahn 1986[36]	16	F	HA, motor	Y	MF (sellar)	NR	NR	NR	NR
Johnson 2005[37]	42	M	C/P, HA	Y	AF (frontobasal)	STR	Partial	N	0.42
Jyoti Das 2009[38]	48	M	C/P, HA, N/V, other	Y	AF (frontobasal)	NR	NR	NR	NR
Karabulut 2000[39]	26	M	HA, N/V, other	Y	IF	NR	NR	NR	NR
Kim 2008[40]	29	M	HA, other	Y	MF (sellar)	GTR	Partial	N	3.00
Klonoff 1990[41]	43	F	C/P, visual, other	N	MF (sellar)	STR	Partial	N	4.83
Koh 2012[42]	17	F	Other	Y	PF	GTR	NR	NR	NR
Lee 1977[43]	43	F	HA, N/V, Sz	N	AF (frontobasal)	NR	Improved	N	0.17
Li 2011[4]	14	M	HA, HCP, motor, other	N	PF (cerebellar vermis)	GTR	NR	NR	NR
Li 2012[44]	14	M	Sz	Y	AF (Sylvian fissure)	GTR	Improved	N	0.25
Liu 2008[45]	57	M	Sz	Y	AF (Sylvian fissure)	STR	Improved	N	5.00
	25	F	HA, visual	Y	MF (sellar)	GTR	Improved	N	1.00
	36	M	C/P, HCP, visual, other	Y	MF (sellar)	STR*	Partial	Y	9.00
	35	M	CNP, HA, HCP, N/V, visual	Y	AF (Sylvian fissure)	STR	Improved	N	11.0
Luan 2012[46]	28	F	Other	N	PF (cerebellar vermis)	GTR	Improved	N	2.50
Mamata 1998[47]	63	F	CNP	N	MF (sellar)	NR	NR	NR	NR
Maulsby 1980[67]	12	F	HA, visual	Y	PF (cerebellar vermis)	STR	Death	NR	0.02
McCoul 2012[48]	50	M	HA	N	MF (sellar)	GTR	Improved	N	3.00
Nakamura 2001[49]	15	F	CNP, HA, motor	Y	MF (sellar)	GTR	Improved	N	1.00
Neugroschl 2002[50]	73	M	Motor, other	N	PF (cerebellar vermis)	GTR	Partial	N	NR
North 1993[51]	4	M	Visual	Y	Other	NR	Partial	N	1.00
Orakcioglu 2008[52]	19	M	C/P, HA, Sz	Y	AF (frontobasal)	NR	Partial	N	9.00
	33	M	Sz	Y	Other	STR	Improved	N	1.50
	16	F	HA	Y	MF (sellar)	STR	Improved	N	2.00
	60	M	CNP, HA, N/V, motor, other	N	PF (cerebellar vermis)	GTR	Improved	N	0.25
	56	M	C/P	Y	MF (sellar)	GTR	Improved	N	0.60
	52	M	Sz, visual, other	N	MF	GTR	Improved	N	0.50
Pant 2008[53]	12	F	HA, N/V, Sz	N	PF (cerebellar vermis)	STR	Improved	N	1.00
Park 2012[54]	28	F	HA, other	Y	MF (sellar)	STR*	Partial	Y	1.67
Plans 2006[55]	53	M	C/P, HCP	Y	PF	NR	Partial	N	0.50
Prahbu 2009[19]	30	F	CNP, motor	N	PF (prepontine)	NR	Improved	NR	NR
Riordan 2016[56]	31	F	CNP	N	MF	GTR	Partial	NR	0.08

Table 1 (*Continued*)

Study and year	Age, years	Sex	Presentation	Rupture	Location	EOR	Outcome	Recurrence	FU, years
Schneider 2012[11]	14	F	HA, HCP	N	AF	GTR	Improved	N	0.50
	11	M	C/P, HA, other	N	AF (Sylvian fissure)	NR	Improved	N	1.00
Skorvlj 2014[57]	51	M	HA, visual	Y	AF (Sylvian fissure)	NR	Partial	NR	1.00
Smith 1991[58]	23	F	HA, visual	Y	MF (sellar)	NR	NR	N	0.06
	24	F	Visual	Y	MF (sellar)	NR	NR	Y	16.0
	43	M	HA, Sz, visual	Y	MF (sellar)	NR	NR	N	1.00
	35	M	Motor, visual	Y	MF (sellar)	NR	NR	N	0.50
	49	M	Sz	Y	MF (sellar)	NR	NR	N	0.50
	45	F	Sz, other	Y	AF	NR	NR	N	1.00
	80	F	Motor	Y	AF (Sylvian fissure)	NR	Death	NR	0.04
Sood 2014[59]	48	M	HA, Sz	Y	AF (frontobasal)	GTR	NR	NR	NR
Stendel 2002[12]	16	F	CNP, motor	N	MF (sellar)	GTR	Improved	NR	NR
Sturiale 2009[60]	37	F	CNP, other	N	MF	GTR	Improved	N	1.00
Tan 2015[61]	52	F	HA, visual	N	MF (sellar)	NR	NR	N	0.25
Tanabe 2016[20]	63	F	CNP	N	PF (prepontine)	GTR	Improved	NR	NR
Titlic 2008[21]	41	M	HA, N/V, HCP	NR	PF (prepontine)	NR	NR	NR	NR
Tun 2008[62]	41	F	HA, visual	N	IF	STR	Improved	N	1.00
van Calenbergh 2004[63]	4	F	CNP, visual	N	PF (brainstem)	DR*	Improved	Y	3.92
Wani 2016[64]	30	M	HCP, motor, visual	Y	PF (cerebellar vermis)	NR	Improved	NR	1.00
Wang 2013[65]	59	F	HA, visual	N	MF	GTR	Improved	N	2.00
Yadiz 2015[66]	53	M	HA, other	Y	AF (Sylvian fissure)	GTR	Improved	N	0.25

Abbreviations: AF, anterior fossa; C/P, cognitive and/or psychiatric; CNP, cranial nerve palsy; DR, drainage; EOR, extent of resection; F, female; FU, follow-up; GTR, gross total resection; HA, headache; HCP, hydrocephalus; IF, interpeduncular fossa; M, male; MF, middle fossa; N, no; N/V, nausea and/or vomiting; NR, not reported; PF, posterior fossa; STR, subtotal resection; Sz, seizure; Y, yes.
*Patient had reoperation.

an observation first described in an earlier study.[16] These slow-growing cysts have variable clinical manifestations and are commonly treated with surgery.

Theories of Development

Theories regarding the development of dermoid cysts acknowledge the presence of ectopic tissue in the central nervous system. However, iatrogenic dermoid cysts have been reported to arise after procedures that introduce epithelium into areas adjacent to neural tissue (e.g., lumbar puncture or percutaneous aspiration of subdural hematoma).[8] The most widely supported theory involves ectopic ectoderm inappropriately sequestered in the neural tube during closure between the third and fifth weeks of gestation.[2,8,10,12–15,32,55,68,69,71] Evidence supporting this theory includes an affinity of dermoid cysts to develop along the midline.[6,8–12,15,68,69] Dermoid cysts have also been associated with congenital anomalies (e.g., Klippel–Feil syndrome or agenesis of the corpus callosum).[2,3,13] Others have hypothesized that mesodermal or mesenchymal cells are necessary during the inclusion of the ectoderm.[2,5,11,12] While not as supported as the *a priori* theory, this mode of development is consistent with laterally positioned (e.g., cerebellopontine angle [CPA] or Sylvian fissure) intracranial dermoid cysts. Lateral and supratentorial dermoid cysts may also represent displaced midline dermoid cysts.[18]

Reports of malignant transformation of intracranial dermoid cysts have also been described, though at much lower rates than intracranial epidermoid cysts.[72–74] Only four cases of malignant transformation of intracranial dermoid cysts have been reported in the literature thus far.[75–78] These cases were associated with either older age (mean age of 50) or recurrence from incomplete capsule removal during prior surgery.

Clinical Presentation

Headaches, seizures, and symptoms of increased intracranial pressure are most common, but other signs and symptoms may manifest depending on the location of the cyst.[1,2,6–8,10–14,32,71] The majority of patients have cranial neuropathies that affect extraocular movements or facial sensation. Visual impairment and trigeminal paresthesia/neuralgia were also common in the cases we reviewed. In contrast, cyst rupture often presents as acute or recurrent meningitis.[4,38–40,54,66,67] Ruptured cysts may also result in unusual presentations, such as hallucinations or cheiro-oral syndrome (thalamic lacunar syndrome resulting in sensory deficits of the fingers and mouth).[31,49]

Asymptomatic patients have also been described in the literature. These patients are often diagnosed incidentally during routine imaging after trauma.[46,68] Traumatic rupture of dermoid cysts may also present as an acute onset headache

Table 2 Summary of patients ($n = 69$)

Age, years	
Mean	33.3
Median	30.0
Range	3–80.0
Sex, *n* (%)	
Male	35 (50.7)
Female	34 (49.3)
Presentation, *n* (%)	
Headache	36 (52.2)
Visual disturbances	23 (33.3)
Cranial nerve palsy	15 (21.7)
Seizure	14 (20.3)
Hydrocephalus	12 (17.4)
Motor/cerebellar	12 (17.4)
Cognitive/psychiatric	12 (17.4)
Nausea/vomiting	11 (15.9)
Other	20 (29.0)
Rupture, *n* (%)	
Yes	34 (49.3)
No	34 (49.3)
Not reported	1 (1.4)
Location, *n* (%)	
Anterior fossa	20 (29.0)
Sylvian fissure	11 (15.9)
Frontobasal	5 (7.2)
Middle fossa	25 (36.2)
Sellar	20 (29.0)
Posterior fossa	20 (29.0)
Cerebellar vermis	9 (13.0)
Fourth ventricle	3 (4.3)
Brainstem	3 (4.3)
Prepontine	3 (4.3)
Interpeduncular fossa	2 (2.9)
Other	2 (2.9)
Extent of resection, *n* (%)	
Gross total	29 (42.0)
Subtotal	15 (21.7)
Drainage of cyst only	1 (1.4)
Not reported	24 (34.9)
Outcomes, *n* (%)	
Symptoms improved	34 (49.3)
Symptoms partially improved	15 (21.7)
Death	2 (2.9)
Not reported	18 (26.1)
Recurrence, *n* (%)	
No	46 (66.7)
Yes	4 (5.8)
Not reported	19 (27.5)

or meningismus.[54] Patients often remain asymptomatic until spontaneous cyst rupture, or until the mass becomes 3 cm in size.[1,6,7,10,68] A correlation between intracranial dermoid cyst size and patient age is supported by the literature.[5,9,10,70] Specifically, patients may not present until the third or fifth decade of life simply because the cyst has not had enough time to reach a size large enough to cause significant mass effect or rupture.[5,9,10,70]

Radiologic Features

Dermoid cysts appear as non-enhancing heterogeneous masses, a finding that is consistent throughout the literature.[7,9,11,14,15] The heterogeneity is attributed to the variable secretions and desquamations the cyst may contain.[1,4,10,13,71] An intensely enhancing dermoid cyst may suggest the presence of malignant transformation.[77] As dermoid cysts are primarily fat-filled capsules, CT imaging most often displays a sharply defined hypodense mass.[1,4,7,9–15,71] Punctate hyperdensities may be visualized and are consistent with calcification of cystic components.[1,4,10,13,71]

Conversely, dermoid cysts appear more variable on MRI. The literature most commonly describes dermoid cysts as displaying T1-hyperintensity and T2-hypointensity.[4,10,27,52] However, heterogeneous signal intensity on T2-weighted MRI is frequently reported, ranging from hypo- to hyperintense and is attributed to the variable density of the fatty components within the cyst.[1,9,32] T1-hypointensity and T2-hyperintensity have also been reported in patients with dermoid cysts, particularly in the solid components of these cysts.[71] Linear peripheral enhancement may also be seen with gadolinium administration.[22] Rupture of cystic contents produces marked hyperintensity on both T1- and T2-weighted MRI, and often with the tell-tale sign of fat-like droplets in the subarachnoid space or ventricular system.[1,9,71] This finding may be useful when determining the location of ruptured cystic components for removal.

Dermoid cysts rarely restrict on diffusion-weighted imaging (DWI).[9] This key feature can be used to differentiate dermoid cysts from epidermoid cysts, which display intense diffusion restriction. However, dermoid cysts may contain thick or solid components that decrease diffusion; thus, mimicking epidermoid cysts.[79] Diffusion sequences were only present in 10 of the articles we reviewed.[11,18,19,22,35,37,52,53,59,66] In those cases, DWI displayed minimal to moderate restriction. A diffusion scan for our patient, at recurrence, demonstrated minimal restriction (►**Fig. 2D**).

Location

Dermoid cysts are commonly found along the midline, a location supported by their theory of development as entrapped ectodermal cells within the growing neural tube.[28,45] Prior studies have reported an infratentorial predilection, particularly near the cerebellar vermis and the fourth ventricle.[16,37] Surprisingly, the incidence of supratentorial dermoid cysts has increased in recent decades.[1,17,80] Supratentorial dermoid cysts are frequently found in the sellar, temporal, and frontobasal regions, and less commonly in the cavernous sinus.[28] The intracranial dermoid cysts we

reviewed were almost equally distributed between the anterior, middle, and posterior cranial fossae; however, there was a clear predilection for specific anatomical regions. Supratentorial dermoid cysts were overwhelmingly located in the sellar region and Sylvian fissure, while infratentorial dermoid cysts were predominately located near the cerebellar vermis. Although Sylvian fissure dermoid cysts have been previously reported to be exquisitely rare, our review suggests that Sylvian fissure dermoid cysts may be relatively common.[18]

Management

Recurrence following subtotal resection was rare, with very few cases identified in our review of the literature.[41,63,81,82] Complete resection may not be possible when the capsule is adherent to neurovascular structures or vital areas (e.g., brainstem). GTR may be associated with greater surgical morbidity.[1] Therefore, the goal in surgical decision-making should be safe, maximal resection, rather than complete resection.[12,13,83]

Surgical approach is dependent on the location of the cyst. Most middle fossa dermoid cysts were resected via a pterional craniotomy.[11,12,32,69,71] Posterior fossa dermoid cysts were often resected via a suboccipital craniotomy.[3,4,10] CPA dermoid cysts were also resected via a standard retrosigmoid approach.[55,84] It must be considered that the chosen trajectory may limit visualization of cystic components, especially those extending into multiple compartments.

The illustrative case herein described highlights the advantages and disadvantages of several approaches to resecting a prepontine dermoid cyst. Intracranial dermoid cysts may be relatively large and involve multiple compartments. In our patient, the dermoid cyst was located in the prepontine cistern and extended into Meckel's cave. It was the senior author's opinion that the bulk of the mass was located near the CPA and, thus, was accessible via a retrosigmoid approach. The retrosigmoid approach allows for maximal exposure of CPA tumors, with direct visualization of cranial nerves V to XI.[85] However, this approach may not be optimal for tumors extending into the petroclival or anterior brainstem region due to the need for increased cerebellar retraction and a long surgical corridor. For these tumors, an anterior and posterior transpetrosal approach may be required. The combined transpetrosal approach provides optimal exposure to zones I to II of the petroclival region, from the dorsum sellae to the jugular tubercle, with minimal need for brainstem retraction.[85] This short, flat route to the anterior brainstem comes at the expense of increased temporal bone removal. Another option is an orbitozygomatic approach, particularly for sellar masses that extend laterally into the cavernous sinus or posteriorly into the cerebral peduncle.[85] However, unfamiliarity with this approach may result in a surgeon favoring a pterional craniotomy.

Medical management may be attempted prior to definitive treatment or in cases not amenable to surgical resection. Of the cases we reviewed, a single patient was managed expectantly, although surgery was eventually performed after serial imaging demonstrated cyst growth in the setting of worsening symptoms.[11] In a single study, carbamazepine was used as the primary treatment in two patients with CPA dermoid cysts presenting as trigeminal neuralgia.[86] Sustained symptom resolution was achieved in both patients at the last follow-up. However, the literature overwhelmingly supports surgical resection as the first-line treatment.

Pathology

Dermoid cysts are most often reported as capsules filled with either a yellow or white substance, with a wide range of viscosities. Dermal appendages to surrounding structures are not uncommon.[8,11] In our patient, microdissection was necessary to safely remove as much of the adherent portions as possible. Closer pathological examination reveals a capsule lined with stratified squamous epithelium.[1,5,7,12,15,69,71] The cystic components contain secretions from sebaceous glands, desquamated material from epithelial cells, keratin, hair follicles, and even teeth, which may explain the common misdiagnosis of teratoma.[1,4,5,7,9,11,12,15,69,71] Teratomas, by definition, must contain non-cutaneous material and can be ruled out by histopathologic evaluation.[12]

Differential Diagnosis

Although histopathological confirmation remains the gold standard in diagnosing dermoid cysts, imaging modalities may be useful in excluding other differential diagnoses. The spectrum of congenital cystic masses includes epidermoid cysts and teratomas.[87] Other potential diagnoses may also include arachnoid cysts and cholesterol granulomas.[9,10,69] Arachnoid cysts may be differentiated from dermoid cysts by the heterogeneous appearance of dermoid cysts on imaging.[9,14,15] Diffusion scans may be used to differentiate epidermoid cysts by their greater degree of restriction as compared to dermoid cysts.[9,14,15] Cholesterol granulomas often show greater inflammatory response on pathological evaluation as compared to dermoid cysts.[9,14,15]

Outcomes

Nine decades ago, postoperative mortality rates for patients with dermoid cysts were as high as 70%.[88] Advancements in microsurgical techniques now allow for safe, maximal resection. Currently, intracranial dermoid cysts are associated with postoperative mortality rates as low as 5%.[80] We identified only two deaths among the cases reviewed, with both cases occurring in the 1980s and one attributed to postoperative pulmonary embolism.[58,67]

Due to the possibility of immunological response from either cyst rupture or surgical removal of dermoid components, patients should be closely monitored for signs and symptoms of aseptic meningitis (Mollaret's meningitis).[1,3,32] Mollaret's meningitis results from meningeal irritation secondary to invasion of the subarachnoid space by oil droplets from a ruptured dermoid cyst.[9,12,15,71] None of the cases we identified reported occurrence of this problem as a consequence of surgical resection. However, one patient experienced delayed ischemic injury from arterial vasospasm after

surgery, which was attributed to dermoid material left in the surgical bed.[32] Thus, proper irrigation of cystic contents during surgery may reduce morbidity by diluting the concentration of dermoid byproducts.

The size of dermoid cysts is related to the growth of epithelium, as well as, the rate of secretion from the glandular components of the capsule.[9,11,12,14,71] Of the patients reviewed, only four recurrences were reported.[45,54,58,63] Three of those recurrences underwent repeat surgery, and all three cases reported favorable outcomes.[45,54,63] Management of the last recurrence was not described.[58] Given that the dermoid capsule represents the metabolically active component of the dermoid cyst, recurrence likely occurs in a time-dependent manner.[89] Although recurrences within a year may be possible, the ectodermal origin of dermoid cysts suggests a slow, linear growth that predisposes individuals to late recurrences.[1,90] Thus, it is plausible that all subtotally resected dermoid cysts may eventually recur if given enough time. Resection of dermoid cysts in deep-seated areas may be limited by incomplete visualization and, therefore, may lead to residual capsule being left behind and a higher likelihood of recurrence.

Conclusion

Intracranial dermoid cysts have variable clinical manifestations. Headache and visual disturbances are most common. Cranial neuropathies were also common. Intracranial dermoid cysts were distributed similarly among the anterior, middle, and posterior cranial fossae but with clear predilections for the Sylvian fissure, sellar region, and cerebellar vermis, respectively. Surgical decision-making should consider the prevalent region where the cyst is located to achieve a safe, maximal resection. Although rates of GTR were moderate, symptomatic relief after surgery was common, and recurrence was rare.

Conflict of Interest

The authors report no conflicts of interest concerning the materials or methods used in this study or the findings specified in this paper.

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