Remote recurrence of craniopharyngioma in the epidural space

Case report

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The case is reported of a 28-year-old man with “ectopic” craniopharyngioma recurring in the epidural space 21 years after the original tumor was resected. Previously described cases of similar remote recurrences as well as some features of the biological behavior of craniopharyngioma are discussed. The rarity of this postoperative complication is addressed.

KEY WORDS • craniopharyngioma • ectopia • tumor recurrence • surgical seeding

Craniopharyngiomas are maldevelopmental tumors of epithelial nature that commonly occur in the suprasellar region. Although the histogenesis of these frequently cystic neoplasms is unclear, well-defined cytoarchitectural features distinguish them from the simpler epithelial cysts arising in this region.6,13,21,25 Typical characteristics range from partially cystic masses containing “machinery oil-like” fluid and the typical adamantinomatous pattern to the more solid, polypoid tumors composed of papillary and trabecular squamous epithelial arrangements. The benign histopathological character of these tumors fails to reflect the well-documented capacity to recur locally.26 Despite this tendency, remote or “ectopic” recurrences are extremely unusual.3,20 We present a case of craniopharyngioma that recurred in the epidural space 21 years after the original tumor was resected.

Case Report

This 28-year-old right-handed white man initially presented at the age of 6 years with complaints of headaches, falls, and a change of personality.

First Admission. The patient's vision was normal, as was a neurological examination at that time. Bilateral carotid angiography and pneumoencephalography revealed a 2.5-cm suprasellar mass.

First Operations. A right frontal craniotomy was performed for partial resection and drainage of a cystic tumor. Histopathological examination confirmed the diagnosis of a craniopharyngioma with areas resembling the adamantinomatous pattern in addition to denser palisades of basaloid cells forming irregular nests and trabeculae. A loosely attached squamoid matrix was often associated with these areas (Fig. 1). Postoperatively, the patient received radiation therapy (2800 rad to the tumor from a cobalt-60 source between October 8, 1969, and June 6, 1970). During this same period, the patient underwent surgery for further partial resection of the tumor. Despite the surgery and the radiation treatment, the patient lost vision in the left eye and developed progressive temporal hemianopsia in the right. This precipitated a third operation at 8 years of age, which involved total resection of the tumor and hypophysectomy. This operation was complicated by panhypopituitarism and seizures, well controlled with mephalobarbitol. The patient subsequently did well, with no recurrence of the tumor as documented by computerized tomography (CT) scans obtained 5, 7, and 9 years postoperatively. His endocrinological status was stable on replacement therapy. His visual deficits remained unchanged with no light perception in the left eye and visual acuity of 20/25 + 2 in the right. He continued to have definite temporal desaturation on the right. He was employed and independent, functioning at the “low average” range of intellectual development at the 15th percentile on the Weschler Adult Intelligence Scale.
Recurrent epidural craniopharyngioma

**Second Admission.** In October, 1990, 21 years after his initial presentation, the patient complained of intermittent frontal and occipital headaches during a routine follow-up visit. A CT scan revealed a partially calcified cystic epidural lesion in the right frontal area (Fig. 2), associated with some scalloping of the inner table and enhancement of its inner margin. A magnetic resonance (MR) image demonstrated a 3.5-cm cystic lesion with a fluid level of high signal intensity on both T₁- and T₂-weighted images (Fig. 3). Incidental changes consistent with postoperative encephalomalacia in the frontal lobes and absence of the anterior clinoid processes were also noted. The differential diagnosis included chronic epidural hematoma or an epidermal/dermoid cyst.

**Second Operation.** On December 14, 1990, a right frontal craniotomy was performed for removal of the cystic mass. At surgery, the cyst contained dark-brown fluid with multiple cholesterol crystals. The wall of the cyst was removed with blunt dissection, and the histopathological features represented a craniopharyngioma with prominent trabeculae of palisaded basaloid cells and nests of squamous cells comprising a less prominent matrix (Fig. 4).

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**Fig. 1.** Photomicrograph obtained in 1969 of the original suprasellar craniopharyngioma. The tumor displays the typical microcystic pattern with spaces lined by palisading basaloid cells. Loosely juxtaposed squamous cells compose the cellular cores of these interconnected trabeculae with a scant stroma. H & E, × 225.

**Fig. 2.** Computerized tomography scan (unenhanced) obtained in 1990 demonstrating a right frontal cystic lesion with calcification of the cyst wall and central portions.

**Fig. 3.** Magnetic resonance (MR) images, coronal view, obtained in 1990. *Left:* T₁-weighted (TR 2.5 msec, TE 90 msec) MR image revealing an extra-axial right frontal mass lesion with high signal intensity. *Right:* T₂-weighted (TR 0.5 msec, TE 20 msec) MR image demonstrating persistent high signal intensity of the mass.

**Fig. 4.** Photomicrographs of the recurrent frontal epidural craniopharyngioma. The tumor has microcysts lined by cuboidal epithelium (left) in juxtaposition to the more typical trabeculae of basal/squamous epithelium (right). Focal nodules of keratin without anuclear squamous cells or any evidence of epithelial surface maturation were occasionally observed within this epithelium. H & E, × 300 (left), × 250 (right).
Postoperative Course. The patient made an uncompli-
cated recovery from surgery and was discharged
home on the 4th postoperative day. At 4½ months after
surgery, he is doing well and is neurologically stable. A
CT scan obtained on the 2nd postoperative day as well
as MR imaging performed 6 weeks after the procedure
confirmed complete removal of the extra-axial mass.

Discussion

Cranioopharyngiomas account for 2% to 3% of all
intracranial tumors,6 but in children this percentage
increases to 5% to 13%.24 They generally occur in a
suprasellar location but can also be intrasellar or extend
into the frontal lobes, temporal lobes, third ventricle,
or posteriorly along the clivus.12,25 Despite their benign
histopathological features, cranioopharyngiomas can be
difficult to remove completely due to their complex
epithelial pattern of growth into adjacent neural struc-
tures.

Rare examples of cranioopharyngioma have been re-
ported in ectopic locations, including the sphenoid
bone,12 paranasal sinuses, nasopharynx,16 cerebel-
lopontine angle,19 and pineal gland.21 In two cases, an
ectopic recurrence of a cranioopharyngioma appeared to
be directly related to a previous surgical procedure:
frontal lobe implantation of cranioopharyngioma by
repeated needle aspirations in one case,3 and ectopic
cranioopharyngioma deposition in the sylvian fissure
following craniotomy for a suprasellar lesion in the
other.21 Cerebrospinal fluid seeding or distal metastases
in cranioopharyngiomas have not been reported.

Our case demonstrates an ectopic cranioopharyn-
ghioma located epidurally, physically separate from the
primary site. As in the other two reported cases, the
most probable explanation is that the tumor grew from
a small "seed" displaced during surgical manipulation.
Presumably, a remnant of tissue was deposited epidi-
rally during one of the three surgical procedures but it
is not clear at which procedure this occurred. If the
recurrence were to be attributed to the first or the sec-
ond operation, one would conclude that the radiation
therapy did not prevent the ectopic growth. While ra-
diation therapy is often considered useful in cranio-
opharyngiomas, it should be kept in mind that the dose
administered to our patient was well below current
standards.24,17,18,27 Radiotherapy is generally consid-
ered effective treatment for cranioopharyngiomas in
terms of survival and also quality of life,1,5,11,17 but total
surgical excision is often recommended when possible.3,4,9,14,15,25,26

Unlike the other two reported cases, the ectopic
tumor recurrence in our patient was primarily cystic.
Its epidural location is also unique. The present case
illustrates that cranioopharyngioma may have a bio-

gical behavior analogous to skin inclusions or to seeding
of basal cell carcinoma associated with intraoperative
tumor manipulation.8,24 This case provides a cautionary
note for the neurosurgeon undertaking the surgical
treatment of cranioopharyngiomas. A residual tumor
mass or a small fragment deposited anywhere in the
surgical field may be sufficient for this benign but
relentlessly growing neoplasm to recur. Careful inspec-
tion and irrigation of the surgical field may prevent this
event.

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References

1. Amacher AL: Cranioopharyngioma: the controversy re-
garding radiotherapy. Childs Brain 6:57-64, 1980
2. Backlund EO: Studies on cranioopharyngiomas. III.
Stereotaxic treatment with intracystic yttrium-90. Acta Chir
Scand 139:237-247, 1973
tion of cranioopharyngioma by repeated needle aspira-
4. Bartlett JR: Cranioopharyngiomas — a summary of 85
cases. J Neurol Neurosurg Psychiatry 34:37-41, 1971
5. Baskin DS, Wilson CB: Surgical management of cranio-
opharyngiomas. A review of 74 cases. J Neurosurg 65:
12-27, 1986
6. Burger PC, Scheithauer BW, Vogel FS: Surgical Pathol-
ysis of the Nervous System and Its Coverings, ed 3. New
secretion of corticotropin-releasing factor as a cause of
Cushing's syndrome. A clinical, morphologic, and bio-
8. Carmel PW: Brain tumors of disordered embryogenesis,
in Youmans J (ed): Neurological Surgery, ed 3. Philadel-
phia: WB Saunders, 1990, pp 3223-3242
9. Carmel PW, Antunes JL, Chang CH: Cranioopharyn-
10. Carpenter RC, Chamberlin GW, Frazier CH: The treat-
ment of hypophyseal stalk tumors by evacuation and
irradiation. AJNR 38:162-177, 1937
11. Cavazzuti V, Fischer EG, Welch K: Neurological and
psychological sequelae following different treatments
of cranioopharyngioma in children. J Neurosurg 59:
409-417, 1983
12. Cooper PR, Ransohof J: Cranioopharyngioma originating
in the sphenoid bone. Case report. J Neurosurg 36:
102-106, 1972
13. Erdheim J: Uber hypophysengangsgeschwulste und Hirn-
cholesteatome. Sitzungb d d Akad der Wissensch. Mathe-
matisch-naturwissenschaftliche Classe 113:537-726,
1904
gement of cranioopharyngioma in children. J Neurosurg
47(2):218-277, 1977
15. Kahn FA, Gosch HH, Seeger JF, et all: Forty-five years
experience with the cranioopharyngiomas. Surg Neurol
1:5-12, 1973
16. Majlessi H, Shariat AS, Katirai A: Nasopharyngeal cra-
nioopharyngioma. Case report. J Neurosurg 49:119-120,
1978
17. Manaka S, Teramoto A, Takakura K: The efficacy of ra-
diotherapy for cranioopharyngioma. J Neurosurg 62:
648-656, 1985
18. Overton MC III, Shellef DD: Recurrent cystic formation
in cranioopharyngioma treated with radioactive chromic
Recurrent epidural craniopharyngioma


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