Clivus Chordoma: A Report of 12 Recent Cases and Review of the Literature

Abstract—Twelve patients with histologically confirmed clivus chordoma were treated at the Johns Hopkins Hospital between 1971 and 1989. Eight of the patients were men and four were women. The mean age at first operation was 51 years (range, 10 to 80). The most common presenting symptoms were headache, diplopia, dysphagia and dysarthria, and facial sensory changes. Computed tomography, with and without contrast enhancement, proved adequate for tumor identification and localization. Magnetic resonance imaging and angiography were occasionally employed to localize the tumors further and to define tumor vascular supply and proximity to vascular structures. Twenty-two resections were performed in 11 patients, and another patient underwent biopsy only. Seven were also treated with radiation therapy. Tumors recurred in eight patients, six of whom underwent further operations. The mean time to first recurrence was 22 months (range 8 to 36 months). Six of the patients are still alive, with a mean follow-up of 31 months (range, 3 to 89 months) from first surgical resection. The mean survival time from first treatment was 31 months (range, 4 to 62 months) among those patients who died. There was no operative mortality. The 5-year cumulative survival rate was 20%. Six patients with long follow-up have had fair to good results, being free of recurrences for at least a year. However, none of the patients returned to their premorbid baseline of activities. Five of the patients had tumors with the histologic diagnosis of chondroid chordoma. Three of these patients are still alive. The mean age at first treatment was 44 (compared with 62 for typical chordomas). The mean time from symptoms to diagnosis was 29 months (typical chordomas, 18 months). The mean length of survival and time to tumor recurrence were not significantly different between chondroid and typical chordomas. (*Skull Base Surgery, Volume 1, Number 4, 1991, p. 200*)

Chordomas are rare, usually benign, neoplasms that are thought to arise from remnants of the embryonic notochord along the spinal axis.¹ Intracranial chordomas usually arise from the region around the clivus and account for about a third of all chordomas in most series.^{2,3} Clivus chordomas seldom metastasize and usually become symptomatic by slow growth and local invasion to affect nearby cranial nerves and brainstem structures. These tumors are difficult to manage because of their critical location and propensity to recur, and they have

been treated in the past with various combinations of surgery and radiotherapy. Over the years, the surgical morbidity has steadily decreased with advancements in technique. However, it is unclear if recurrences or long-term survival have been affected by this change.

Twelve cases of clivus chordoma have been treated at the Johns Hopkins Hospital since 1971, seven of whom had their initial operation within the last 5 years by the joint skull base team composed of a neurosurgeon and head and neck surgeon. In this article we report the clinical presen-

tation, treatment, and outcome of our series of cases, along with comments based on an extensive review of the literature. The effects of recent improvements in management and current expectations for outcome and survival are discussed.

CLINICAL MATERIAL AND METHODS

Twelve patients with histologically proven chordomas involving the clivus and adjacent skull base structures treated at John Hopkins between 1970 and 1989 were involved in the series. Hospital records, surgical pathology reports, and any autopsy reports were obtained on each of the patients, and follow-up data were obtained from clinic records and the Johns Hopkins Tumor Registry. There were eight men and four women. Five of the tumors were determined histologically to be chondroid chordomas, according to surgical pathology reports from the Department of Pathology at Johns Hopkins. One patient had previously undergone surgical resection and stereotactic radiosurgery with a gamma knife elsewhere, 1 year before his operation here. The information on each patient is summarized in Table 1.

RESULTS

Patient Presentation

The mean age at onset of symptoms in our patients was 47 years (range, 8 to 80). This is similar to other series,^{3,4} in which symptoms most commonly began in the fourth and fifth decades of life. Two-thirds of the patients were men. Most large series in the literature have reported either a slight excess of males^{3,5} or an equal number of both sexes.⁶

The mean time from onset of symptoms to diagnosis was 25 months (range, 1 to 60). The most common presenting symptoms in our patients were headache (50%), facial pain, numbness or paresthesias (50%), and diplopia (42%). Other common symptoms, none of which were sensitive or specific for clivus chordoma, included dysphagia and dysarthria, visual complaints, ataxia, extremity weakness, and hoarseness (Table 2). One patient presented at 8 years of age with loud snoring and poor school performance. A nasopharyngeal mass was discovered on otolaryngologic examination, which was initially diagnosed as hypertrophic adenoiditis. On biopsying the lesion, it was found to be a chordoma, and computed tomography (CT) scan revealed a large tumor involving the clivus and sphenoid sinus with bony invasion into the nasopharynx and palate.

The neurologic signs elicited on physical examination at the time of presentation could be divided into those attributable to tumor compression in the region of the basisphenoid, or upper clivus, and the basiocciput, or

lower clivus. Eight (67%) of our patients presented predominantly with basisphenoid signs, which included upper cranial nerve (I–VI) dysfunction. Papilledema, optic nerve atrophy, and pituitary dysfunction have been reported⁷ but were not detected in our patients. Three patients (25%) presented with basioccipital signs, including lower cranial nerve (VII–XII) dysfunction, pyramidal tract abnormalities (including hemiparesis, hyperreflexia, clonus, and Babinski sign) and cerebellar signs. The frequency of presenting signs and comparison with the literature are presented in Table 3.

None of our patients had clinical evidence of tumor metastasis to distant sites. Most series in the literature have reported metastasis rates of 0 to 8% for cranial chordoma.^{5,8} A large series from the Mayo Clinic reported only two cases of metastasis among 55 clivus chordomas.³ Therefore, although metastases have been documented in clivus chordoma, the incidence is quite small.

Radiologic Features

Tumors were usually well visualized and localizable by CT, with or without contrast. CT was also useful for detecting calcification, cystic tumors, and bone erosion. In one case, however, the CT scan showed no abnormality, so a magnetic resonance imaging (MRI) scan was performed, which showed a tumor mass on the right clivus. MRI has also been helpful to clarify further the anatomy and relationship of the tumor to nearby structures. Angiography has also been helpful in several cases to define the vascularity of the tumor and to identify the proximity of the tumor to nearby vascular structures. Others have also found CT to be the radiologic examination of choice for clivus chordomas.⁵

Surgical Treatment

All except one of our patients underwent surgery for resection of the tumor (Table 1). The other one had only biopsy through a transpalatal approach. The initial operations included three midfacial deglovings, two transphenoidal, two infratemporal, one posterior fossa, one bifrontal, and one right frontal approach. Another had subtotal tumor resection through an unspecified craniotomy at another hospital. One patient also underwent C1–C4 laminectomy and tumor debulking from an infratemporal exploration. Only one resection was thought to be total at the time of operation (this patient had a recurrence).

Five patients had repeat operations for tumor recurrence. One patient had two repeat midfacial deglovings, one had a left temporal craniotomy, and one had a right frontotemporal craniotomy followed by reexploration through a right frontoparietotemporal craniotomy. One patient with a previous midfacial degloving underwent midline transpalatal, transpharyngeal approach with man

Table 1. Clinical Summary of 12 Patients Treated for Clivus Chordoma*

Patient	Age (yr)	Sex	Sx to Dx	Initial Symptoms, Signs	Surgical Approach	Radiation	Recurrences	Follow-up†	Histology
1	80	M	5	Headache, rt. central scotoma, unsteady gait, left VI	Transsphenoidal (1986)	None	None	Alive at 22 mo	Typical
2	75	F	48	Diplopia, facial numbness, retro-orbital pain, left VI (dysphagia)	Left subtemporal (1984)	6488 r	1	Dead at 33 mo	Typical
3	64	F	11	Headache, dysarthria, ataxia, facial weakness, bilateral V, IX, X, right peripheral VII, decreased vestibuloocular reflex	Posterior fossa (1982) Left temporal (1985)	None	2	Dead at 62 mo	Typical
4	30	M	1	Blurred vision	Bifrontal & right temporal (1983) Right frontotemporal (1984) Right frontotemporoparietal (1984)	4500 r	1	Dead at 27 mo	Typical
5	68	F	60	Diplopia, retro-orbital headache, left facial paresthesias, bilateral VI	Right frontal (1971) Midfacial degloving (1985)	None	None	Alive at 89 mo	Typical
6	40	M	60	Loud snoring, nasopharyngeal mass	Midline transpharyngeal (1987)	None	2	Dead at 21 mo	Chondroid
7	10	F	24	Left neck and face pain, dysphagia, dysarthria, hoarseness, weight loss, left VII, bilateral IX–XII	Midfacial degloving (1986) Transpalatal biopsy (1979)	None	None	Alive at 3 mo	Chondroid
8	69	M	5			1980 r	None	Dead at 4 mo	Typical
9	21	M	30	Neck pain, headache, dysarthria, dysphagia, left-sided weakness, diplopia, left VI, central VII, bilateral IX–XII	Left infratemporal (1980) Midline transpharyngeal (1982) Left infratemporal (1983) Midline transmandibular (1983)	5600 r	3	Dead at 41 mo	Chondroid
10	72	M	6	Diplopia, right facial pain and numbness, right III, VI (weak rt. gag, rt. tongue atrophy)	Transphenoidal (1988)	None	1	Alive at 8 mo	Chondroid
11	38	M	24	Diplopia, decreased vision in left eye, left facial paresthesia, left VI	Midfacial degloving (1985) Midfacial degloving (1988) Midline transmaxillary (1989)	5360 r	2	Alive at 45 mo	Chondroid
12	45	M	60	Headache, facial pain, partial left III, left VI	Subtotal resection (1988) Midfacial degloving (1989)	40 Gy	1	Alive at 16 mo	Typical

*Sx: signs; Dx: diagnosis; rt: right; mo: months

†Follow-up time from first operation.

*Roman numerals = cranial nerve dysfunction, parentheses = subsequent symptoms and signs.

Table 2. Presenting Symptoms in 12 Patients with Clivus Chordoma and Cumulative Data from the Literature

<i>Symptom</i>	<i>Present Series (%)</i>	<i>From Literature* (%) (range)</i>
Headache	6 (50)	46 (12–87)
Facial pain or numbness	6 (50)	10 (8–14)
Diplopia	5 (42)	52 (14–81)
Dysphagia or dysarthria	3 (25)	12 (7–25)
Other visual complaint	3 (25)	29 (14–63)
Unsteady gait	2 (17)	13 (6–29)
Extremity weakness	1 (8)	18 (4–41)
Hoarseness	1 (8)	5 (3–14)

*There were 196 patients combined from references 4–8, 10–12.

Table 3. Neurologic Signs at Presentation in 12 Patients with Clivus Chordoma and Cumulative Data from the Literature

<i>Clinical Sign</i>	<i>Present Series (%)</i>	<i>From Literature* (%) (range)</i>
Basisphenoid signs		
Cranial nerve VI	7 (58)	52 (14–58)
Cranial nerve V (sensory or motor)	7 (58)	19 (16–33)
Visual acuity or field deficit	2 (17)	28 (11–50)
Cranial nerve III*	2 (17)	24 (19–56)
Basioccipital signs		
Cranial nerves VII–XII	3 (25)	33 (32–57)
Pyramidal tract signs	1 (8)	31 (29–41)
Cerebellar signs (ataxia)	1 (8)	29 (4–50)

*There were 121 patients combined from references 4; 5, 10–12.

*Including ptosis, pupillary defects, and extraocular muscle deficits.

dibulotomy and glossotomy. Another patient underwent a similar transpharyngeal approach, followed by infra-temporal and transmandibular procedures.

Postoperatively, improvements in symptoms were generally modest. One patient underwent midfacial degloving and orbital decompression and subsequently reported improved vision in the left eye, which persisted for 3 years until he had a recurrence with a return of decreased vision in that eye. Another patient had a temporary relief of facial pain, which recurred within weeks after surgery. Most patients had only temporary or subtle improvements in previously existing cranial nerve deficits. This was especially true of patients with lower cranial nerve involvement, and persistent problems with dysphagia, dysarthria, and aspiration were common.

Postoperative Complications

There were no perioperative deaths, although three patients died in the hospital following biopsy or surgery,

due to tumor recurrence. Postoperative complications were uncommon, and usually consisted of small cerebrospinal fluid leaks necessitating serial spinal taps or a lumbar drain. One patient had postoperative seizures in the recovery room following foramen magnum and C1–C4 decompression, which were stopped after 8 hours with anticonvulsant therapy. He later developed pseudomonal pneumonia and died 2 months after his fifth tumor resection of overwhelming peritonitis following subtotal gastrectomy for duodenal ulcer. Postmortem examination revealed a large recurrent chordoma compressing the pons and medulla.

One patient required a tracheostomy and gastrostomy, due to lower cranial nerve dysfunction after a repeat operation through a left temporal craniotomy. She also developed a new right hemiparesis, left sixth cranial nerve palsy, and a small pontine hemorrhage by CT scan. A tarsorrhaphy of the left eye was later required for seventh cranial nerve dysfunction. She later died of tumor recurrence and hydrocephalus. Another patient required a postoperative tracheostomy and prolonged intubation due to lower cranial nerve involvement after a second operation for tumor resection. He had a recurrence 2 months later with lethargy, confusion, and multiple cranial nerve palsies. CT and MRI showed hydrocephalus and tumor compression of the left brainstem. A ventriculoperitoneal shunt was placed and 2 months later he developed deep vein thromboses, necessitating heparin therapy. He subsequently died of intracranial hemorrhages involving the right basal ganglia, sylvian fissure and inferior frontal lobe.

Radiation Treatment

Seven of the patients also underwent radiotherapy. The radiotherapy was administered after surgical debulking in all but one patient, who received 5360 r at an outside hospital before undergoing surgical resection here 13 months later for symptomatic recurrence. The radiation dose ranged from 1980 to 6488 r. Most radiotherapy was by standard external beam irradiation, although one patient received 6488 r of heavy charged particle irradiation at another hospital following subtotal resection. Another patient underwent stereotactic gamma irradiation with a dose of 40 Gy at an outside institution before coming here for surgical resection 13 months later. In addition, two patients had ¹²⁵I seeds implanted in the tumor bed at reoperation for tumor debulking. One of these patients lived an additional 14 months after implantation, and the other lived 4 months.

Outcome

Of the 11 patients with operations for tumor resection and one with transpalatal biopsy, six are still alive, after

a mean follow-up of 31 months, with a range of 3 to 89 months. Among the six patients who have died, the mean length of survival from first operation was 31 months, with a range of 4 to 62 months. The 5-year probability of survival was about 20% (Fig. 1). This was similar to the 23% 5-year progression-free survival rate reported by Fuller and Bloom on 13 recent cases⁹ and probably represents the best current prediction for survival.

Recurrences

All but two patients experienced tumor recurrences, among those who lived or had follow-up for greater than 1 year from their first operation. Tumor recurrence was defined as a return of symptoms and signs, with verification of tumor regrowth radiologically. The mean time from initial treatment to tumor recurrence was 22 months. There were five patients who experienced a second recurrence after the second operative resection, and the mean time to second recurrence was 9 months. One patient had a third recurrence 3 months after a third operation.

Chondroid Chordoma

Five of our patients had chondroid chordomas. The mean age at initial treatment was 44 years compared with a mean age of 62 years among patients with typical chordomas. Four had tumor recurrences following their initial surgery, and three had at least one repeat operation. Three patients with chondroid chordoma are still alive with follow-up of 3, 8, and 45 months. One died at 21 months after the first operation, following a second attempt at resection. Another died 41 months after the first operation, following three subsequent operations for tumor debulking.

Figure 1. Cumulative survival probability curve for 12 patients with clivus chordoma and follow-up ranging from 3 to 89 months.

DISCUSSION

Clivus chordoma is an uncommon tumor that is characterized by slow growth and insidious onset of symptoms related to compression of cranial nerves and adjacent brainstem structures. The most common presenting symptoms were headache and diplopia in our series and in the literature.^{5,7,10} Over half of our patients also presented with facial pain, numbness, or paresthesias. Patients usually presented with longstanding symptoms. The mean time from onset of symptoms to diagnosis in our patients was 25 months. In the literature, it is interesting that the mean time to diagnosis or treatment in series published before 1980 was usually around 40 months,^{4,11,12} whereas in two series reported within the last decade this time was around 20 months,^{5,8} which is comparable to our results. This is most likely a result of improved diagnostic methods, CT and MRI scanning in particular, which have allowed earlier and more accurate diagnosis of smaller skull base lesions.

Neurologic signs at presentation reflected the tumor location. Two-thirds of our patients presented with signs attributable to basisphenoid tumors, which tended to present with abnormalities of cranial nerves II–VI. The tumor location in these patients, radiographically and at surgery, was typically in the region of the sella, sphenoid sinus, and upper clivus. These patients can also demonstrate endocrine abnormalities due to pituitary involvement. None of our patients presented with pituitary dysfunction, although one patient developed an elevated prolactin level 9 months after tumor resection. One quarter of our patients presented with basioccipital tumors, which included abnormalities of cranial nerves VII–XII, as well as pyramidal tract and cerebellar signs. These patients had tumors at the lower clivus, cerebellopontine angle, or apical petrous bone. With subsequent recurrences, patients tended to develop symptoms and signs suggesting progressive involvement of additional cranial nerves and brainstem structures. This was particularly true of patients who initially presented with upper cranial nerve signs, then later developed lower cranial nerve and brainstem involvement.

Clivus chordomas with histologic features of chondroid differentiation have been reported to have a more indolent course and better prognosis than typical chordomas.³ We had five such patients in our series and they tended to present at an earlier age (44 years versus 62 years for typical chordomas) but had no fewer recurrences or longer survival than typical chordomas. Chetiyawardana¹³ also failed to find a more favorable outcome among chondroid chordomas, although Rich et al⁸ reported a longer mean survival among four chondroid patients, and Raffel et al⁵ found a significantly decreased incidence of recurrences and mortality among six of these patients. These conflicting data could be due to variable criteria for the histologic diagnosis of chondroid chordoma among different institutions, or due to other variables, such as

age, which have not been adjusted for in comparing survival data. A larger number of chondroid chordoma patients identified with standardized criteria for histologic diagnosis will need to be followed before the natural history of this histologic variant is clear.

Despite their usually benign histologic features, clivus chordomas have historically been a neurosurgical enigma because of their difficult location and propensity to recur despite extensive resection. Untreated, the natural history of clivus chordoma is dismal, with a mean survival of less than 1 year in one study from the Swedish National Series.¹⁴ In those patients who underwent surgical resection as late as the 1950s, perioperative deaths were very common,^{11,15} due to the proximity of the tumors to critical brainstem structures. With the development of microsurgical techniques, operative mortality has been dramatically decreased and more radical tumor excisions have been realized. Despite these changes, the long-term survival of these patients has not been improved. Our mean length of survival after first operation, among patients who have died, was 31 months. Among reports on clivus chordoma since 1980, Reddy et al¹⁶ stated a mean survival of 56 months, although the mean patient age was only 34 years, compared with 51 for our series. Fuller and Bloom⁹ had a mean age of 46 years, and the mean survival was 34 months, similar to our results. Thus, the longer survival reported by Reddy et al¹⁶ may have been due to a younger patient population. Among earlier series, Kamrin et al¹⁰ reported a mean survival of 40 months in 1964 (mean age in mid 30s), Falconer et al¹² reported a 19-month mean survival in 1968 (Mean age, 45 years). Therefore there were probably age factors to account for apparent differences in survival among reported series, but there has been no clear trend toward increased long-term survival in recent years.

Symptomatic tumor recurrences were very common in our series, even in cases in which resection was thought to be extensive or total. This has also been experienced by other groups, who have consistently reported recurrence rates of over 50% with no downward trend in recent years.^{3,5,10,11,16} The mean time to first recurrence was 22 months, which was not improved from the 24 months reported by Poppen and King¹¹ in 1952. Among these patients who have more than one recurrence, the time to second recurrence (mean, 9 months) was always shorter than the time to initial recurrence. Interestingly, there was a distinct correlation between the time to first recurrence and the time to second recurrence. Those patients who had longer initial symptom-free intervals also had longer times to recurrence after the second operation. This observation may reflect biologic properties of the individual tumors, such as their growth rate and proximity to critical neural structures.

Radiation therapy as an adjunct to surgery was suggested by Dahlin and MacCarty¹⁷ in 1952. Heffelfinger et al¹³ showed longer survival times in patients with surgery and radiation than with radiation alone. Kamrin et al¹⁰

suggested that larger tumor doses, in excess of 5000 r, lead to more prolonged remissions than smaller doses. Among deceased patients in our series who received less than 2000 r, one patient received no radiotherapy and lived 62 months from her initial operation, but the other two lived only 4 and 21 months. In contrast, three patients received 4500 r or more and had a mean survival of 34 months (range, 27 to 41). Those patients who had larger radiation doses did not, however, have longer symptom-free intervals. Therefore, radiation therapy in excess of 4500 r, in addition to surgical resection, appeared to improve long-term survival modestly but did not lengthen the time to tumor recurrence. Raffel et al⁵ also found no correlation between radiation dose and the length of the symptom-free interval and recommended a minimum dose of 5500 r. In an attempt to deliver higher radiation doses with acceptable neurologic sequelae, heavy particle proton beam irradiation has been used and found to be effective.¹⁸ In a recent update of patients being followed for heavy particle irradiation following surgery, the 5-year actuarial local control rate was 82%.¹⁹ One of our patients received 6488 r by proton beam following surgery, and she lived 33 months.

Our approach to management of clivus chordomas has been radical tumor excision, followed in most cases by radiotherapy. Total gross tumor removal is the goal of surgery. Recent preferences for surgical access have been the midline approaches, including transsphenoidal resection, midfacial degloving, and transpharyngeal/transpalatal procedure with or without mandibulotomy and glossectomy. A combined neurosurgery and head and neck surgery team was employed for all of these approaches, and with microsurgical techniques the perioperative morbidity was acceptable. Furthermore, the latter two approaches allowed more complete excision of midline tumors. Postoperative radiotherapy with standard external beam irradiation of at least 5000 r is desirable. Local tumor control has clearly been more successful with heavy particle irradiation than previous modalities, but there are only a few institutions that currently have this capability.

Currently, the best management of patients with clivus chordoma is radical surgical removal of the tumor, followed by high-dose radiation therapy. Even with this radical approach, the recent outlook for management of clivus chordoma has been dismal, with further advances in surgical technique unlikely to lead to better long-term results. However, the newly developed proton beam radiation technology promises to improve the lot of these patients. Whether stereotactic radiosurgery or other innovations will also lead to future advances in the management of clivus chordoma remains to be seen.

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REVIEWER'S COMMENTS

This article records in considerable detail the progress of 11 cases of clivus chordoma, one of the most refractory problems to confront the skull base surgeon. Important points borne out by this study are that treatment is worthwhile but that to be effective it requires radical surgery followed by a full dose of radiotherapy. The multiplicity of the surgical approaches used demonstrates that we are still a long way from the ideal treatment for this tumor.

What is surprising is the infratemporal fossa approach of Fisch was not used more extensively in this series.

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