Surgical Treatment of Chordoma and Chondroma of the Skull Base

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A "chordoma" affecting the basisphenoid or basioccipital bones is rarely reported, mainly because of the feeling that little can be done to help these patients either by operation or by radiation therapy. Forti and Venturini could find only 240 cases in the world literature, while Schisano and Tovi found only 10 cases in Olivecrona's series of 6700 cases of verified brain tumor, an incidence of 0.15%. Zoltán and Fényes, from Budapest, surveyed all the operated cases published in the literature up to 1959 and concluded that "the diagnosis of cranial chordoma is extremely difficult, its complete surgical removal is impossible, and the prognosis is hopeless." Kamrin, et al., stated that "except for emergency situations demanding surgical decompression for the preservation of life or of vital structures, surgical treatment of non-sacroccygeal chordoma has little to offer."

We are reporting a small series of nine cases seen in this unit since 1953, because at least five of these patients showed sufficient improvement after operation to resume their work. Three of these patients, however, have since died. Admittedly the follow-up periods are short (2 to 6 years), but our experience suggests that, provided these cases can be diagnosed preoperatively, palliation by an appropriate surgical procedure can be achieved in a reasonably high proportion of cases.

These tumors appear in either sex commonly in middle life. They tend to occur in, or close to, the midline in either the basisphenoid or basioccipital bone. Luschka was the first to describe them in 1856, while Virchow a year later called them "echondroses physalifora," believing them to be of cartilaginous origin. Ribbert, however, is credited with establishing their origin from notochordal remnants. Since then, an extensive literature on them has been built up from which it appears that the notochord traces a sigmoid-shaped course through the body of the sphenoid and the basilar part of the occipital bone. In sequence, it lies close to the floor of the sella turcica, next in relation to the clivus, and finally passes through the odontoid process of the axis to the vertebral column.

Cranial chordomas can arise from notochordal remnants in any part of their cranial course. Although the notochord itself is a midline structure, the remnants may appear slightly to one side. Therefore, we have found it useful to subdivide the "chordomas" into three groups, as follows:

**Group 1.** Sellar chordomas, arising in relation to the sella turcica

**Group 2.** Parasellar chordomas, situated immediately lateral to the sella turcica

**Group 3.** Clival chordomas, arising in the clivus posterior to the sella turcica.

Other classifications have included a group of "chordomas" projecting into the sphenoid air sinus and even the nasopharynx, but our case material indicates that these are probably extensions of the other groups (as in our Case 3). Our classification is admittedly arbitrary and incomplete, for one group may merge into the other, but it has the merit of paralleling the neurological syndromes presented by these tumors.

We have summarized the salient features of each of our nine cases in Tables 1 and 2 and shall discuss the problems of diagnosis and treatment individually.

**Group 1: Sellar Chordomas and Chondromas**

**Case 1.** A 53-year-old woman was referred to us in 1953 because of bitemporal...
TABLE 1

Clinical features of chordomas

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age, Sex</th>
<th>Symptoms and Signs</th>
<th>Preop Duration of Symptoms</th>
<th>X-Ray Appearances</th>
<th>Pathology</th>
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<td></td>
<td></td>
<td></td>
<td>Plain X-Ray</td>
<td>Arteriography</td>
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Sellar Group

| 1        | 53 F     | chiasmal compression, postmenopausal hypopituitarism  | 3 mos                      | normal             | suprasellar tumor  |
| 2        | 42 F     | chiasmal compression, hypopituitarism                 | 6 yrs                      | irregular, calcified | suprasellar mass   |
| 3        | 56 M     | chiasmal compression, hypopituitarism, nasopharyngeal extension, CSF rhinorrhea, meningitis | 5 yrs                      | extensive bone destruction with irregular calcification | suprasellar mass   |

Parasellar Group

| 4        | 47 M     | optic tract compression, hypopituitarism, unilateral 3rd nerve palsy | 2½ yrs                     | parasellar erosion, no calcification | chordoma           |
| 5        | 23 M     | papilledema, unilateral 3rd, 5th, 6th nerve involvement | 1 yr                       | parasellar erosion, irregular calcification | chordoma           |
| 6        | 45 F     | papilledema, unilateral 3rd, 5th, and 6th nerve involvement, brain stem compression | 9 yrs                      | parasellar thinning, no calcification | chordoma           |
| 7        | 30 M     | optic tract compression, hypopituitarism               | 1 yr                       | parasellar & clival erosion, partially calcified post-clival mass | chordoma           |

Clival Group

| 8        | 51 M     | papilledema, bilateral 6th nerve palsy, unilateral 3rd, 4th, 5th pareses, brain stem compression | 3 yrs                      | parasellar & clival erosion, partially calcified post-clival mass | chordoma           |
| 9        | 54 F     | post-menopausal hypopituitarism, bilateral 6th nerve palsy, unilateral 3rd, 4th nerve pareses & 6th nerve palsy, brain stem compression | 6 yrs                      | parasellar & clival erosion, partially calcified post-clival mass | chordoma           |

hemianopia of 3 months duration. An ossifying spinal meningioma at the T-3 level had been removed 8 months earlier. The menopause had been passed at the age of 49.

Examination. X-rays of the skull showed a normal sella turcica, but air encephalography disclosed that the anterior part of the third ventricle was displaced upward and backward, while left carotid arteriography showed a small uniform tumor "stain" immediately above the sella turcica during the venous phase of filling. A suprasellar meningioma was suspected.

Operation. Through a left frontal craniotomy, a vascular tumor was found encircling the left internal carotid artery; it was deemed irremovable. After operation the patient exhibited a left pulsating exophthalmos and died 3 hours later.

Postmortem examination. The carotid arteries and the optic chiasm were found to be embedded in a soft, pink, friable tumor. Its cells were of widely varying size. The chief type was large and rounded with a single eccentric nucleus and abundant cytoplasm; cytoplasmic vacuoles created the physaliferous appearance characteristic of a chordoma. No mitotic figures were present, and there was little extracellular mucinous material.

Comment. The significance of a recent spinal meningioma eludes us. Even in retrospect it does not seem possible to have made
Chordoma and Chondroma of Skull Base

TABLE 2
Results of treatment

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Pre Cranietomy Treatment</th>
<th>Craniotomy &amp; Result</th>
<th>Post Cranietomy Treatment</th>
<th>Verified Pathology</th>
<th>Present Status</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>—</td>
<td>frontal; irremovable vascular tumor; died 3 hrs postop</td>
<td>—</td>
<td>chordoma</td>
<td>died 3 hrs postop</td>
</tr>
<tr>
<td>2</td>
<td>—</td>
<td>frontal; removal of tumor dome; long convalescence, meningitis</td>
<td>radiation; no improvement</td>
<td>chordoma</td>
<td>died 5 yrs postop; syrinx improved course</td>
</tr>
<tr>
<td>3</td>
<td>transorbital ethmoidectomy, temporary improvement; later radiation, no improvement</td>
<td>frontal; partial intracapsular removal, later more radical intracapsular removal; long convalescence, meningitis</td>
<td>—</td>
<td>chordoma</td>
<td>died 4 mos postop</td>
</tr>
<tr>
<td>4</td>
<td>radiation therapy, no improvement</td>
<td>frontal; biopsy; long convalescence</td>
<td>—</td>
<td>chordoma</td>
<td>died 6 mos postop</td>
</tr>
<tr>
<td>5</td>
<td>—</td>
<td>lateral; intracapsular removal; marked improvement</td>
<td>—</td>
<td>chordoma</td>
<td>alive 4 yrs postop; active, working</td>
</tr>
<tr>
<td>6</td>
<td>—</td>
<td>lateral; intracapsular removal; marked improvement</td>
<td>radiation; no determinate value</td>
<td>chordoma</td>
<td>alive 4 yrs postop, improved</td>
</tr>
<tr>
<td>7</td>
<td>—</td>
<td>lateral; intracapsular removal; marked initial improvement</td>
<td>—</td>
<td>chordoma</td>
<td>died 2 yrs postop after recurrence &amp; 2nd operation</td>
</tr>
<tr>
<td>8</td>
<td>—</td>
<td>lateral; intracapsular removal; marked initial improvement</td>
<td>—</td>
<td>chordoma</td>
<td>died 2 yrs postop after 2nd operation</td>
</tr>
<tr>
<td>9</td>
<td>radiation therapy no improvement</td>
<td>lateral; intracapsular removal; marked improvement</td>
<td>—</td>
<td>chordoma</td>
<td>alive 3 yrs postop, active housework</td>
</tr>
</tbody>
</table>

the correct preoperative diagnosis in this case.

Case 2. A 42-year-old woman, referred to us in 1959, gave a 6-year history of intermittent headache and amenorrhea, with recent gain in weight.

Examination. Bitemporal hemianopia with bilateral optic atrophy was found. Skull x-rays showed an irregularly spherical, partially calcified mass, 4 to 5 cm in diameter, projecting upward and backward in the midline from the dorsum sellae, which was thinned (Fig. 1). The clivus was normal. Air encephalography showed an upward and backward displacement of the third ventricle.

Operation. A right frontal craniotomy disclosed a gray-colored solid tumor partly investing the right optic nerve and internal carotid artery. Only 1 cm of the dome of the tumor was removed before rock-hard tissue was reached. The tumor, a mixture of calcified nodules and firm gelatinous material, was histologically characteristic of a chordoma. Subsequently, the patient was given 5000 r of supervoltage radiation therapy in 42 days, but no improvement occurred, and the hypopituitary features increased.

The patient survived for 5 years and died following a head injury.

Postmortem examination. A 4 × 5 cm tumor mass arising from the base of the skull completely obscured the pituitary fossa (Fig. 2). The tumor was bossellated, hard, and glistening white. The third cranial nerves were elongated and stretched over the tumor, and there was a deep indentation in the brain overlying the tumor mass. The mammillary bodies were flattened, and the region of the tuber cinereum and the pituitary stalk could not be identified. Microscopic examination of this tumor showed chondrocytes embedded in a cartilaginous matrix with areas of calcification and ossification. The appearance was that of a benign chordoma.

Comment. The correct diagnosis of either chordoma or chondroma was possible on radiological grounds.

Case 3. In 1958, when 51-years old, this man had been seen elsewhere for left frontal pain and a left ptosis of 6 days’ duration.
His early history, resulting in removal of a chordoma, has already been reported.\textsuperscript{2,3} By 1960, x-rays had shown erosion of the anterior wall, floor, and dorsum of the sella turcica with destruction of the sphenoidal and adjacent left ethmoidal air sinuses. A transorbital ethmoidectomy opening into the sphenoidal air sinus revealed a firm blue cystic swelling with mucoid contents. The cyst was opened, its mucoid contents aspirated, and part of its wall removed.\textsuperscript{3} The histological report was of a chordoma, and within a few days the headache and oculomotor signs had disappeared. Following operation, however, the patient developed well-marked signs of hypopituitarism for which he was given cortisone and later other hormonal therapy. A course of Cobalt\textsuperscript{60} irradiation was given. Shortly afterwards intermittent cerebrospinal rhinorrhea appeared.

\textbf{Examination.} Early in 1963 a bitemporal hemianopia developed associated with bilateral primary optic atrophy, and the patient, now 56 years old, was admitted to the Neurosurgical Unit. He was almost blind. X-rays showed increased bony destruction in the same sites as before. The dorsum sellae was greatly thinned; there was linear calcification adjacent to it. Air encephalography outlined a suprasellar extension of the tumor, indenting the anterior end of the third ventricle. The aqueduct and fourth ventricle were in their normal situations.

\textbf{Operation.} On October 4 and December 13, frontal explorations were performed from the right and left sides respectively. There was a gelatinous tumor beneath the chiasm and the left optic nerve. During the first exploration, sufficient tumor was removed intracapsularly to relieve pressure on the right optic nerve. In the second, a more radical removal leaving a cavity 2 1/2 to 3 cm in diameter was made. The convalescence was protracted by postural hypotension, diabetes insipidus, and a bout of meningitis. In due course these were partially controlled by endocrine therapy and antibiotics, and there was some improvement in the vision of the right eye. Four months later, however, the patient died after a short period of mental confusion.

\textbf{Postmortem examination.} Extensive de-
struction of the base of the skull was seen in
the region of a greatly expanded pituitary
fossa. A mass of purplish tumor lay beneath
the optic chiasm and extended down into the
region of the sphenoidal sinuses and into the
left nasal cavity. The carotid arteries were
embedded in tumor tissue, and the mass also
extended upward into the third ventricle al-
most as high as the massa intermedia. The
tuber cinereum, mammillary bodies, and pi-
tuitary gland could not be identified. Micro-
scopically, the tumor consisted of cells of
greatly varying size, with distension of the
cytoplasm by vacuoles. Some tumor cells
were multinucleated or contained gigantic bi-
zarre masses of nuclear material. No mitotic
figures were present, and there was a moder-
ate amount of extracellular mucinous mate-
rial; the appearance was that of chordoma.

Comment. The course of this patient’s ill-
ness from first to last was nearly 6 years,
and in retrospect the diagnosis was apparent
radiologically by the second year.

Group 2: Parasellar Chordomas
and Chondromas

Case 4. A 47-year-old man was referred to
us in 1958 with a 4-month history of pain
and slight proptosis (2 mm) of the left eye-
ball and diplopia with slight ptosis indicat-
ing a left third nerve paresis.

First examination. X-rays of the skull
showed that the left side of the sella turcica
was deepened, and the left anterior and
posterior clinoid processes were eroded (Fig.
3). The visual fields were normal. Air en-
cephalography and left carotid arteriography
suggested a suprasellar as well as a left lat-
eral extension of a pituitary tumor, which
 provisionally was regarded as an “invasive”
chromophobe adenoma. A course of conven-
tional radiation therapy (3500 r) was fol-
lowed by temporary improvement of the dip-
lopia. The pain had disappeared prior to this
treatment, but absent libido and other fea-
tures of hypopituitarism supervened.

Second examination. Within 2 years a
progressive left third nerve palsy appeared
associated with an incongruous right homon-
ymous hemianopia. Repeat air encephalo-
graphy indicated that the suprasellar and left
lateral extensions had become larger, al-
though no air entered the ventricles.

Operation. A left frontal craniotomy re-
vealed a large tumor displacing the left optic
erve upward and inward, and also bulging
laterally from the region of the cavernous
sinus into the left temporal lobe. A partial
intracapsular removal of this tumor was per-
formed; the tumor tissue was of tough con-
sistency.

Immediately postoperatively the patient
was drowsy, with a right hemiparesis. Three
days later, after a left carotid arteriogram
had shown increased elevation of the left
middle cerebral artery and also further dis-

Fig. 3. Case 4. Lateral and Towne’s view of the skull showing deepening of the sella turcica and erosion
of the anterior and posterior clinoid processes on the left side.
placement of the anterior cerebral artery to the right, the craniotomy was reopened. Instead of the anticipated clot, the temporal pole was found swollen. Excision of 4 cm of its anterior portion disclosed a 5-cm tumor projecting into the mesial side of the temporal lobe from the cavernous sinus. Its contents were again tough. The microscopic appearance of this tumor was characteristic of a chordoma. There were cells of varying sizes containing cytoplasmic vacuoles, with some of the larger cells markedly distended and having a "signet-ring" appearance. Much extracellular mucinous material was present, as were areas of necrosis and old organized hemorrhage. No mitotic figures were seen.

Following reoperation the patient remained comatose for 2 months before making slow improvement. He was able to walk but had left third nerve palsy, right hemiparesis, marked dysphasia, and homonymous hemianopia. He died 6 months after operation.

Postmortem examination. A tumor was found replacing the pituitary gland and adjacent base of the skull.

Comment. The radiological appearance was considered to be that of a chromophobe adenoma projecting laterally into the temporal lobe. Initially there had been no endocrine symptoms, but these appeared after the radiation therapy. Even at the time of operation the diagnosis of "invasive pituitary adenoma" was still being entertained, and later when the pathological report became available the patient's general state appeared too parlous to warrant further intervention. Even in retrospect it would have been difficult to have anticipated the correct diagnosis. However, it was known before operation that the tumor was primarily parasellar. Therefore, instead of a subfrontal approach with its limited success, a combined subfrontal and subtemporal approach might have given better exposure of the tumor with chances of better palliation.

Case 5. A 23-year-old man had a 1-year history of headache and double vision. For 4 months he had noticed numbness in the upper two-thirds of the right side of the face, as well as vertiginous sensations on quick movements of the head.

Examination. There was partial paresis of the right third, fifth, and sixth cranial nerves. The visual fields were normal. X-rays showed a mass about 5 cm in diameter containing patchy calcification and situated just above the base of the skull in the region of the foramen lacerum. There was erosion of the right side of the sella and the apex of the petrous bone that extended laterally to the right foramen ovale. The superior orbital fissure was enlarged and the walls of the right optic canal decalcified. A soft-tissue mass projected into the right sphenoidal air sinus.

Right carotid arteriography showed forward and inward displacement of the right internal carotid artery both in its course through the base of the skull and within the cavernous sinus; there was marked elevation of origin of the middle cerebral artery. Air encephalography was not performed.

Operation. Right lateral craniotomy with elevation of the temporal lobe disclosed an extradural mass occupying the general region of the cavernous sinus. The dura over it was incised anteroposteriorly, and yellow gelatinous tissue interspersed with semiporous calcified material was scraped out from within the capsule, leaving a cavity about 4 cm in diameter. We then could see the internal carotid artery in the cavity, with an attenuated nerve (probably the third) lateral to it. The tumor consisted of small irregularly shaped cells lying in lacunae in a cartilaginous matrix. The cells and nuclei were fairly uniform in size and shape, and no mitotic activity was present. In some areas there was well-marked granular calcification and ossification. The appearance was that of a benign chondroma.

Postoperatively the cranial nerve paresis persisted but the headaches ceased. The patient soon resumed his old employment and, after 15 months, married. He stated that his marital functions were normal. When he was last seen 5 years after operation, there was no evidence of further deterioration. Skull x-rays showed some increase in the density of calcification, but the size of the tumor was unchanged.

Comment. This parasellar tumor eroded the base of the skull. The radiological evidence in this case enabled us to make the correct diagnosis of either a chordoma or chondroma, and the degree of operative pal-
liation achieved without radiation therapy was marked.

Case 6. A 45-year-old woman had developed diplopia 9 years before being referred to us, which had been treated by advance-
ment-recession operations of the right eyeball muscles. This had relieved the diplopia for 5 years, when a further operation on the eyeball muscles was without benefit. The signs by then included a right third nerve pa-
resis. A year later, aching pain developed in the right eyeball, with bouts of blurred vi-
sion. Two years later, shortly before her ad-
mission, the patient developed recurrent at-
tacks of tingling in the right side of the face, and vertiginous sensations on sudden move-
ment of her head.

Examination. The patient was a healthy
looking woman with mild papilledema, no-
mal visual fields, but slight reduction of vi-
sual acuity in the right eye (6/9, J.4 cor-
rected). The right eyeball was slightly prop-
tosed. There were partial right 3rd and 6th nerve pareses with hyperalgesia over the right trigeminal distribution. The right Ba-
binski test was positive. X-ray of the skull
showed thinning and decalcification of the skull base in the region of the foramen lac-
erum. The bony changes involved the right anterior clinoid process, the apex of the pe-
trous bone, and the floor of the right optic
channel. There was no intracranial calcifica-
tion. Arteriography showed changes similar
to those in Case 5, with the addition that the artery in the region of the tumor was attenu-
ated. During air encephalography, no air
centered the ventricles, and the pontine and the interpeduncular cisterns only filled to the
left of the midline. The lumbar cerebrospinal
fluid contained one cell and 40 mg% of pro-
tein.

Operation. Right lateral craniotomy re-
vailed findings similar to those in Case 5. The temporal pole was amputated to facili-
tate exposure. An intracapular removal ap-
proximately 4 cm in diameter was per-
formed. Bleeding from the interior of the
tumor cavity was difficult to control. The mi-
croscopic appearance of the tumor was char-
acteristic of a chondroma, with small tumor
cells of fairly uniform shape and size lying in lacunae in a uniform cartilaginous matrix. A
little granular calcification was present, but
no ossification.

Postoperatively, supervoltage radiation
therapy (4500 r) was given over the course of
a month. The visual acuity in the right eye returned to normal, the papilledema dis-
appeared, but the occulomotor nerve paresis persisted. The trigeminal impairment also disappeared. Six months after operation, the patient had two grand mal epileptic attacks. She resumed her housework, but had post-
arative depressive episodes. She appeared
to have a functional intermittent weakness of
the right limbs; there was no alteration in
the tendon jerks. The plantar responses were
flexor. The patient is still alive 5 years after
operation. She has normal vision and is on
tranquilizer and anticonvulsant drugs; there
is no x-ray evidence of enlargement of the
tumor.

Comment. It is difficult to decide how
much the radiation therapy contributed to
the patient’s improvement. Certainly the
major part of her improvement resulted from
operation. It is interesting that no calcifica-
tion was present in the tumor, and the cor-
rect diagnosis was not possible from the skull x-rays. However, the angiographic and
air studies did indicate the presence of a par-
asagittal tumor which, as it was not calcified,
could not be distinguished from an epider-
moid, a parasellar meningioma, or a pitui-
tary tumor.

Case 7. A 30-year-old man had com-
plained for 1 year of increasing inability to
see objects on the right and more recently of
blurred vision in the left eye. There had
been no headaches. The patient had been
impatient for 7 years.

Examination. The patient had an incon-
gruous right homonymous hemianopia with
a central scotoma in the left eye, and mild
hypopituitarism. Visual acuity was 6/6, J.1,
on the right and reduced to finger counting
on the left. X-rays showed erosion of the cli-
vus with a partially calcified mass posterior
to it (Fig. 4 left). The erosion had spread
into the left parasellar region, involving the
left petrous apex, the left clinoid processes,
the left superior orbital fissure, and the floor
of the left optic canal. Tomograms showed
that the dorsum sellae was thinned but in its
correct position (Fig. 4 right). The left ca-
rotid arteriogram showed forward and up-
ward displacement of the intracranial carotid
artery and its bifurcation. The basilar artery as shown by vertebral angiography was displaced backward behind the partially calcified mass. The lumbar encephalogram showed marked backward displacement of the fourth ventricle and aqueduct with elevation of the floor of the third ventricle and obliteration of its anterior end; the left temporal horn was displaced backward and upward. The pontine cistern was compressed.

Operation. A left lateral craniotomy with elevation of the temporal lobe revealed that the tentorium was bulging upward. It was incised anteroposteriorly, thus opening into a yellow, gelatinous extradural tumor about 3.5 cm in diameter. It lay in front of the brain stem and in the left parasellar region. The interior of the tumor was scooped out together with some small sheets of cortical bone from the posterior part. Anteriorly the cavity was formed by bleeding cancellous bone and posteriorly by dura. Hemostasis was secured with oxycel. Microscopic examination of the tumor showed a characteristic chordoma. Cells varied greatly in size, the largest being those grossly distended by cytoplasmic vacuoles and having a 'signet ring' appearance (Fig. 5 left). No mitotic figures were found. There was much extracellular mucin; granular calcification and ossification were also present in some areas.

The patient made a good recovery. He was left with a minimal third nerve paresis, but his vision showed some improvement, and he noticed that he could now appreciate colors with the right eye. The right upper quadrant of the visual field had returned. X-rays 1 year later showed some regeneration of the clinoid processes and of the clivus. He returned to work as a storeman.

Second examination. Two years later the patient was readmitted to the Neurosurgical Unit because of deterioration of vision in his good, right eye for 1 month. Visual acuity was 6/9, J.1, with difficulty on the right and reduced to finger counting on the left. The right temporal field was now gone. X-rays of the skull showed further calcification above the sella turcica and clivus, while left carotid arteriography and air encephalography confirmed the presence of a large left parasellar mass.

Second operation. The left lateral craniotomy was reopened, the temporal lobe elevated, and the tumor thus exposed. The interior of the tumor was again curetted; this provoked bleeding which was controlled with difficulty. The histological appearances of the tumor had not changed. The patient died 2 days later.

Postmortem examination. A blood clot 5 \( \times 3 \times 3 \) cm was found posterior to the dorsum sellae indenting the hypothalamic region. Clot was also found in the basal cisterns and within the ventricular system. There was a small pituitary gland within the sella turcica.

Comment. The radiological evidence indicates that, although this tumor may have arisen primarily from the clivus in the posterior fossa, it had extended forward to involve structures in the left parasellar region. The symptoms were connected principally with this parasellar extension. Thus, the mild hypopituitary features were probably related to compression of the hypothalamus. The ra-
diological findings were diagnostic, and enabled a planned operation to be performed. This benefitted him, and so radiation therapy was not given.

**Group 3: Clival Chordomas and Chondromas**

*Case 8.* A 51-year-old man had developed double vision 3 years before being seen by us and, 6 months before, “turns” suggestive of transient basilar artery ischemia. Some of these “turns” were occasioned by quick movements of the head, others appeared spontaneous; in them he exhibited bilateral oculomotor pareses of a bizarre type, nystagmus, ataxia of all limbs, dysarthria, and bilateral extensor plantar responses, associated occasionally but not always with clouding of consciousness. Such attacks would last several minutes and recur with increasing frequency up to several times a week. More recently his right face had become numb.

**Examination.** The patient was a well-built man who had bilateral low-grade papilledema. The visual fields were normal. There was a complete right sixth nerve palsy and a partial left sixth nerve paresis. There was diminished sensibility in the right trigeminal area with weakness of the right masticatory muscles. Between his “turns” there were no abnormal signs in the limbs. X-rays of the skull showed gross destruction of the dorsum sellae and clivus with a few flakes of calcification situated above and behind the sella. Carotid and vertebral arteriography as well as air encephalography outlined a mass projecting backwards from the region of the destroyed clivus slightly more to the right than to the left of the midline (Figs. 6 and 7).

**Operation.** A right lateral craniotomy was performed, with findings and procedure similar to those in Case 7, and the tumor was a typical chordoma.

At 6 hours after operation, the patient developed a clot in the anterior part of the right temporal lobe, necessitating reexploration and amputation of the anterior 5 cm of

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**Fig. 5. Left:** Section of the chordoma from Case 7. The tumor cells vary considerably in size. Some are small with scanty cytoplasm while others are greatly distended by large cytoplasmic vacuoles. There is much extracellular mucinous material. **Right:** Section of the chondroma from Case 9, showing the tumor cells lying in a hyaline cartilaginous matrix. Granular calcification and early ossification are present in the lower part. H. & E., ×400.
the right temporal lobe. Thereafter he made a good recovery, but with a persistent right sixth nerve weakness and analgesia of the third division of the trigeminal nerve which had been cut when the dura over the tumor was incised. Vision declined in the right eye to 6/18 for no obvious reason. Three months later he returned to work as a coal miner.

The patient’s vision continued to deteriorate, however, and 1 year later he complained of hoarseness and dysphagia. After another 6 months, visual acuity was found to be reduced to finger counting in the right eye and 6/60, J.20, in the left. He had a left homonomous hemianopia with a right ophthalmoplegia. Trigeminal analgesia became complete in the first and third divisions, but only partial in the second division. Palatal weakness developed on the right side, as well as weakness of the right trapezius and sternomastoid muscles, and hemiparesis and hemianesthesia in the left limbs.

It was now 2 years after the first operation and x-rays of the skull showed that the tumor had increased in size.

Second operation. The craniotomy was reopened. A hemorrhagic tumor was found lying in front of the brain stem and beneath

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**Fig. 6.** Case 8. Right carotid arteriogram with cross compression showing unfolding of both carotid syphons and elevation of each carotid bifurcation.

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**Fig. 7.** Case 8. Left: Lumbar air encephalogram showing air in the third ventricle, aqueduct, and fourth ventricle. The posterior end of the third ventricle and the aqueduct are stretched upward and posteriorly by the underlying tumor. Air in the pontine cistern helps to visualize the extent of the tumor. Right: The vertebral arteriogram confirms the presence of a mass arising from the clivus.
the optic chiasm, which was prefixed. An attempt to remove more tumor substance and decompress the left optic nerve was not successful owing to the excessive vascularity of the tumor. Postoperatively, the patient was found to be blind in the left eye. His general condition continued to deteriorate and he died 2 weeks after operation.

Postmortem examination. A tumor 7×4 cm was found extending from the posterior part of the anterior cranial fossa almost to the foramen magnum. The bone in the mid-line of the skull around and including the pituitary fossa had been distorted and destroyed by tumor which was confirmed as a chordoma.

Comment. The signs of chordoma in this case were those of compression of the fifth and sixth cranial nerves associated with "turns" deemed to be due to basilar artery ischemia from compression of the brain stem. There were no obvious endocrine features, but the radiological appearance was characteristic of a chordoma. The patient benefited from the operation for only 12 months.

Case 9. A 54-year-old woman, 6 years before referral, had developed a left sixth nerve palsy. X-rays at that time had shown a calcified mass immediately behind the clivus with destruction of the dorsum sellae. The patient had been referred from one hospital to another with various diagnoses ranging from craniopharyngioma to chordoma, but she was deemed inoperable. Some 18 months before her admission, her gait had become unsteady, and shortly afterwards she developed numbness of the right side of her face, as well as bilateral sixth nerve palsies. Then, 6 months before admission, she was given a course of Cobalt 60 therapy (4600 r) which temporarily epilated her scalp. There was no improvement. Subsequently her menses ceased, and she attempted suicide; this led to her admission to the Maudsley Hospital under psychiatric care.

Examination. The patient appeared to be a very depressed woman with dysarthria. The scalp was still epilated. The visual fields, optic discs, and visual acuity were normal. There were bilateral sixth nerve palsies and slight bilateral nerve deafness. Sensation was lost in the right trigeminal area with weakness of the right masticatory muscles. The tongue protruded slightly to the left. All four limbs were ataxic, and the gait was extremely unsteady. Both plantar responses were extensor. X-rays of the skull showed a partially calcified mass, arising from the clivus and extending into the right side of the pituitary fossa (Fig. 8 left). The basisphenoid and dorsum sellae were eroded. The right carotid arteriogram was relatively normal, but the right vertebral arteriogram showed that the basilar artery was displaced backward about 2 cm and also curved markedly around the left side of the tumor (Fig. 9). Air failed to enter the ventricular system during lumbar encephalography. The lumbar fluid had 80 mg% of protein.

Operation. A right lateral craniotomy was performed. Amputation of the anterior 4 cm of the temporal lobe revealed a large tumor situated extradurally in the region of the clivus and the right parasellar region. Its interior was scooped out, as in the preceding cases, the removal being subtotal. Microscopic examination showed a cellular chordoma with marked variation in size of cells and nuclei. No mitotic figures were seen. In some regions the matrix was partially calcified and a little ossification was present (Fig. 5 right).

The patient's convalescence was protracted for several months. She was in coma for the first 3 days. A right-sided tarsorrhaphy was performed for a corneal ulcer. A year later she was able to look after her 13-year old son. Movements of the left eye had improved, and x-rays of the skull showed that the residual tumor mass was smaller (Fig. 8 right). When last seen nearly 4 years after operation, she had no complaint of headache. She has become a little more unsteady on her feet and has developed a right facial palsy and a right palatal weakness. The right eye was adducted 30°, but otherwise vertical and medial movements are full. There was still paralysis of the right trigeminal nerve and slight bilateral deafness. Apart from ataxia and bilateral extensor plantar responses there were no abnormal neurological signs in the limbs, although skull x-rays showed some increase in the size of the calcified mass.

Comment. The radiological appearances in this case were characteristic of a "chor-
doma,” although histologically the lesion was classified as a chondroma. The tumor did not respond to radiation, but the patient was greatly improved by operation.

**Discussion**

In this series of nine cases, five showed a marked initial improvement following an intracapsular removal of the tumor, which has been maintained 4 to 5 years later in three instances. Two of these patients (Cases 5 and 6) lead very active lives. Operation, even though palliative, seems to have been worthwhile in this group. Of the remaining four cases, one survived for 5 years in an unimproved state after the removal of the “dome” of the tumor (Case 2), one tumor was deemed irremovable (Case 1), a third was merely biopsied (Case 4), while the patient who had already been operated upon through the nose, developed cerebrospinal rhinorrhea with meningitis and a prolonged

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**Fig. 8. Case 9.** *Left:* Lateral x-ray showing calcification in a chondroma arising from the clivus. *Right:* A comparable film, 1 year later, showing a reduction in the size of the tumor bulk.

**Fig. 9. Case 9.** Right vertebral arteriogram showing backward displacement of the basilar artery which is curving around the left side of the tumor.
convalescence following the intracranial procedure. The over-all picture of surgery in these cases is, therefore, somber, but not hopeless. However, the precise nature of the lesion gives an answer to the different outcomes, for all four cases with long survival had "chondromas" and not true "chordomas." It is generally not possible to distinguish these two types preoperatively.

All tumors in this small series when first seen were reported as chordomas, but a review has shown that four were chordomas. Indeed, many of the cases illustrated in the literature as chordomas are in fact chordomas, and it seems clear that these two sets of lesions have been confused in the past (see Cases 2 and 5, Poppen and King). In reviewing Olivecrona's cases, Leitholf tried to distinguish the two lesions, but he described them in the same situations as having similar symptoms and x-ray appearances. It may well be, therefore, that these two types of lesions are in reality both derived from notochordal remnants.

Although differing in their microscopic characteristics, the two kinds of tumor have many points in common. They occur in the same sites, produce the same syndromes, and usually have similar radiological appearances. At operation they look alike. Both are resistant to radiation and will respond, we feel, to the same palliative surgical approach. For these reasons we have grouped them together, especially as the chordoma is an unusual type, and very distinct from the more common osteochondroma seen in the paranasal sinuses. However, although some chordomas are poorly calcified, the calcification increases with years without the tumor necessarily enlarging. When densely calcified, as in Fig. 1, the radiographic appearances are pathognomonic.

Granted, therefore, that these two tumors are in reality variants of one neoplastic process, our subdivision of them according to their site of origin in the base of the skull does help explain the variation in the clinical syndromes, not only in our own case material, but in that in the literature. Briefly, the sellar group is characterized by signs of hypothalamic dysfunction and of chiasmal compression with bitemporal hemianopia. The parasellar group may involve, unilaterally, the third, fourth, and sixth cranial nerves, producing various degrees of squint, ptosis, and pupillary abnormality, and the optic tract where the tumor produces an incongruous homonymous hemianopia, pressure on the Gasserian ganglion or its peripheral branches resulting in numbness of the face, and encroachment of the pituitary gland or hypothalamus leading to hypopituitarism. The clival group is characterized by signs of brain stem compression producing ataxia, signs of pyramidal dysfunction, and involvement of various nerves in the posterior fossa such as the sixth and eighth cranial nerves. A fairly characteristic feature appears to be bilateral involvement of the sixth cranial nerves. Thus, some reported cases resembled cerebellopontine angle tumors.

Although subdivision of cranial "chordomas" into these three groups helps to explain the variations in their neurological presentation, it is arbitrary and has many limitations. A tumor, in its slow relentless growth, arises in one site and slowly spreads into another with alteration of the clinical picture. In our case material, two of the nine patients had definite signs of raised intracranial pressure (Cases 6 and 8). One (Case 3) presented initial evidence of sphenoidal sinus and nasopharyngeal tumor, and was so reported. However, even at that stage the skull x-rays showed destruction of the floor, walls, and dorsum of the sella turcica. Similar destruction has been present in other cases reported with nasopharyngeal extensions.

Often the preoperative diagnosis of these cases may be difficult. This was especially so in our first case where the plain x-rays showed a normal sella turcica, and the air encephalogram only a suprasellar tumor compressing the chiasm and causing bitemporal hemianopia. Generally, however, there is considerable destruction of bone around the sella turcica, the suprasellar region, or the clivus, but the differential diagnosis from other lesions in these regions such as invasive pituitary adenomas, craniopharyngiomas, and epidermoids may be difficult. The diagnosis becomes easier if, in addition to erosion of the bone in these regions, there is an irregularly calcified mass behind the clivus, particularly if it appears to have displaced backward some flakes of cortical bone from the clivus. However, our small series shows that it is impossible on this basis
to distinguish between classical "chordomas" and "chondromas." Thus, of the five classical "chordomas," two showed no calcification (Cases 1 and 4), while one "chondroma" (Case 6) was similarly without calcification.

The radiological appearances were characteristic of either chordoma or chordoma in six cases (Cases 2, 3, 5, 7, 8 and 9). The differentiation from other calcified tumors in this neighborhood, notably the craniopharyngiomas, offered no difficulties because of the associated bony erosion. The correct diagnosis, however, was not possible in the remaining three cases. In Case 1 the plain x-rays were normal, and arteriography showed a small suprasellar mass indistinguishable from a suprasellar meningioma. In Case 4 differentiation from an "invasive" pituitary adenoma was not possible. In Case 6 the radiological studies revealed a parasellar mass without calcification and with some local skull erosion, an appearance which we have also seen in a paraspinal epidermoid. Thus, our findings agree with those of others\textsuperscript{15} that a correct preoperative diagnosis is not always possible. They also show that it is not possible to distinguish chordomas from chondromas.

The radiological studies, however, did always indicate the site and approximate size of the intracranial lesion. In the case of the parasellar and clival lesions, the combination of carotid arteriography and of vertebral arteriography was particularly useful, for the basilar artery was characteristically bowed backwards by the tumour.

Although radiotherapy has been advocated for these tumors\textsuperscript{7,14} the degree of benefit reported has not been impressive. It was given in 5 of our 9 cases without any really worthwhile improvement. Zoltán and Fényes\textsuperscript{13} reported a single case in which seeds of radioactive yttrium were implanted at operation with some apparent benefit, but the follow-up was only 9 months.

Our small series of cases suggests that some useful palliation is often possible after appropriate neurosurgical intervention. In cases where the clinical features and radiological findings indicate that the tumor is in the sella, a subfrontal approach as for a pituitary tumor will probably suffice. In those cases where the tumor is in a parasellar or clival portion, however, the lateral or temporal approach is indicated. The essentials of this approach are that the bone flap should be taken down to the floor of the middle cranial fossa so that the anterior part of the temporal lobe can be elevated, revealing the tumor covered over by its dural investment. This investment, which usually includes the anterior part of the tentorium, is then incised in an anteroposterior direction to permit an intracapsular removal of the tumor. Amputation of the anterior part of the temporal lobe facilitates exposure and minimizes risk of a hematoma developing within the retracted temporal lobe. We have had no experience of transcervical transclival craniotomy, an approach pioneered by Stevenson and his colleagues at San Francisco.\textsuperscript{11} We imagine, however, that this approach would not be adequate for the complete removal of the tumors that we encountered.

**Summary**

Nine cases of chordoma or chondroma arising from the skull base have been described and subdivided into three groups:

1) Sellar, characterized by optic chiasmal compression and signs of hypopituitarism

2) Parasellar, characterized by ocular motor nerve pareses, optic tract compression, and hypopituitarism

3) Clival, characterized by bilateral sixth nerve paresis and brain stem compression without hypopituitarism or chiasmal compression.

The two types of tumor are often indistinguishable preoperatively, except when a chordoma is densely calcified and then presents a characteristic x-ray appearance. Some chordomas, however, are poorly calcified. Histologically, the two types of tumor have often been confused in the literature, and probably both arise from notochordal elements.

The chordomas have the better prognosis. Our experience indicates that useful palliation can sometimes be accomplished by intracapsular removal of the tumor.

**Acknowledgments**

We wish to thank our various colleagues
Chordoma and Chondroma of Skull Base

who have entrusted us with their patients and helped us with this paper. Mr. P. H. Schurr, F.R.C.S., operated on Cases 2 and 6, while Dr. R. D. Hoare undertook the neuroradiological studies.

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