STEREOTACTIC DIAGNOSIS AND RADIOACTIVE TREATMENT IN A CASE OF SPHENO-OCCIPITAL CHORDOMA

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ALTHOUGH in 1958, at the time we diagnosed our case, chordomas had been known for 100 years, they are still looked upon, especially those of the spheno-occipital region, as one of the most hopeless problems in neurosurgical diagnosis, therapy and prognosis. There seems to be general agreement that the diagnosis of cranial chordoma is extremely difficult, its complete surgical removal is impossible, and the prognosis is hopeless, though not everyone shares the extreme view put forward by Kotzareff,36 who said that it is impossible to diagnosis chordoma in a living patient, or that of Stanton53 that “practically all spheno-occipital cases are inoperable.” Even Congdon,12 who described the follow-up of 25 years in a case of nasopharyngeal chordoma operated on several times, concluded that: “it is doubtful if clival and cervical chordomas which involve the central nervous system can be cured by present methods.” In view of these facts the stereotactic surgical method employed in our case seems to be the method of choice in regard to both diagnosis and therapy, although the prognostic aspects of the problem are still far from being solved.

Historical Note. Virchow60 (1857) is often credited with the discovery of chordoma, although it is likely that he saw and described this jelly-like outgrowth of the cranial base only after Luschka39 (1856) had done so. At any rate, Virchow60 certainly misinterpreted the structure of such tumors, believing them to be cartilaginous in origin: this explains the term suggested by him (echondrosis physaliforma) to denote such neoplasms. In 1858 Müller42 recognized that these growths are of notochordal origin, but this correct view failed to gain widespread acceptance for a long time. Such outgrowths in the base of the skull had been seen only at autopsies.

The first case of expansive chordoma was described by Klebs34 (1864), in which symptoms of increased intracranial pressure were noted. Ribbert47,48 (1894, 1895) recognized these outgrowths as neoplasms, and suggested their correct name, i.e. chordoma (designated previously by Müller). He even produced experimental lesions in the nucleus pulposus, which is of notochordal origin.

The first case of malignant cerebral chordoma associated with neurological symptoms was described by Grah124 in 1903. In America the first case of chordoma was published by Jelliffe and Larkin33 in 1912, and the first case of
spheno-occipital chordoma was reported by Daland in 1919; the first case to be recorded in the British Isles was that of Burrow and Stewart in 1923.

**Occurrence.** Chordoma is a rare kind of tumor. It occurs at sites along the notochord during ontogenetic development: from the nasopharynx, through the spheno-occipital synchondrosis, down to the sacrococcyx. It is remarkable that 84 per cent of all chordomas occur at the cranial and sacrococcygeal ends of the notochord.58

**Absolute Frequency.** The number of reported cases of chordoma has, of course, been increasing, obviously because of improvement in diagnosis. This is clear from the data of Utne and Pugh,58 shown in Table 1.

**Relative Frequency.** In Ribbert’s47 autopsy material ecchondrosis spheno-

<table>
<thead>
<tr>
<th>Author</th>
<th>Year</th>
<th>Cranial Chordomas</th>
<th>Total Cases</th>
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<tbody>
<tr>
<td>Coenen11</td>
<td>1925</td>
<td>21</td>
<td>47</td>
</tr>
<tr>
<td>Mabrey49</td>
<td>1935</td>
<td>49</td>
<td>150</td>
</tr>
<tr>
<td>Faust et al.20</td>
<td>1944</td>
<td>96</td>
<td>252</td>
</tr>
<tr>
<td>Tonelli26</td>
<td>1948</td>
<td>118</td>
<td>317</td>
</tr>
<tr>
<td>Utne and Pugh58</td>
<td>1955</td>
<td>197</td>
<td>505</td>
</tr>
</tbody>
</table>

occipitalis occurred in 2 per cent, and in Stewart and Burrow’s55 material in 1.5 per cent of the cases. Harvey and Dawson27 found among 16,000 tumors of all regions of the body only 14 definite and 5 doubtful chordomas. Petit-Dutaillis et al.44 encountered in over 3,000 cases of cerebral tumors only 4 histologically verified chordomas (1.33 per million). In the material of Tönns55 there were 0.2 per cent of chordomas among 6,000 cerebral tumors.

**Age.** Chordoma may occur at any age. Hennig31 described a sacrococcygeal chordoma in a fetus aged 7 months. Rubaschow49 excised a tumor from the upper jaw of an infant 24 hours old, which proved histologically to be a chordoma. The oldest patient with a spheno-occipital chordoma was aged 82 years. The greatest incidence of sphenoid chordomas occurs in the third and fourth decades, the average age at onset being 40 years.

**Size.** The largest chordoma known filled the entire middle fossa of the skull,28 and the smallest was almond-size and caused symptoms of the cerebellopontile angle.6

**Duration.** The longest duration of symptoms was in the case of nasopharyngeal chordoma reported by Congdon12 with duration of 25 years, and in that of Bailey and Bagdasar7 (Case 1), whose patient had complained of headaches for 18 years. The shortest duration was in Stanton’s33 case, in which symptoms of increased intracranial pressure were of 4 months’ duration and the postoperative survival was 2 months (a total of 6 months), and the case of Uhr and Churg,57 in which death followed approximately 6 months after onset of symptoms.

The average duration of life of patients who had involvement of cranial