

CHORDOMA

A Clinicopathologic and Prognostic Study of a Swedish National Series

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Chordoma is a rare tumor with slow growth developing from remnants of the notochord and thus appears in close relation to the axial skeleton. A Swedish national series collected over a period of 13 years, comprising 51 patients with a follow-up time of 8–20 years, was studied. Histologically all tumors had a typical appearance except two, which had the characteristics of “chondroid chordoma”. Fifty-seven per cent of the tumors were located in the sacrococcygeal region, 27 per cent in the spheno-occipital region and 16 per cent in the vertebrae. The peak age incidence was in the 6th and 7th decades. Male: female ratio was 1:1. The main symptoms were pain and neurologic disturbances. Skeletal destruction was noted radiographically in most instances. Intratumoral calcifications were rarely seen. The treatment was surgery, radiotherapy or a combination of these. There was only one long-term survivor without evidence of disease, a patient operated on for cervical chordoma 14 years earlier. Six other patients lived 8–18 years after diagnosis with chordoma. All other patients were dead; 39 died of chordoma, five of unrelated causes. Distant metastases were noted in 29 per cent. Chordoma constituted 17.5 per cent of all primary malignant bone tumors of the axial skeleton. The yearly incidence was 0.51 per million inhabitants in Sweden.

Key words: bone tumors; chordoma; histology, prognosis, treatment

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Chordoma is a rare tumor appearing in close relation to the axial skeleton and commonly believed to arise from notochordal remnants. Extensive reviews and studies of chordoma have been published since 1894 when Ribbert gave the tumor its name; since then at least 700 cases of chordoma have been reported (although it is obvious that many reports and reviews deal with the same chordomas). The tumor is most often located in the sacrococcygeal area (approximately 50 per cent), somewhat less commonly in the spheno-occipital area (approximately 35 per cent) and more rarely in the vertebrae (approximately 15 per cent) (Mabrey 1935, Littman 1953, Dahlin 1970). Men appear to be more exposed than women (ratio 2:1) and the most afflicted decades are the third and fourth for

spheno-occipital chordomas and the fifth and sixth for sacrococcygeal chordomas. The clinical course is generally prolonged with a strong tendency for recurrences. Metastases are reported to appear in from less than 10 per cent to 60 per cent of the patients (Wood & Himadi 1950, Dahlin & McCarthy 1952, Higinbotham et al. 1967, Wang & James 1968, Paavolainen & Teppo 1976). The treatment mostly advocated in the literature is surgical ablation (e.g. Littman 1953). However, the frequently large size of the sacrococcygeal tumors and the inaccessible location of the vertebral and spheno-occipital ones make surgery difficult. Thus, follow-up studies have revealed a very high recurrence rate and an almost invariably fatal outcome, also in cases without metastasis. However, in the case of sac-

rococcygeal tumors, radical surgery has recently been shown to be much more possible than was formerly thought (Stener & Gunterberg 1978). Radiotherapy in high doses seems to restrain the tumor but is not to be relied upon for cure (e.g. Wood & Himadi 1950, Dahlin & McCarthy 1952, Higinbotham et al. 1967, Pearlman & Friedman 1970, Weldinger 1975).

The purpose of this paper is to give a correlated clinicopathologic review of all chordomas diagnosed in Sweden during a 13-year period (1958–1970), with a follow-up period of 8–20 years; to calculate the incidence of chordoma in Sweden; and to describe the natural history of chordoma as well as the results of traditional treatment. Altogether, 51 previously unreported cases are presented.

MATERIAL AND METHODS

All cases of chordoma reported to the Swedish Cancer Registry during the period 1958–1970 were studied. (All malignant tumors diagnosed in Sweden have been reported to the Registry since the year 1958. A double reporting system is used involving clinicians, pathologists and cytologists throughout the country).

For the years 1958 through 1970 a total of 979 primary malignant bone tumors were reported, of which 290 were confined to the axial skeleton (face, skull, vertebrae, sacrum and pelvic bones). Fifty-one of the tumors were reported with a diagnosis of chordoma.

The medical records including roentgenographic statements were available for all patients, as well as autopsy reports when postmortem examination had been done (20 cases). The patients were followed until death or until mid-1978. A complete follow-up was achieved.

Biopsy, operation and autopsy specimens were re-examined. All tissue blocks were collected and new sections were made and stained with hematoxylin and eosin, van Gieson, PAS according to McManus with and without diastase, Alcian blue at pH 2.5 and 8.0, or 0.5, with and without prior digestion with bovine testicular hyaluronidase, and with the CEC-technique according to Scott. The results of the histochemical study characterizing the glucosaminoglycan content of chordomas have previously been reported (Kindblom & Angervall 1975).

RESULTS

Pathology

Of the 51 tumors registered at the Swedish Cancer Registry in the years 1958–1970, two cases were excluded at the re-examination; one of them was diagnosed as a chondrosarcoma and in the other insufficient material was available for diagnosis. Two cases of chordoma diagnosed during this period from our own files, primarily diagnosed as adenocarcinoma and myxoid liposarcoma, respectively, were added to the series, which thus consisted of 51 cases of chordoma.

Gross appearance

In cases with a detailed macroscopic description the chordomas were usually described as lobulated and with grayish transparent myxoid or more firm, chondroid, cut surfaces. The margins against surrounding soft tissues were often considered to be distinct, while a distinct demarcation from the involved bone usually was lacking. The size of the tumors could only be roughly estimated from reports of the operation or histopathologic examination or from the radiographic examination. The size of the sphenoccipital chordomas varied from a "pea" to one tumor "filling the whole of the middle cranial fossa" with an estimated median size of 3–4 cm; the vertebral tumors ranged from "two marbles" to "a Tangerine"; the sacrococcygeal chordomas varied in size from 4 to 18 cm in largest diameter with a median of 10 cm.

Light microscopic appearance

The tumors characteristically showed fibrous strands dividing the tumor into lobules, which were built up by abundant mucin-containing solid sheets or delicate strands of tumor cells. The tumor cells were mostly stellate or polygonal with eosinophilic, finely vacuolated cytoplasm. The characteristic syncytial strands passed from the periphery to the center of the lobules. The characteristic physaliphorous cells with large intracytoplasmic vacuoles were seen in most instances but were rarely a dominating cell type.

Cellular and nuclear pleomorphism was usually not prominent and the mitotic activity was low; however, in three of the sacrococcygeal chordomas there were cellular areas with prominent pleomorphism and abundant mitotic figures.

Staining with the PAS-method with and without prior digestion with diastase showed abundant glycogen in many of the tumor cells. The result of the histochemical study characterizing the glucosaminoglycan content of the mucous matrix has previously been reported (Kindblom & Angervall 1975). The staining results obtained in this study indicated the presence of chondroitin-4 and -6 sulphate and in some instances also keratosulphate.

Two of the spheno-occipital chordomas had an appearance of a "chondroid chordoma" as described by Heffelfinger et al. (1973), i.e. mixed with the typical chordoma structures were, in one case, areas of mature apparently chondromatous hyaline tissue with foci of calcification and in the other case partly myxoid, partly hyaline chondromatous areas with distinct cellular and nuclear pleomorphism as in a chondrosarcoma.

The metastases examined had an appearance consistent with that of the primary tumors, and evidence of dedifferentiation in metastases or recurrences was never recorded.

Anatomical distribution

This is shown in Figure 1. Twenty-nine chordomas were situated in the sacrococcygeal region (57 per cent); 14 in the spheno-occipital region (27 per cent) and eight in the vertebrae (16 per cent) (seven in cervical vertebrae and one in a thoracic vertebra).

Age and sex

The age distribution is shown in Figures 2 and 3. The age ranged from 6 to 87 years. The peak incidence was in the 6th and 7th decades. The mean age in the total material was 57 years. The mean age was 63 years in the sacrococcygeal group, 53 years in the spheno-occipital group and 47 years in the vertebral group. The male/female ratio was 26/25.

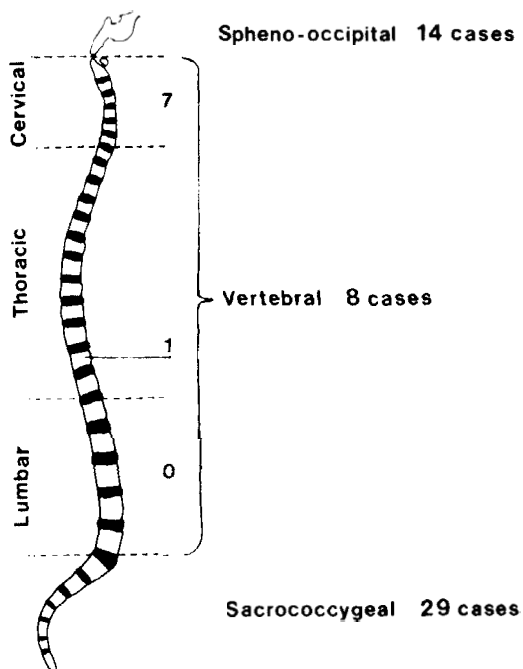


Figure 1. Anatomical distribution of 51 chordomas diagnosed in Sweden 1958-1970.

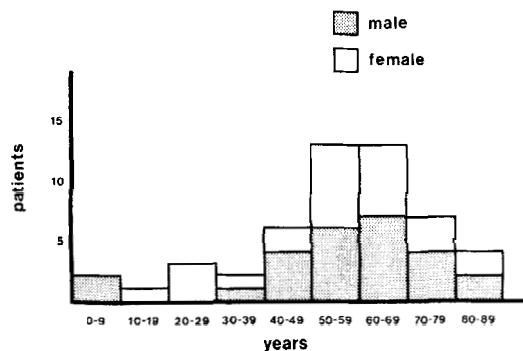


Figure 2. Age and sex distribution of 51 patients with chordoma diagnosed in Sweden 1958-1970.

Symptoms and signs

Sacrococcygeal group - 29 patients. The interval between initial symptoms and date of diagnosis ranged from 10 months to 10 years, median time 1 year. The early symptoms are given in Table 1. Two patients died without known symptoms of tumor; the diagnosis was made after autopsy.

At the time of diagnosis sacrococcygeal pain was experienced by almost all patients. Nine patients had noted rectal and/or urinary dysfunction.

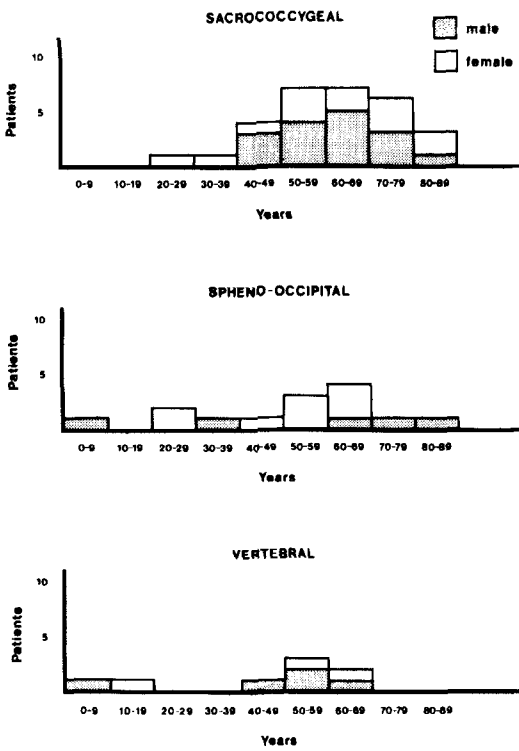


Figure 3. Age and sex distribution of the patients in relation to site of 51 chordomas diagnosed in Sweden 1958-1970.

tion. At clinical examination a palpable mass in the sacral or gluteal region was recorded in 12 patients. Twenty-six patients were examined by rectal palpation, 24 of these had a presacral tumor. All patients presenting a tumor posteriorly had a palpable presacral expansion as well. Six patients had deficient sensibility of sacral dermatomes. Two of these had deficient motor function of the lower extremities as well. Vesical or rectal function was not investigated by laboratory methods.

Spheno-occipital group – 14 patients. The interval between initial symptoms and date of diagnosis ranged from 2 weeks to 5 years, median time 1 year. The early symptoms are given in Table 2. Two patients died without known symptoms of tumor; the diagnosis was made after autopsy.

At clinical examination disturbance of cranial nerve function was recorded in nine patients.

Most commonly observed was diplopia (eight patients), owing to loss of function of the oculomotor and abducense nerves. Two of these patients had visual impairment as well. The trigeminal nerve was affected in five patients and the facial nerve in two patients. The other cranial nerves were affected more infrequently. Three patients did not demonstrate cranial nerve affection but had other signs of disturbance of the central nervous system such as ataxia, epilepsy, hemiparesis or cerebellar herniation.

Vertebral group – 8 patients (seven cervical and one thoracic vertebral lesion). The interval between initial symptoms and date of diagnosis

Table 1. Early symptoms in 29 patients with sacrococcygeal chordoma

Early symptoms	No. of patients
Local pain	7
Local pain and sciatica	8
Constipation	7
Urinary dysfunction	2
Local mass	3

Table 2. Early symptoms in 14 patients with spheno-occipital chordoma

Early symptoms	No. of patients
Diplopia and headache	4
Trigeminal neuralgia	1
Facial nerve palsy	1
Tinnitus	1
Acute cerebellar herniation	2
Psychomotor epilepsy	1
Ataxia and spasticity	2

Table 3. Early symptoms in 8 patients with vertebral chordoma

Early symptoms	No. of patients
Local pain	3
Nasopharyngeal obstruction	2
Cervical mass	1
Hemi- or paraparesis	2

ranged from 3 months to 2 years, median time 10 months. The early symptoms are listed in Table 3.

Two patients had a history of trauma.

A 65-year-old man had a traffic accident resulting in a fracture of the C₂-vertebra. Soon after the accident he noted a tumor of his neck. Four years later he experienced pain in this region and progress of tumor size. Five years after the accident, biopsy revealed the true nature of the tumor.

A 15-year-old girl fell and hit her head on a stone floor. She experienced immediate headache, dizziness and weakness of her left hand. During the following months there was a slow progress of paresis of both left extremities and later also of her right extremities. A laminectomy was performed because of suspicion of disc protrusion between the C₃ and C₄ vertebrae, resulting in a slight improvement in her condition. Further investigation revealed a pharyngeal mass and 5 months after her trauma a chordoma in the C₃-C₄ region was removed. The patient was symptom-free 14 years postoperatively.

At the time of diagnosis clinical examination revealed hemiparesis in two patients and transversal medullary lesion in one patient. One patient had an affection of the glossopharyngeal nerve with paralysis of the soft palate. Two patients had visible epipharyngeal tumor and one only had stiffness of her neck.

Radiography

Sacrococcygeal chordomas – 29 cases. Plain radiography was performed in 26 patients. A partial destruction of the sacrum and occasionally also of the coccyx was visible in 24 cases. In two cases the destruction was missed initially but clearly demonstrated at re-examination of the films.

Thin calcifications within the tumor were recorded in four cases.

Angiography was performed in four patients. No pathological tumor vessels could be detected, only dislocation of pelvic arteries. In two further cases angiography was performed at recurrence of the tumor, also without any demonstrable tumor vessels.

Barium enema revealed dislocation of the rectum in 7 out of 12 examined patients.

Myelography was performed in four patients in all of whom an occlusion of the dural sac distally could be seen.

Spheno-occipital chordomas – 14 cases. Plain radiography was performed in 11 patients. Destruction of the dorsal aspect of the sella tursica and clivus regions was seen in seven patients. In four patients no skeletal abnormality could be detected. Thin tumor calcifications were seen in three patients. A soft tissue mass was seen in the epipharynx on the films of two patients.

Angiography was performed in nine patients. No pathologic tumor vessels could be detected. A dislocation of the internal carotid artery was described in four patients and a dislocation of the basilar artery in one patient.

Ventriculography was performed in nine patients in eight of whom an expansive lesion was disclosed.

Vertebral chordomas – 8 cases. Plain radiography was performed in all patients. In three an evident skeletal destruction was detected initially. In the patient with a thoracic chordoma destruction was not seen until 4 months after the first examination. In two cervical chordomas only minor changes, interpreted as a dilatation of intervertebral foraminae were noted. In two patients a retropharyngeal soft tissue mass was the only radiographic finding.

Angiography was performed in two patients. In one a dislocation of a vertebral artery was evident; in the other no pathological changes were seen.

Gas myelography was performed in three patients. In one cervical case a disc protrusion was suspected and in the thoracic case a dislocation of the dural sac was seen. The third myelography was a failure.

Treatment and follow-up

Sacrococcygeal chordomas – 29 patients. Six patients were treated surgically and 12 were treated by surgery and radiotherapy. The high recurrency rate necessitated 33 operations on these 18 patients. Seven patients were treated by radiation only. Four patients were not treated. The average survival time was 4.6 years. Twelve patients lived 5 years or more and four patients were still alive 8 to 10 years after diagnosis, all with evidence of chordoma (Figures 4 and 5). Three of these four

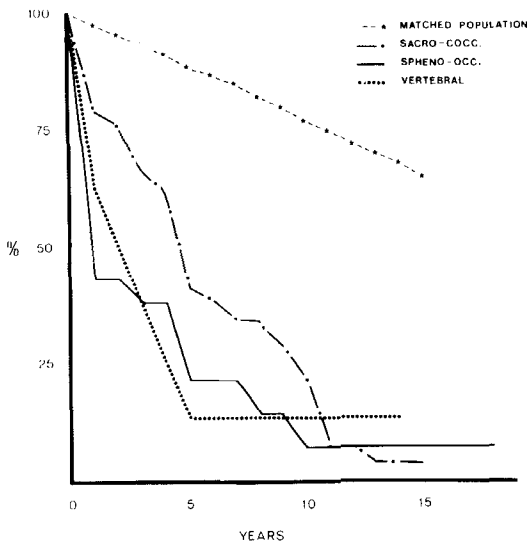


Figure 4. Survival curves in relation to anatomical location of 51 chordomas diagnosed in Sweden 1958-1970. The computed survival curve of a matched population is also given.

patients had both surgery and radiotherapy. Two of the 25 dead patients died of unrelated causes. For details see Table 4.

Spheno-occipital chordomas – 14 patients. Seven patients were treated surgically and two patients were treated by surgery and radiotherapy. Altogether 15 operations were done on these nine patients. No patient had radiotherapy as the only treatment. Five patients were not treated. The average survival time was 3.3 years. Three patients lived 5 years or more and two were still alive 9 and 18 years after diagnosis with evidence of chordoma (Figures 4 and 5). The first patient was treated surgically and the second had both

surgery and radiotherapy. Two of the 12 dead patients died of unrelated causes. For details see Table 5.

Vertebral chordomas – 8 patients. Four patients were treated surgically and two patients were treated by surgery and radiotherapy. Altogether nine operations were done on these six patients. Two patients were treated by radiation only. The average survival time was 3.5 years. There was only one long-time survivor in this group: a patient who was operated on for cervical chordoma was still living without evidence of disease 14 years postoperatively. One of the seven dead patients died of unrelated cause. For details see Table 6.

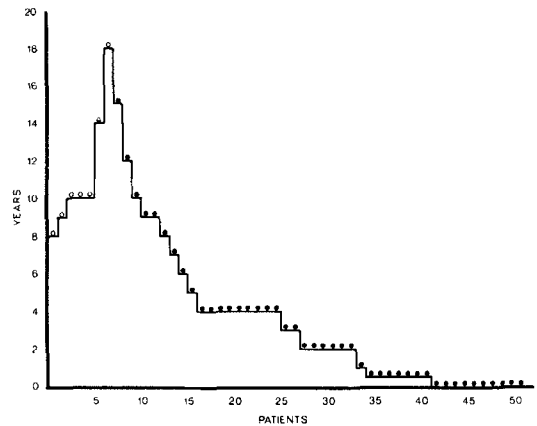


Figure 5. Survival pyramid for 51 patients with chordoma diagnosed in Sweden 1958-1970. Every filled circle represents a dead patient, every unfilled circle a living patient with evidence of chordoma. The unfilled triangle symbolizes the only living patient without signs of chordoma. The vertical axis indicates survival time.

Table 4. Data on treatment and survival of 29 patients with sacrococcygeal chordomas

Treatment	No. of patients	No. of operations	Radiation dose (rad)	Mean survival (years)
Surgery	6	17		5.7
Radiotherapy	7		2700-6000	5.4
Surgery + Radiotherapy	12	16	*	6.6
No treatment	4			0.6

*Data on doses not available.

Table 5. Data on treatment and survival of 14 patients with spheno-occipital chordomas

Treatment	No. of patients	No. of operations	Radiation dose (rad)	Mean survival (years)
Surgery	7	12		3.4
Radiotherapy	0			
Surgery + Radiotherapy	2	3	*	4 and 18, resp.
No treatment	5			<1

*Data on doses not available.

Table 6. Data on treatment and survival of 8 patients with vertebral chordomas

Treatment	No. of patients	No. of operations	Radiation dose (rad)	Mean survival (years)
Surgery	4	7		6.1
Radiotherapy	2		5000-5400	1.4
Surgery + Radiotherapy	2	2	*	<1
No treatment	0			

*Data on doses not available.

Metastases

The overall rate of metastases was 29 per cent. The postmortem examination rate was 39 per cent.

Sacroccygeal chordomas – 29 patients. The rate of metastases was 45 per cent (13/29). In eight patients the metastases were observed prior to death, 1 to 9 years after diagnosis. In five patients the metastases were detected at postmortem examination (examination rate 41 per cent). The sites of the metastases were the lungs (five patients) skeleton (four patients), liver (three patients), peritoneum (two patients) and pancreas (one patient). Two patients had metastases in several regions.

Spheno-occipital chordomas – 14 patients. No metastases were observed in this group. The postmortem examination rate was 36 per cent (5/14).

Vertebral chordomas – 8 patients. The rate of metastases was 25 per cent (2/8). The regional lymph glands were involved in both patients and one of them also had pulmonary metastases, which were detected at postmortem examination (examination rate 37 per cent).

Survival

The survival in relation to tumor site is given in Figure 4. The computed survival rate of a population with the same age and sex distribution as that of the chordoma patients is also given in Figure 4.

The survival pyramid of all 51 chordoma patients is given in Figure 5 and survival pyramids in relation to mode of treatment in Figure 6.

Incidence

The yearly incidence was 0.51 cases per one million inhabitants. Chordomas constituted 17.5 per cent of all primary malignant bone tumors of the axial skeleton (face, skull, vertebrae and pelvic bones) and 5.2 per cent of all primary malignant bone tumors reported to the Swedish Cancer Registry during the period 1958–1970.

DISCUSSION

The distinct predilection for the ends of the spinal column, the gross appearance of a soft, lobulated, translucent, greyish tumor and the characteristic microscopic appearance of lobules composed of syncytial strands of cells and physaliferous cells in

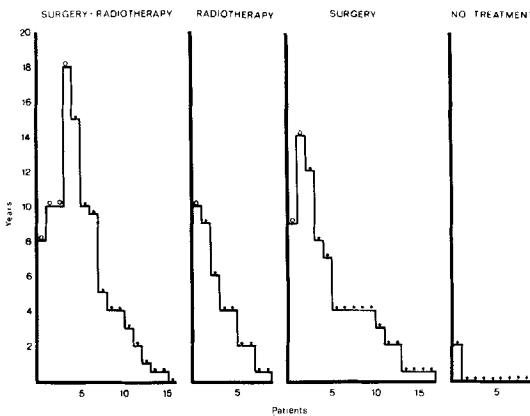


Figure 6. Survival pyramids in relation to treatment of 51 patients with chordoma diagnosed in Sweden 1958–1970. Every dot represents a dead patient, every unfilled circle a living patient with chordoma. The unfilled triangle symbolizes the only living patient without signs of chordoma. The vertical axis indicates survival time.

an abundant mucoid matrix usually make it possible for the pathologist aware of this tumor to make a correct diagnosis. Thus, 49 of the 51 cases reported as chordomas could be accepted as such at review. However, this study and other observations by us have shown that chordoma may be erroneously diagnosed as a myxoid soft tissue tumor such as myxoid liposarcoma and myxofibrosarcoma or chondrosarcoma or even adenocarcinoma. The estimated incidence figure of 0.51 per million population is thus a minimum figure. In the only previously reported population study the estimated incidence was half the one found in our study (Paavolainen & Teppo 1976).

The presence of areas with prominent pleomorphism and high mitotic activity in three sacrococcygeal chordomas did not alter the clinical course in these cases. This is in accordance with previous observations by Dahlin & McCarthy (1952). A feature of prognostic significance, however, is the presence of chondroma- or chondrosarcoma-like areas within chordomas at the skull base – by Heffelfinger et al. (1973) named “chondroid” chordomas. These tumors often show an indolent course. In the present series only two of the speno-occipital chordomas had this appearance, one of which was a small tumor accidentally found at autopsy; the other patient

died after 8 years from a recurrence (the average survival time for the speno-occipital chordomas was 3.3 years).

The value of histochemical characterization of the glucosaminoglycan contents of these tumors has previously been emphasized (Kindblom & Angervall 1975): the identification of acid sulphated glucosaminoglycans helps to exclude the possibility of myxoid soft tissue tumors, such as myxoid liposarcoma and myxofibrosarcoma, but does not help to distinguish chordoma from chondrosarcoma.

The anatomical distribution and the age at onset were consistent with previous reports. However, the male predominance previously shown by several investigators (Mabrey 1935, Dahlin 1970, Higinbotham et al. 1967, Ariel & Verdu 1975, among others) could not be found in the Swedish series.

The symptoms at the time of diagnosis can be attributed to the size of the tumor and the location. The sacrococcygeal chordomas were usually already large when the patient first sought medical care, mostly because of local pain and symptoms caused by mechanical pressure on sacral nerves, giving sciatica and bladder and bowel disturbances. The smaller size of the skull base chordomas is obviously due to their deleterious location, giving earlier focal symptoms in the central nervous system.

The observation that trauma in two cases preceded the development of chordoma may be of etiologic interest. Chordomas appearing after trauma have previously been reported in as many as 20 per cent of cases (Gray et al. 1975). Furthermore, there are reports on experimental induction of chordoma by opening intervertebral discs and exposing the nucleus pulposus (Ribbert 1894, Congdon 1952).

The characteristic radiographic finding of the sacrococcygeal chordomas was bone destruction, which was observed in 86 per cent of the cases. Other findings reported to be characteristic of sacrococcygeal chordoma are expansion of cortex, trabeculation and calcification (Hsieh & Hsieh 1936, Utne & Pugh 1955). In our series expansion of the cortex or trabeculation was not specifically reported by the radiologists. Calcifications were noted in only 15 per cent and were

thus not a common finding. Radiographic bone involvement was also a fairly common feature of the spheno-occipital chordomas (7 out of 11 radiographically examined patients). Ventriculography proved to be a helpful method of demonstrating the intracranial neoplasms (in eight out of nine examined patients). Today one might expect CT-scan to be an equally effective method. Vertebral chordomas seem not to differ radiographically from other tumors of the spine. In the present series only three of the eight patients had evident bone destruction initially, although minor abnormalities could be noted in the films of a further four patients.

Our follow-up results showing a very high recurrence rate and a poor prognosis are consistent with most previous reports. The surgical intention was in most instances tumor enucleation or palliative resection and in exceptional cases radical removal. This seems only to have been successful in a cervical chordoma, where the patient is alive without evidence of disease 14 years postoperatively. The treatment of chordoma most often advocated in the literature is radical removal. The localization of the tumor in the axial skeleton, however, makes this kind of surgery hazardous as important nerve structures may have to be sacrificed and stabilizing structures of vital importance may be damaged. In the sacrococcygeal region extensive resections have proved successful (Hays 1953, Stener & Gunterberg 1978) as all of the sacrum except for the upper half of the first segment may be sacrificed without fatal compromise of stability. The nerves to the bladder and rectum may also be sacrificed and still the patient's life may be surprisingly little affected (Gunterberg 1976). However, a big tumor destroying also the first sacral segment cannot be radically removed with preservation of functioning lower extremities, and the situation is similar in the region of the skull base and vertebrae where vital structures must be preserved. Thus, other methods of treatment must be taken into account.

In many instances radiation has been added to subtotal removal of chordoma, and radiation as the only therapy has also been documented (e.g. Wood & Himadi 1950, Dahlin & McCarthy 1952, Higinbotham et al. 1967, Pearlman &

Friedman 1970, Weldinger 1975). The results vary but in general it seems as if surgery in combination with radiotherapy is superior to radiotherapy alone (Weldinger 1975). The radiation doses recommended are high, the rate of success with tumor doses of over 8000 rad being 80 per cent, whereas smaller doses offer little likelihood of tumor restraint (Pearlman & Friedman 1970). The differing doses given in this series together with the varying locations and sizes of the tumors make it difficult to draw any definite conclusions concerning the benefit of radiation in the treatment. However, it can be noted that of the seven patients still living more than 8 years after diagnosis, four had received combined surgery and radiotherapy and one radiation as the only treatment (Figure 6).

This study confirms that the malignant characteristics of chordoma lie in their critical location, locally aggressive nature and high recurrence rate, although the course is generally long. The high recurrence rate is naturally a result of the impossibility of removing the tumor radically or failure to do so. This might in some instances be due to an unsuitable biopsy route, which if not removed together with the tumor will most probably be the site of recurrence as tumor cells always can be expected to contaminate a biopsy route. Thus, sacrococcygeal tumors, for example, should not be biopsied via the rectum as this would require the rectum to be included with the specimen at the final procedure. A plan to radically remove the tumor must be the first step in any treatment program of chordoma whenever possible. This must be thoroughly considered by the surgeon or team of surgeons who are presented with a case of chordoma.

The rate of metastases varies considerably in different reviews. This seems to be closely related to the follow-up time and the rate of postmortem examinations. In the study by Higinbotham et al. (1967) 43 per cent metastasized in a series of 46 cases and in another series of 20 patients reported by Paavolainen & Teppo (1976) metastases occurred in 60 per cent. In our series 29 per cent of the patients developed metastases, often late in the course; the true incidence is probably higher since only 39 per cent of the patients were examined post-mortem.

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