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# CHORDOMA IN FINLAND

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During the period 1953–1971, 20 cases of chordoma were reported to the Finnish Cancer Registry. Twelve of the patients were males. The mean annual (crude) incidence of chordoma in Finland was $0.30/10^6$ in males, and $0.18/10^6$ in females. Fifteen of the tumours were sacral, three vertebral, and two cranial. Local recurrences were common, and distant metastases were observed in 60 per cent of the cases; this exceeds the proportion usually mentioned in the literature. The commonest treatment was surgery combined with postoperative high-dose irradiation. The relative 5-year survival rate was 35 per cent, and the 10-year rate 18 per cent.

*Key words:* chordoma; bone tumours; histology; treatment; prognosis

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Chordomas arise in the remains of the embryonic notochord. These tumours are rare: up to 1969, less than 700 cases had been published in the literature (Beaugié et al. 1969). Rissanen & Holsti (1967) have reported seven cases of sacrococcygeal chordoma from Finland. This paper concerns the 20 cases of chordoma reported to the Finnish Cancer Registry during the period 1953–1971.

## PATIENTS AND METHODS

During the period 1953–1971, the Finnish Cancer Registry received reports on 20 cases of chordoma. The hospital records concerning the diagnostics and treatment of chordoma were compiled, and the patients were followed up until death, or until June 30, 1974; a complete follow-up was achieved. All tumours had been histologically verified. In 19 instances, re-examination of the sections (L.T.) confirmed the diagnosis of chordoma. In one case, although neither tissue nor slides were obtained, the clinical findings, and the original statement of the pathologist made regarding a specimen taken at operation, were typical of a chordoma.

## RESULTS

Chordomas constituted 1.5 per cent of all the malignant bone tumours reported to the Finnish Cancer Registry during the period 1953–1971. The mean annual (crude) incidence of chordoma in Finland was $0.30/10^6$ in males, and $0.18/10^6$ in females.

Twelve of the patients were males, eight were females (male/female ratio 1.5). Table 1 indicates the age distribution of the patients. The mean age of the patients was 55.5 years. The anatomical distribution of the tumours is given in Table 2.

*Table 1. Age and sex distribution of the patients.*

| Age (years) | Males | Females | Total |
|---|---|---|---|
| 0– 9 | – | 1 | 1 |
| 10–19 | – | – | – |
| 20–29 | – | – | – |
| 30–39 | – | – | – |
| 40–49 | 4 | 1 | 5 |
| 50–59 | – | 4 | 4 |
| 60–69 | 7 | 1 | 8 |
| 70–79 | 1 | 1 | 2 |
| 80– | – | – | – |
| All ages | 12 | 8 | 20 |

*Table 2. Number of patients by anatomical site of the tumour and sex in a series of 20 cases of chordoma.*

| Anatomical site | Males | Females | Total |
|---|---|---|---|
| Sacral | 10 | 5 | 15 ( 75 %) |
| Vertebral | 1 | 2 | 3 ( 15 %) |
| Spheno-occipital | 1 | 1 | 2 ( 10 %) |
| Total | 12 | 8 | 20 (100 %) |

*Table 3. Presenting symptoms in a series of 20 patients with chordoma.*

| Symptom | Sacral N = 15 | Vertebral N = 3 | Cranial N = 2 |
|---|---|---|---|
| Pain | 11 | 2 | – |
| Abnormal growth | 9 | – | – |
| Urinary incontinence | 2 | – | – |
| Fever | – | 1 | – |
| Tetraparesis | – | 1 | – |
| Visual impairment | – | – | 2 |
| Medullary compression | – | – | 2 |

### *History and symptoms*

*Sacral chordomas.* Six patients (40 per cent) gave a history of trauma in the region of the subsequent tumour. The period of time between the trauma and the diagnosis of chordoma varied from one month to 7 years (median 4 years).

Eleven patients (73 per cent) complained of pain, this being the most common symptom (Table 3). A tumour-like growth in the sacral region, alone or accompanied by pain, had been noticed by nine patients (60 per cent); in two patients, the tumour had induced urinary incontinence.

In eight instances, the first diagnosis was incorrect: sciatic syndrome (3), dermoid cyst (4) or prostatic hyperplasia (1). One patient had been subjected to an operation for disc prolapse before the institution of a correct diagnosis of chordoma.

*Vertebral chordomas.* The patient with a thoracic tumour had a history of pain and fever (Table 3); a tumour was palpable on the left side, and initially a false diagnosis of renal abscess was made. Progressive pain experienced in swallowing was the symptom of one of the patients with a cervical tumour. The other, who gave a history of neck distension-like trauma one year earlier was suffering from progressive tetraparesis (Table 3).

*Spheno-occipital chordomas.* Both tumours had induced visual impairment, along with various neurological symptoms (Table 3).

*Duration of symptoms.* Wide variation was apparent in the duration of the symptoms: $< 3$ months in three cases, 3–6 months in eight cases, 6–12 months in seven cases, and more than 1 year (6 years) in one case; the median was 6 months, and the mean 12.7 months.

### *Findings*

On first examination, 13 patients had a palpable tumour dorsally, in the gluteal or sacral region. In eight instances, the tumour could also be found by rectal palpation. A massive tumour was observed in the hypopharynx of one patient with a cervical chordoma.

The destruction of bone, suggestive of a tumour, was observed at the first X-ray examination in 11 patients (73 per cent) with a sacral tumour. One of the cervical

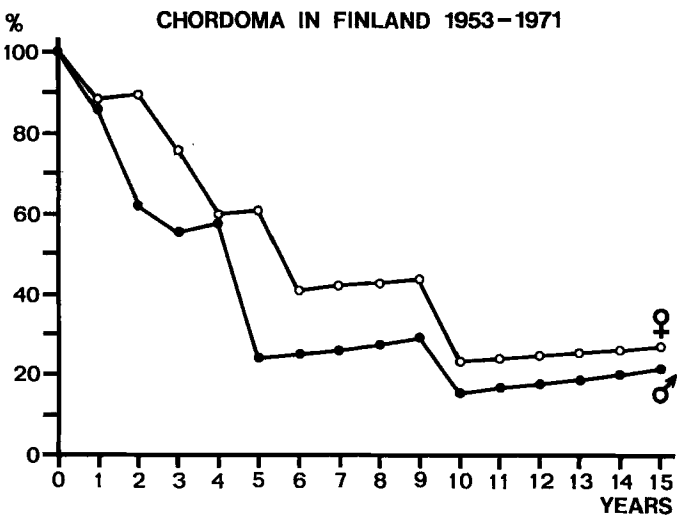


*Figure 1. Relative survival curves (calculated by the actuarial method) of all cases of chordoma diagnosed in Finland during the period 1953–1971 (12 males, 8 females).*

tumours had induced destruction of the corpus C IV, visible on tomography.

*Histology*

The first histological diagnosis was obtained from a biopsy specimen in eight cases, from an operation specimen in 11 cases, and at autopsy in one case. In 18 cases, one or more tissue specimens from the primary tumour were available for histological study; in one case, the only specimen was from the recurrent tumour. Sixteen chordomas were considered as "typical", taking the form of lobular tumours composed of loosely-arranged cells, with rather plemomorphic nuclei, and often foamy cytoplasm; mitoses were few. Varying amounts of intercellular homogenous mucin-like material were discernible, together with necrosis.

Three tumours exhibited atypical features: The general structure of one of these tumours resembled adenocarcinoma, but small areas, with typical chordomatous pattern, revealed the real nature of the tumour. The two remaining tumours displayed mostly undifferentiated tissue with areas suggesting chordoma.

*Mode of treatment*

Twelve of the 15 patients with *sacral* chordomas were initially subjected to operation; five of these operations were considered macroscopically radical. Postoperative radiotherapy was instituted in nine cases (Table 4).

One of the three patients with *vertebral* tumours (located cervically) was

*Table 4. Mode of treatment in a series of 20 patients with chordoma. Figures in parentheses indicate postoperative radiotherapy.*

| Localization of the tumour | No. of patients | Radical operation | Palliative operation | Radiotherapy alone | No treatment |
|---|---|---|---|---|---|
| Sacral | 15 | 5 (4) | 7 (5) | 2 | 1 |
| Vertebral | 3 | 1 | – | 1 | 1 |
| Spheno-occipital | 2 | – | 1 | – | 1 * |

* Diagnosed at autopsy.

*Table 5. Localization of the metastases in a series of 20 patients with chordoma.*

| Localization of the metastases | Localization of the primary tumour | | | |
|---|---|---|---|---|
| | Sacral N = 15 | Vertebral N = 3 | Spheno-occipital N = 2 | Total N = 20 |
| Bone | 8 | – | – | 8 |
| Lungs | 5 | – | – | 5 |
| Liver | 2 | 1 | – | 3 |
| Lymph nodes | – | 2 | – | 2 |
| Skin | 2 | – | – | 2 |
| Chest wall | 2 | – | – | 2 |
| No metastases | 6 | – | 2 | 8 |

the subject of radical operation, and one patient (cervical tumour) received radiotherapy alone; no treatment was given in the case with a thoracic tumour (Table 4).

An operation was performed on one of the *spheno-occipital* chordomas, but the other remained untreated (Table 4).

Treatment by X-ray or telecobalt, 3,000–8,200 rad, was administered both to the nine cases subjected to postoperative irradiation, and to the three cases given radiotherapy only. In one case, radiotherapy was supplemented by cyclophosphamide (6,000 mg).

The treatment of locally recurring sacral chordomas was as follows: one case operation only, two cases operation supplemented with postoperative radiotherapy, two cases radiotherapy, and in three cases no treatment. The radiation dose was 3,000–8,000 rad X-ray or telecobalt. Radiotherapy, alone or in combination with cytostatic agents (cyclophosphamide, 5-fluorouracil), was the mode of treatment applied at the metastatic stage of the disease.

*Clinical course*

In 15 cases out of 17, a local recurrence appeared after the initial treatment. The lapse of time from the first treatment to the recurrence was $< 1$ year in three cases, 1–3 years in eight cases, 3–6 years in three cases, and $> 6$ years in one sacral case (12 years).

Distant metastases were observed in 12 cases (60 per cent); in seven of them, several metastases were observed. In three instances the metastases were studied histologically. The commonest sites of metastases were bone, lungs and liver (Table 5). The period of time from primary treatment to appearance of the metastases ranged from one month to nearly 20 years. None of the metastases was present at the time of diagnosis.

The 5-year relative survival rate of the patients was 35 per cent, and the 10-year rate 18 per cent (Figure 1). Three patients were alive at the end of the follow-up period; only one of them was tumour-free.

In one case, hypernephroma was diagnosed four years after a sacral chordoma. Widespread metastases developed, and the patient died from her renal tumour; the chordoma remained local.

## DISCUSSION

The Finnish Cancer Registry covers the entire country. It receives reports of cases of malignant neoplasms from hospitals, pathological laboratories and practitioners. Various check-ups have indicated that the number of cases not reported to the Registry is negligible. It

can be concluded that the series of patients presented in this paper probably represents all of the cases of chordoma diagnosed in Finland during the period concerned, and accordingly that calculation of an incidence rate is justified.

Incidence rates of chordoma have seldom been presented. In Sweden, the mean annual incidence in 1958–1968 was $0.49/10^6$ (Larsson & Lorentzon 1974). The rate in Finland ($0.30/10^6$ in males and $0.18/10^6$ in females) was only one-half of that in Sweden; on the average, only one case of chordoma will be diagnosed annually in Finland (population 4.6 million). In the material compiled from the Swedish Cancer Registry from 1959 to 1965, chordomas constituted 3.9 per cent of all bone tumours (Cancer Incidence in Sweden 1959–1965). In Finland, the corresponding figure was 1.5 per cent.

The anatomical distribution of tumours along the spine is related to the source from which the patients are drawn (radiotherapy clinic, neurological unit, cancer registry, etc.). Most often the sacrococcygeal region is affected; this was also found to apply to our series. The age and sex distribution of our series corresponds with many other series which have indicated a male preponderance, and the highest incidence in middle and old age (e.g. Higinbotham et al. 1967).

As wide variations were apparent in the modes of therapy, and the patients concerned underwent treatment over a long period of time, this series does not provide a basis for any conclusion being drawn as to the most effective treatment. Of course, a radical operation seems desirable, and probably offers the only hope for the patient's permanent cure (Gentil & Coley 1948). Unfortunately, anatomical circumstances mean that radicality is the exception rather than the rule (Pearlman & Friedman 1970). Spheno-occipital and vertebral chordomas are hardly ever curable. Although the value of postoperative radiation is difficult to assess, it should be attempted particularly in cases of non-radical operation (Pearlman & Friedman 1970). despite the radio-resistance offered by chordomatous tissue in general.

The reported frequencies of metastases vary, ranging from 0 per cent (Dahlin & MacCarty 1952) to 43 per cent (Higinbotham et al. 1967). During the course of the disease, up to 60 per cent of our cases developed distant metastases; in only three instances, however, was this confirmed by biopsy or at autopsy. On occasion, it has been stated that chordoma is a semi-malignant neoplasm. We regard this as an underestimation: the high frequency of local recurrences, and the fact that metastases are often encountered, clearly indicate that chordoma is a malignant tumour. Admittedly, rather long periods of survival are attainable; this means that chordomas grow slowly. Nonetheless, this demands active treatment of the recurrences, and even of the metastases, although sooner or later the disease may prove fatal. In a series of 46 patients reported by Higinbotham et al. (1967), 9 per cent of the patients were free from disease after a follow-up period of five years. The corresponding figure in our series was 5 per cent after 3 years (1/20).

The histological details of the chordoma do not provide a basis for evaluation of the patient's prognosis (cf. Heffelfinger et al. 1973); a tumour with very little cellular pleomorphism can cause the death of the patient within a few months. The prognosis is largely determined by the extent of the tumour at the time of diagnosis (upon which depends the radicality or non-radicality of the operation), and probably also by some unknown factors of the host, the patient himself.

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