

Cryosurgical Treatment of Sacrococcygeal Chordoma

Report of Four Cases

JAKOB DE VRIES, MD, PhD,* JAN OLDHOFF, MD, PhD,†† AND HENK N. HADDERS, MD, PhD†

Sacrococcygeal chordoma is a rare malignant neoplasm situated in a location adjacent to important structures. Distant metastases are usually rare and occur late. The treatment of choice usually consists of radical surgery, sometimes followed by radiotherapy. Extensive surgical resection is difficult and often causes bladder and/or bowel dysfunction, and the local recurrence rate remains high. In an attempt to diminish both risks, the authors introduced cryosurgery *in situ* as a new treatment modality for chordoma in the sacrococcygeal region. From 1974 to 1980, four patients (two male, two female) with sacrococcygeal chordoma were treated with cryosurgery without resection. Two patients had extensive tumors (>10 cm) and could be treated only palliatively. Two other patients with smaller tumors (<10 cm) had radical cryosurgical treatment. Both patients are disease-free 10 and 7 years after cryosurgical treatment. One of the palliatively treated patients is alive with local recurrence 4 years after cryosurgery, the other died of tumor after 5 years. In a cryosurgical lesion, the tissue is completely devitalized; however, the architecture of the tissue in peripheral nerves, large vessels, and bone is preserved and remains as a perfect autograft. Frozen tissue is very susceptible to the hematogenous spread of infection. Therefore, infection prevention is of utmost importance. The authors believe that cryosurgery should have a place in the treatment of sacrococcygeal chordoma.

Cancer 58:2348-2354, 1986.

CHORDOMA IS AN UNCOMMON NEOPLASM that is believed to be derived from areas of primitive notochordal tissue. Although this tumor may arise anywhere along the spinal column, it is most often found at the ends of it: the sacrococcygeal area and the base of the skull (85%). It represents approximately 2% of all malignant tumors of the bone. The main problems are associated with its critical location adjacent to important structures and its locally aggressive nature causing extremely high recurrence rates. Consensus exists about the fact that treatment is very difficult. Susceptibility for radiation therapy is low. In the literature, radical surgical excision with wide margins of excision is advocated as the treatment of choice for sacrococcygeal chordoma. Nevertheless, percentages of local recurrence after radical surgery remain high.¹⁻³

Evaluation of the effects of various modes of surgical treatment is difficult because it is a rare tumor and because

of the length of follow-up required.⁴ Furthermore, extensive surgery in the sacrococcygeal region leads inevitably to damage of important structures resulting in bladder and/or bowel dysfunction.^{5,7} The high local recurrence rate and the complications of extensive surgery led us to investigate another treatment modality: cryosurgery, *i.e.*, tissue destruction using extreme cold.

Cryosurgical treatment of bone tumors is well known from the work of Marcove and Miller^{8,9} and Gage and Erickson.¹⁰ These authors consider locally aggressive and low-malignancy bone tumors particularly suitable for cryosurgical treatment (*e.g.*, giant cell tumors, chondroblastoma, chondrosarcoma Grade I, *etc.*). The method generally consists of surgical exposure of the tumor followed by rapid freezing through direct application of liquid nitrogen (-196° C) with or without previous excochleation of the tumor. Spontaneous thawing to normal temperature is then allowed.⁹⁻¹² Cell death in the cryolesion is achieved by two different types of mechanisms: physicochemical and vascular.¹³ Surrounding structures are protected by isolation material and temperature control. After cryosurgery, the architecture of bone, peripheral nerves, and great vessels remains intact as a perfect autograft.^{11,13} Cryosurgical damages to nerves or great vessels are, therefore, reversible. In vessels, the autograft functions

From the *Division of Oncology, Departments of Surgery and †Pathology, University Hospital, Groningen, The Netherlands.

†Head Professor.

Address for reprints: J. Oldhoff, MD, PhD, Division of Oncology, Department of Surgery, University Hospital, Oostersingel 59, 9713 EZ Groningen, The Netherlands.

Accepted for publication April 11, 1986.

TABLE 1. Cryosurgical Treatment and Follow-Up of Four Patients

Patients	Age/sex (yr)	Tumor		Therapy	Complications	Follow-up			Remarks
		Location	Size (cm)			Years	Local recurrence	Metastases	
1	10/F	S3	4	Radical cryosurgery <i>in situ</i> via dorsal route in one session	Temporary bladder dysfunction in the postoperative period	12	No	No	NED
2	53/M	S1	14	Intra-arterial CDDP infusion Rectosigmoid resection Nonradical cryosurgery via dorsal and ventral route in two sessions Radiotherapy (60 Gy)	Aggravation of the already-existent bladder and bowel dysfunction	5	1.5 yr	No	Died of recurrent tumor
3	64/M	S3	8	Radical cryosurgery <i>in situ</i> via dorsal route in one session	Temporary bowel dysfunction and permanent bladder dysfunction; wound infection after 3 mo, due to neglected urinary infection	7	No	No	NED Needs self catheterization
4	66/F	S2	10	Nonradical cryosurgery <i>in situ</i> via dorsal and ventral route in two sessions	Temporary bladder dysfunction, temporary motor dysfunction in roots L5S1; wound infection	4	3.5 yr	No	Irradiating pain from recurrent tumor

NED: no evidence of disease; CDDP: Cis-diammino-dichloroplatinum (cisplatin).

immediately after thawing; in peripheral nerves, function returns after regrowth of the axons. So the treated area in cryosurgery can be larger than the area treated by resection. Furthermore, in cases of sacrococcygeal chordoma there is no need for resection of the sacrum because the frozen tissue is dead. This diminishes the operative trauma. A cryosurgical lesion in a hollow organ (bladder, bowel) will cause a perforation and subsequent problems plus infection of the frozen sacrum; therefore, these organs should be carefully protected during cryosurgery.

Patients and Treatment

From 1974 to 1980, four patients in our hospital were treated cryosurgically for a sacrococcygeal chordoma (Table 1). There were two male and two female patients, aged 10 to 66 years. In Patients 2 and 4, the tumors were so extensive (>10 cm) that neither resection nor cryosurgery could be performed radically. They were treated pal-

liatively by cryosurgery without resection of the tumor. Patients 1 and 3, with smaller tumors (<10 cm), could be treated radically and are described in the Case Reports section.

Presenting symptoms of all four patients consisted of pain and tenderness in the sacrococcygeal region and in Patient 2, bladder dysfunction already existed on admission. All patients presented a sacral tumor at rectal examination and diagnosis was confirmed histologically after incisional biopsy via the dorsal route.

Cryosurgical treatment consisted of the exposure of the tumor from the dorsal route, followed by pouring liquid nitrogen in a plastic "tube" fixed on the tumor surface (Figs 1A-1G). The plastic "tubes" were made from plastic bottles after removing top and bottom (Figs 1B and 1C). Surrounding tissues were protected by polystyrene material.¹⁴ Temperature was recorded at several points around the tumor to measure the extend of the cryolesion. Dependent on tumor size, the freezing time was about 1



FIGS. 1A-1G. Cryosurgery *in situ*. (A) The tumor is isolated from surrounding soft tissues. (B and C) A plastic bottle is chosen that snugly fits to the surface of the tumor. (D) Polystyrene isolation material is wrapped around the bottle to prevent freezing of surrounding tissue. (E) Liquid nitrogen is poured into the bottle to freeze the tumor. Descending nitrogen damps should be woven away continuously, because it can cause secondary freezing of surrounding tissues. (F and G) White frozen tumor before spontaneous thawing.

hour. Thereafter, spontaneous thawing was allowed to take place, which also took about 1 hour. The wound was closed over vacuum drainage.

In Patients 2 and 4, the tumor was so huge (>10 cm) that radical therapy was impossible, even after cryosurgical treatment *via* laparotomy and the dorsal route, in two

separate sessions about 3 weeks apart. Because of the huge dimensions of the tumor in Patient 2, a rectosigmoid resection was done as a first stage in the therapeutic sequence.

Case Reports

Case 1

A 10-year-old girl had complained of pain in the sacrococcygeal region for several months when she was sent to our department in 1974. Pain sensations increased at sitting and at defecation; micturition was not disturbed. At rectal examination, a sacral tumor was discovered. An x-ray revealed that an osteolytic process was present in the sacrum with soft tissue mass in the presacral space (Fig. 2). Through a small dorsal incision, a biopsy specimen was taken that revealed the diagnosis: chordoma (Fig. 3). Treatment consisted of radical cryosurgery *in situ* (without resection) (Figs. 1A-1G); the sacrum was exposed via a dorsal incision and the rectum was mobilized from the tumor area, which was subsequently frozen with liquid nitrogen. During this procedure, the surrounding soft tissues and rectum were protected with polystyrene material. Temperature was recorded with several thermocouples. Spontaneous thawing was allowed and the wound was closed over vacuum drainage. In the postoperative period, the patient needed urinary catheterization because of urinary retention. After 1 month, the bladder function returned to normal. This patient is well and has had no complaints for more than 10 years after cryosurgery, and is without signs of tumor recurrence (Figs. 4A and 4B).

Case 3

A 64-year-old man presented to our department with complaints of constipation and pain at sitting and lying down. At rectal examination, a sacral tumor was found. An x-ray of the sacrum showed a large osteolytic process involving at least S2 to S5 (Fig. 5). The tumor was at least 8 cm in diameter. A dorsal open biopsy revealed the histologic diagnosis of chordoma.

Treatment consisted of radical cryosurgery (freezing of the tumor *in situ* with liquid nitrogen). In the postoperative period, he had an *incontinentia alvi* and urine retention, which required catheterization. Fecal continence returned in a few weeks but bladder dysfunction persisted and intermittent self-catheterization remains necessary. The wound healed at first and the patient was discharged from the hospital 4 weeks after cryosurgery, but returned 3 months later with a severe urinary infection and an abscess in the frozen area. At operation, the necrotic part of the sacrum was removed with consequent delayed wound healing. No signs of tumor recurrence are present (7 years after cryosurgery, Fig. 6).

Results

Follow-up of these four patients was 4 to 12 years after cryosurgery (Table 1). The two patients (Patients 1 and 3) who had adequate cryosurgery show no evidence of



FIG. 2. Sacrococcygeal chordoma in Patient 1 (x-ray tomography) showing osteolytic process in sacrum with soft tissue mass in presacral space.

disease after 12 and 7 years, respectively. Of the two other patients with not radically treated huge tumors, one (Patient 4) had a local recurrence 4 years after cryosurgery and one (Patient 2) died 5 years after cryosurgery from local disease.

In the postoperative period, all patients had temporary bladder dysfunction that required urinary catheterization. In Patient 4, these problems persisted. Two wound infections were recorded. One occurred 3 months after primary wound healing and was accompanied by a neglected severe cystitis due to bladder dysfunction, the other occurred in the patient who also underwent a rectosigmoid resection.

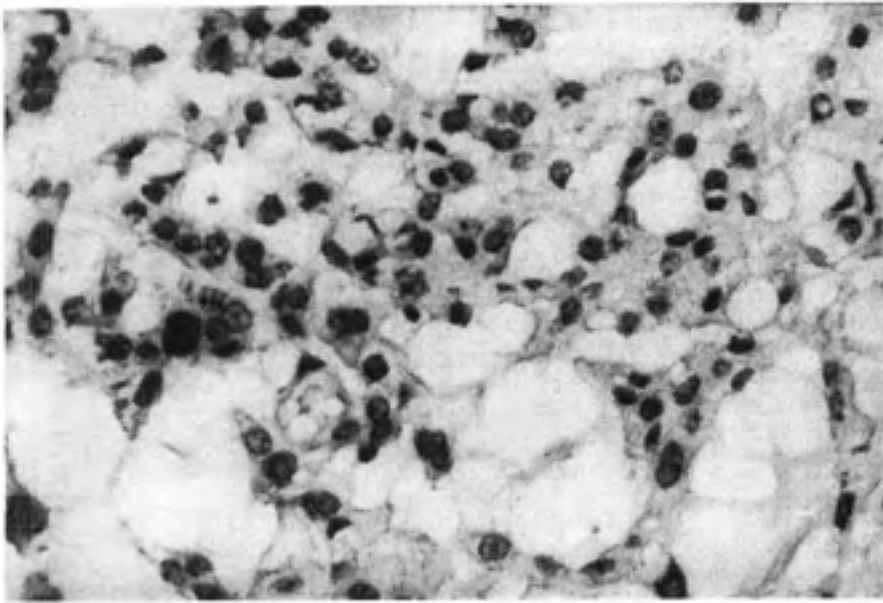
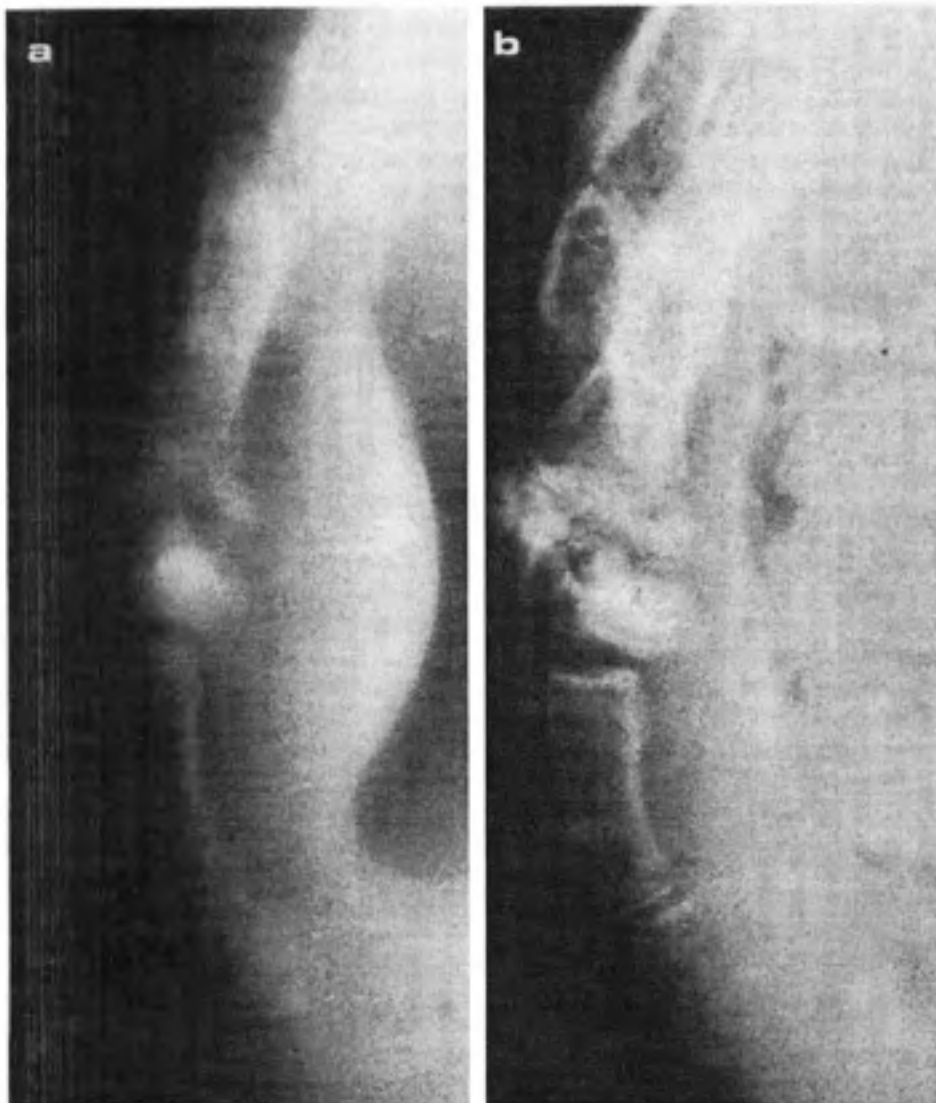


FIG. 3. Histologic aspect of chordoma with typical vacuolized (physaliphorous) cells.



FIGS. 4A AND 4B. X-ray follow-up of sacrococcygeal chordoma in Case 1 before (A) and 10 years after (B) cryosurgical treatment; there are no signs of recurrence.

FIG. 5. Frontal view of a huge sacrococcygeal chordoma in Case 3 before treatment.



Discussion

Many authors have advocated surgical removal of sacral chordomas.¹⁵⁻²⁰ However, the localization, the extent of the tumor, and concern about the loss of sacral nerve roots have led to incomplete resections.¹⁻³ In our opinion, this is the main reason for the high percentages of local recurrence after surgical treatment of chordoma. Adjuvant radiation therapy does not appear to be of curative benefit. Furthermore, extensive surgery leads to damage of important structures and results in bladder and/or bowel dysfunction.

Because cryosurgery in the treatment of bone tumors is particularly effective in locally aggressive lesions, and because damage to nervous structures can be reversible, we chose this treatment modality in the management of sacrococcygeal chordoma.

Due to the small figures in this presentation, conclusions are premature, but in our opinion, these first results are encouraging. However, as all patients showed bladder dysfunction after this treatment, there is a high risk of urinary infection. In this patient group, with a large amount of (cryo)necrotic tissue, the urinary infection can lead to secondary (hematogenous) infection resulting in complete necrotic breakdown, particularly of the frozen bone. So precautions against infection should be taken during the operation and for a long time, postoperatively. Preoperatively, these patients should have an empty large



FIG. 6. X-ray follow-up of sacrococcygeal chordoma in Case 3: a large defect in the sacrum after cryosurgical treatment and subsequent infection that led to necrotic breakdown. However, there were no signs of recurrence.

bowel, and during the operation, a suprapubic urinary catheter should be given. This can be removed when bladder function has normalized, which occurred in three of four patients, confirming that nerve lesions can be reversible.

Cryosurgical treatment is not a difficult method but needs vigorous temperature control, protection of rectum and surrounding soft tissue and skin, strict asepsis during operation, and infection prevention, postoperatively. As mentioned before, there is no need for resection of the sacrum, this increases the operative trauma.

REFERENCES

- Huvos AG. Chordoma. In: Huvos AG, ed. *Bone Tumors: Diagnosis, Treatment and Prognosis*. Philadelphia: WB Saunders, 1979; 373-391.
- Eriksson B, Gunterberg B, Kindblom LG. Chordoma: A clinicopathologic and prognostic study of a Swedish National Series. *Acta Orthop Scand* 1981; 52:49-58.
- Karakousis CP, Park JJ, Fleming R, Friedman M. Chordomas: Diagnosis and management. *Am Surg* 1981; 47:497-501.
- Mindell ER. Chordoma. *J Bone Joint Surg [Am]* 1981; 63:501-505.
- Kaiser TE, Pritchard BJ, Unni KK. Clinicopathology study of sacrococcygeal chordoma. *Cancer* 1984; 54:2574-2578.
- Swinton NW, Lehman G. Presacral tumors. *Surg Clin North Am* 1958; 38:849-857.
- Gunterberg B, Kewenter J, Petersen I, Stener B. Anorectal function after major resection of the sacrum with bilateral or unilateral sacrifice of sacral nerves. *Br J Surg* 1976; 63:546-554.
- Marcove RC, Miller TR. The treatment of primary and metastatic localised bone tumors by cryosurgery. *Surg Clin North Am* 1969; 49:421-426.
- Marcove RC. A 17-year review of cryosurgery in the treatment of bone tumors. *Clin Orthop* 1982; 163:231-234.
- Gage AA, Erickson RB. Cryotherapy and curettage for bone tumors. *J Cryosurg* 1968; 1:60-65.
- Oeseburg HB. Cryochirurgische behandeling van enkele beentumoren. Thesis, Groningen, 1977; 55-60.
- Oeseburg HB, Rogge CWL, Schraffordt Kooops H et al. Cryosurgical treatment of aneurysmal bone cysts. *J Surg Oncol* 1978; 10:9-13.
- Ablin RJ. *Handbook of Cryosurgery*. New York: Marcel Dekker, 1980; 15-68.
- Oldhoff J. Cryochirurgische behandeling van benigne bottumoren. In: Visser GJP, Vermeij A, Scherpbier AJJA, eds. *Chirurgisch Jaarboek 1982*. Assen: van Gorcum, 1983; 69-73.
- MacCarthy CS, Waugh JM, Mayo CW, Coventry MB. The surgical treatment of presacral tumors: A combined problem. *Proc Staff Meet Mayo Clinic* 1952; 27:73-84.
- Schackelford RT, Martin Rhode C. Sacrococcygeal chordoma: Its surgical treatment. *Ann Surg* 1955; 141:952-966.
- Gray SW, Singhabandhu B, Smith RA, Skandalakis JE. Sacrococcygeal chordoma: Report of a case and review of the literature. *Surgery* 1975; 78:573-582.
- Localio SA, Francis KC, Rossano PG. Abdominosacral resection of sacrococcygeal chordoma. *Ann Surg* 1967; 166:394-402.
- Localio SA, Eng K, Ranson JHC. Abdominosacral approach for retrorectal tumors. *Ann Surg* 1980; 191:555-559.
- Rich TA, Schiller A, Smit HD, Mankin HJ. Clinical and pathologic review of 48 cases of chordoma. *Cancer* 1985; 56:182-187.