Cryosurgical Treatment of Sacrococcygeal Chordoma

Report of Four Cases

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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.


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.1–5

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.6,7 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 Miller8,9 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 excoclleration 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.11,12 Cryosurgical damages to nerves or great vessels are, therefore, reversible. In vessels, the autograft functions
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 palliatively 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. 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
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 re-
section was done as a first stage in the therapeutic se-
quence.

Case Reports

Case 1

A 10-year-old girl had complained of pain in the sacrococ-
cygeal region for several months when she was sent to our de-
partment 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 osteo-
lytic 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 (with-
out 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 pro-
tected 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 com-
plaints 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 incontinence alvi and urine retention, which required
catheterization. Fecal continence returned in a few weeks but
bladder dysfunction persisted and intermittent self-catheteriza-
tion 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 cryosur-
gery, 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
disease after 12 and 7 years, respectively. Of the two other
patients with not radically treated huge tumors, one (Pat-
ient 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 infec-
tions 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.

FIG. 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.
Discussion

Many authors have advocated surgical removal of sacral chordomas. However, the localization, the extent of the tumor, and concern about the loss of sacral nerve roots have led to incomplete resections. 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
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