J Korean Med Sci 2000; 15: 601-3
ISSN 1011-8934

Hemophilic Pseudotumor of the Ulna Treated with Low Dose Radiation Therapy
: A Case Report

We report a case of hemophilic pseudotumor in the ulna of a 6-year-old boy treated with radiation therapy. A total dose of 900 cGy in 6 fractions was given in 6 consecutive days. Progression of cystic changes was halted within a month. New bone formation and trabeculation were found on the 4th month. Complete healing of the lesion and bony replacement were found on the 12th month. The patient was followed up to 72 months and there was no evidence of recurrence and no bone growth disturbance. Radiation therapy can be an effective alternative modality in treating hemophilic pseudotumor.

Key Words: *Hemophilia; Pseudotumor; Radiotherapy*

Jin Oh Kang, Yoon Je Cho*, Myung Chul Yoo*, Seong Eon Hong

Departments of Radiation Oncology and Orthopedic Surgery*, KyungHee Medical Center, Seoul, Korea

Received: 27 December 1999
Accepted: 8 March 2000

Address for correspondence
Jin Oh Kang, M.D.
Department of Radiation Oncology, KyungHee Medical Center, 1 Hoiki-dong, Dongdaemun-gu, Seoul 130-702, Korea
Tel: +82.2-958-8664, Fax: +82.2-962-3002
E-mail: kangjino@khmc.or.kr

INTRODUCTION

Hemophilic pseudotumor of the bone is a rare disease occurring in approximately 1% of patients with severe hemophilia as a long-standing complication. It is defined as a progressive cystic swelling by recurrent hemorrhage with roentgenographic evidence of bone involvement (1).

There are very limited number of reports about radiation therapy for hemophilic pseudotumors because such patients are referred only when both surgery and conservative managements are no longer feasible.

We report a case of hemophilic pseudotumor successfully treated with radiation therapy.

CASE REPORT

A 6-year old boy with hemophilia A was admitted with a history of the left elbow swelling of more than 1 year duration. At the time of administration, he complained of a painful mass in his left elbow. Physical examination revealed a 4 cm-sized tender mass without limitation of motion in his elbow. Laboratory tests were as follows: prothrombin time, 88.9 sec; factor VIII, 1.0%; factor IX, 63%; and factor VIII inhibitor, 0.18 BU/mL. X-ray showed widening of the olecranon notch with periosteal reactions around the huge cyst in the proximal ulna (Fig 1A).

The initial therapeutic recommendations were to perform curettage and bone graft but his parents refused operation due to operation risks. He was managed with factor VIII 50 U/kg, twice a day due to severe factor VIII deficiency and was referred to the Department of Radiation Oncology.

Radiation therapy was then instituted. Total dose of 900 cGy in 6 fractions over 6 consecutive days was given by 6 MV X-ray with SSD 100 cm, AP 1 port technique. The radiation portal was designed to include the roentgenographic lesion with adequate margins in all directions but the epiphyseal plate of the distal humerus was shielded. The patient complained of slight aggravation of left elbow swelling during the course of radiation therapy, which soon disappeared afterward. Factor VIII was discontinued shortly after radiation therapy.

The follow up X-ray showed that expansion of the cystic lesion was halted and trabecula density in the cyst slightly increased within a month. The X-ray taken in 4 months of radiation therapy revealed size reduction of the cystic lesion and definite increase of trabecular density within the cyst (Fig 1B). Further follow up X-ray showed a definite evidence of new bone formation at 13 months after radiation therapy (Fig 1C) and hemophilic

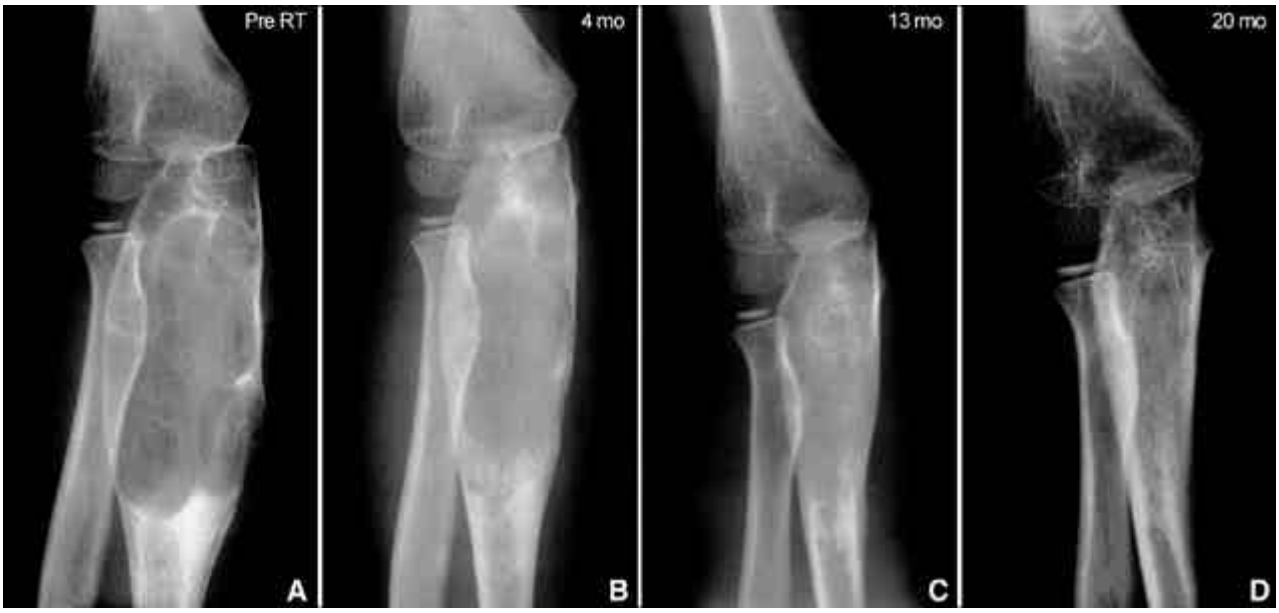

Fig. 1. A) Pre-RT: Widening of olecranon with periosteal reactions around the huge cyst in the proximal ulna. B) Post-RT 4th month. C) Post-RT 13th month. D) Post-RT 20th month: the cyst is completely replaced with new bone.

cyst was completely replaced with new bone at 20 months after radiation (Fig 1D). Until 72 months after radiation therapy, the patient had been doing well with his elbow without any evidence of recurrence.

DISCUSSION

For the diagnosis of hemophilic pseudotumor of bone, considering 10 to 20 percent of hemophilia patients have antihemophilic factor inhibitors, invasive diagnostic techniques such as aspiration are not warranted because severe complication risk is so high. Therefore, noninvasive techniques such as x-ray should be selected first. Periosteal reaction is frequent and regarded as a key sign. In early stage, periosteal reactions might be so minimal that soft-tissue mass is the only finding. In later stage, the entire length of bone can be involved and periosteum can become quite thick up to a few centimeters to show semilunar calcification. These semilunar calcifications developed along the bony cortex are called "struts" (2). This characteristic finding is not found in early stage and is persistent in well-developed old pseudotumors.

The mechanism of pseudotumor of bone is not understood well. It was postulated that recurrent intraosseous hemorrhage into closed spaces prohibits blood reabsorption and promotes bone destruction thus showing its nature to continuously enlarge. Hemorrhage in subperiosteal space also occurs frequently. It was assumed that, in subperiosteal hemorrhage, secondary bony changes occur even in early stage of pseudotumor, because periosteum could produce higher pressure than hematomas in the muscle. Secondary bony changes such as bone necrosis may be caused by blood vessel compression. In this type of pseudotumor, direct erosion by hematoma also affects bone.

Irradiation has been attempted in several reports and it seems effective and some investigators have advocated the use of radiotherapy (3-7). The mechanism of hemophilic pseudotumors to respond to radiation therapy is not known. Brant and Jordan (2) suggested the radiation of 1,000-2,000 cGy lead to an endarteritis in an acute bleeding hematoma. Reinhold (8) suggested that the radiation may directly injure the fine vessels supplying the pseudotumor causing fibrosis, leading to eventual healing. Meyers and Hakami (3) suggested that radiation affects the cellular component of the pseudotumor, primarily fibroblasts, causing fibrosis.

The optimal radiation dose for hemophilic pseudotumor is not defined. We used 900 cGy in this case, reasoning that 900 cGy might not disturb epiphyseal growth. A dose of 900 cGy can be enough to bring about endarteritis or fibroblast growth. Although dose of such range is not thought to cause any changes in epiphysis, we shielded the epiphyseal plate of distal humerus to further decrease the risk of possible bony growth disturbance.

We have followed this patient for up to 72 months and there has been no evidence of recurrence or growth disturbances in his ulna. Authors suggest that radiation therapy can be an effective alternative modality in treating hemophilic pseudotumor especially for the patients

who have lesions in the bone of further growing potential and who have antihemophilic factor antibodies and significant surgical risks.

REFERENCES

1. Gilbert MS. *Characterizing the hemophilic pseudotumor. Ann NY Acad Sci 1975; 40: 311-5.*
2. Brant EE, Jordan HH. *Radiologic aspects of haemophilic pseudotumors in bone. Am J Roentgenol Radium Ther Nucl Med 1972; 115: 525-39.*
3. Meyers L, Hakami N. *Pseudotumor of hemophilia in the orbit: the role of radiotherapy in management. Am J Hematol 1985; 19: 99-104.*
4. Correra A, Buckley J, Roser S, Schreiber A, Syrop S. *Radiotherapy of a pseudotumor in a hemophiliac with factor VIII inhibitor. Am J Pediatr Hematol Oncol 1984; 6: 325-7.*
5. Castaneda VL, Parmley RT, Bozzini M, Feldmeier JJ. *Radiotherapy of pseudotumors of bone in hemophiliacs with circulating inhibitors to factor VIII. Am J Hematol 1991; 36: 55-9.*
6. Hilgartner MW, Arnold WD. *Hemophilic pseudotumor treated with replacement therapy and radiation. Report of a case. J Bone Joint Surg Am 1975; 57: 1145-6.*
7. Chen YF. *Bilateral hemophilic pseudotumors of the calcaneus and cuboid treated by irradiation: case report. J Bone Joint Surg Am 1965; 47: 517-21.*
8. Reinhold HS. *The influence of radiation on blood vessels and circulation IV. Structural changes in blood vessels. Curr Top Radiat Res Q 1974; 10: 58-74.*