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TREATMENT OF HEMOPHILIC PSEUDOTUMOR WITH LOW – DOSE RADIOTHERAPY *

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SUMMARY: Özbek N, Ünsal M, Kara A, Gümrük F, Gürgey A. (Hematology Unit, Department of Pediatrics, and the Department of Radiology, Hacettepe University Faculty of Medicine, Ankara, Turkey). Treatment of hemophilic pseudotumor with low-dose radiotherapy. Turk J Pediatr 1996; 38: 91-94.

Hemophilic pseudotumor is one of the most serious complications of hemophilia and is usually treated with extensive surgery. A new treatment approach is radiotherapy. Patients with long-bone pseudotumors are usually treated with high doses of radiotherapy greater than 1500 cGy. We treated a 13-year-old hemophilic boy who had a pseudotumor of the tibia with low-dose radiotherapy (600 cGy). There was no complication during the two-and-a-half-year follow-up. Improvement of both the clinical and radiological status of the patient was noteworthy. We would like to suggest the use of low-dose radiotherapy in patients with hemophilic pseudotumors.
Key words: hemophilia, pseudotumor, external radiotherapy.

Hemophilic pseudotumor is one of the most serious complications of hemophilia. It was first described by Starker[1] in 1918 in the femur of a 14-year-old boy with hemophilia. It is sometimes confused with various malignant and benign bone diseases. The lower extremities and pelvis are most frequently affected, but almost all sites may be involved[2]. Recurrent hemorrhage into an encapsulated hematoma causes destruction and finally cyst formation in the bone. Increased levels of Factor VIII (F-VIII) inhibitor may render F-VIII supplementation ineffective and surgery impossible[3]. In some cases pseudotumors can become large enough to threaten the patient's life.

Hemophilic pseudotumors are usually resistant to medical treatment. Radical surgical procedures such as amputation are often needed in these cases[4]. However, in recent articles it has been reported that radiotherapy and/or F-VIII supplementation were successfully used in some patients[5]. The present report describe a patient who presented with a pseudotumor in the tibia and was treated with a lower dose of radiotherapy.

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Case Report

A 13-year-old male patient who had been diagnosed with severe hemophilia-A at the age of three months (F-VIII level 1.2 U/dl) was admitted to the hospital with hemarthrosis of the left knee which developed after blunt trauma to that region. Fresh frozen plasma three times per day was transfused for a week to the patient for the treatment of hemarthrosis. Neither pain relief nor reduction in the size of the knee was achieved during the course of treatment. Radiography of the left knee showed a lytic lesion in the tibial tubercle. The continuity of the bone cortex, especially in the anterior part, was disturbed. There were radiolucent cysts with a septal formation in the proximal one-third of the tibia which was diagnosed as a hemophilic pseudotumor (Fig. 1). Factor VIII inhibitor was negative at that time. Since replacement therapy was ineffective, the patient was given a total of 600 cGy of Cobalt 60 photons in four fractions (150 cGy/day) to the left knee for four consecutive days. He was able to walk without pain and the knee swelling was resolved a week after the beginning of radiotherapy. The last physical examination revealed neither deformity nor disturbance of growth. The lytic lesion was significantly diminished with new periostal lamellary bone formation on the radiograms obtained 1.5 and 2.5 years after radiotherapy (Figs. 2 and 3). There was a minimal defect in the tubulation of bone.

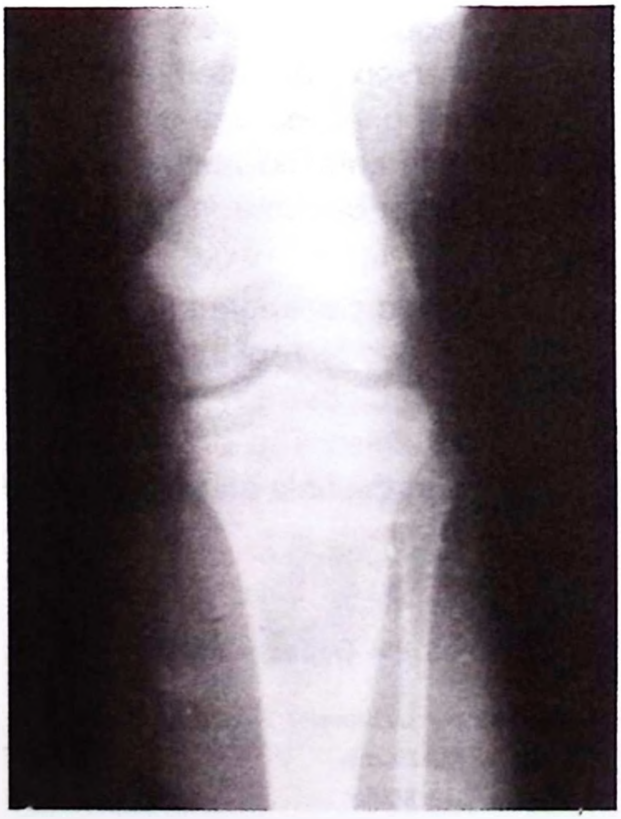

Fig. 1: Posteroanterior view of the left knee before therapy.

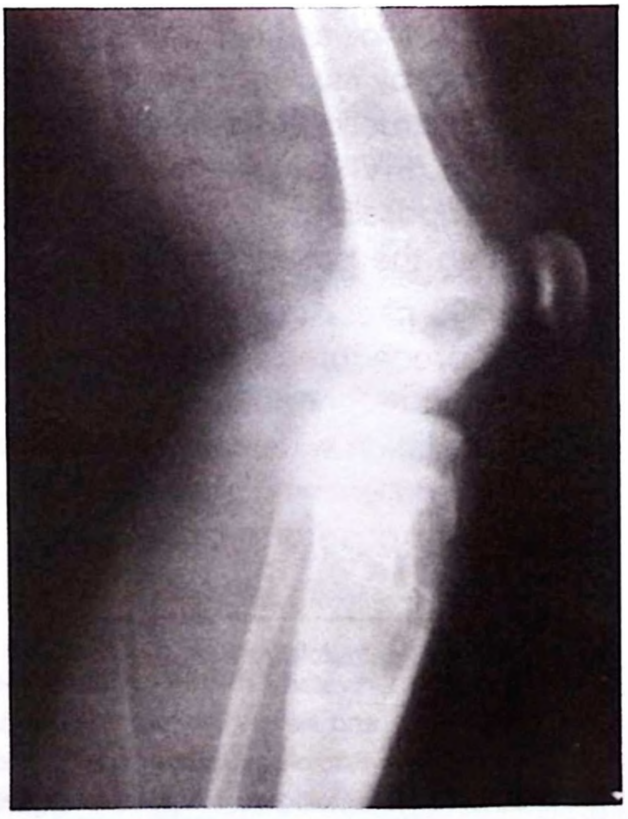

Fig. 2: Lateral view of the left knee one-and-a-half years after radiotherapy.

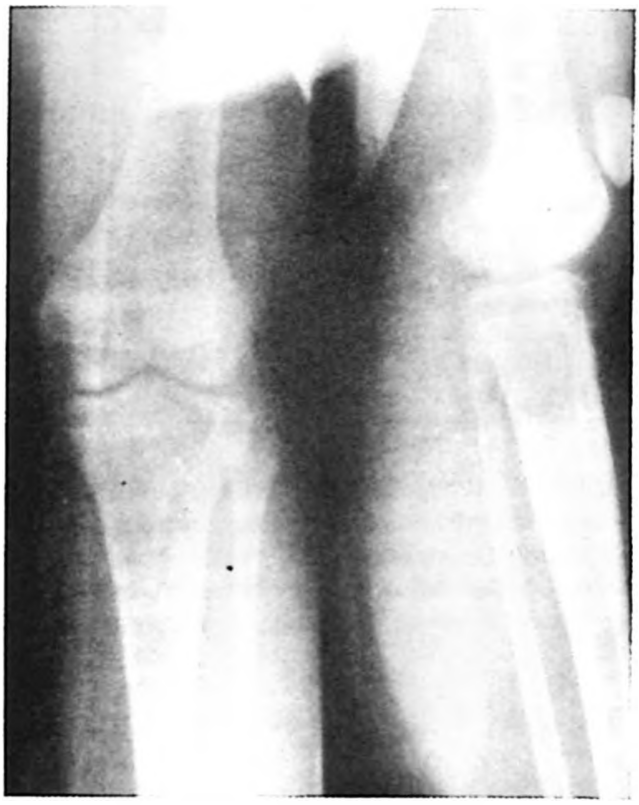

Fig. 3: Lateral and posteroanterior view of the left knee two-and-a-half years after radiotherapy.

Discussion

Replacement therapy has been effective in small-sized pseudotumors. However, large lesions are usually treated with surgical intervention. Bleeding episodes may be impossible to control with replacement therapy in patients with high titers of inhibitors to F-VIII or factor IX. Therefore, some investigators have advocated the use of radiotherapy. Despite the fact that F-VIII inhibitor was negative in our patient, we could not achieve satisfactory results with appropriate replacement therapy. In the literature it has been reported that patients with long-bone pseudotumors were usually treated with high doses of radiotherapy up to 1500 cGy. In a recent article, Castaneda et al.[3] reported good results with a lower dose of radiotherapy in a mandibular pseudotumor, but they used higher doses up to 1500 cGy for long-bone pseudotumors. Our patient was an adolescent boy with accelerating growth that could be affected by high doses of radiotherapy up to 10 Gy. Therefore, we decided to treat our patient with low-dose radiotherapy. As soon as the therapy was completed, the pain was alleviated and the patient was able to walk. It was noteworthy that he did not develop any symptom or hemarthrosis in his left knee within two-and-a-half years following radiotherapy. His last physical examination showed that growth of the tibia was not affected. He is now able to walk with a slight limp.

We would like to suggest the use of a lower dose of radiotherapy in patients with hemophilic pseudotumors.

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