Management of Hemophilic Pseudotumor by Limited Surgery : A Case Report

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A case of infected hemophilic pseudotumor of the femur occurring in an 18 year-old man following an attempted biopsy of the tumor mass was successfully treated by limited exploration and removal of the infected tumor and bone instead of amputation of the limb. The wound later healed with regression of the tumor. No recurrence of the tumor has been observed for two and a half years.

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Hemophilic pseudotumor is a progressive cystic swelling involving muscle, produced by recurrent hemorrhage and accompanied by radiographic evidence of bone involvement, characterized by Fernandez De Valderanna and Mathew[1]. Starker had described the first pseudotumor in 1918[2]. Since then over 60 cases have been described. Gunning estimated an incidence of 1% in severe hemophiliacs[3]. The outcome reported by Marder and Shulman of amputation or death was 58 percent of their reported cases[4]. We are reporting a case of pseudotumor in the femur of an 18 year-old man who had moderately severe Factor VIII insufficiency (hemophilia A) and who was successfully treated by cryoprecipitate and limited surgery.

CASE REPORT

An 18 year-old Thai male, was first admitted to the Medical Department, Siriraj Hospital in September 1977. He presented with a 2 month history of swelling of the right knee which was increasing in size and pain until he was unable to get about. There was no history of any trauma to the knee. He was a member of the family of 8 siblings; 7 males, 1 female and there was a history of abnormal bleeding in 4 out of the male members (Fig. 1). Physical examination revealed a fusiform enlargement of right lower thigh and knee. The mass was rather firm and hard, nonfluctuant but slightly tender with the resulting limitation of knee movement. Roentgenogram of the right femur revealed severe destruction of metaphysis of the

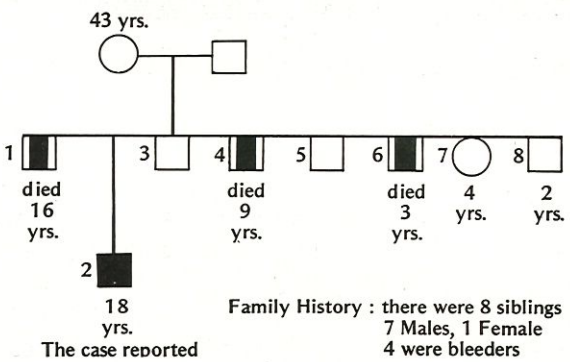

Fig. 1 Pedigree of the patient's family.

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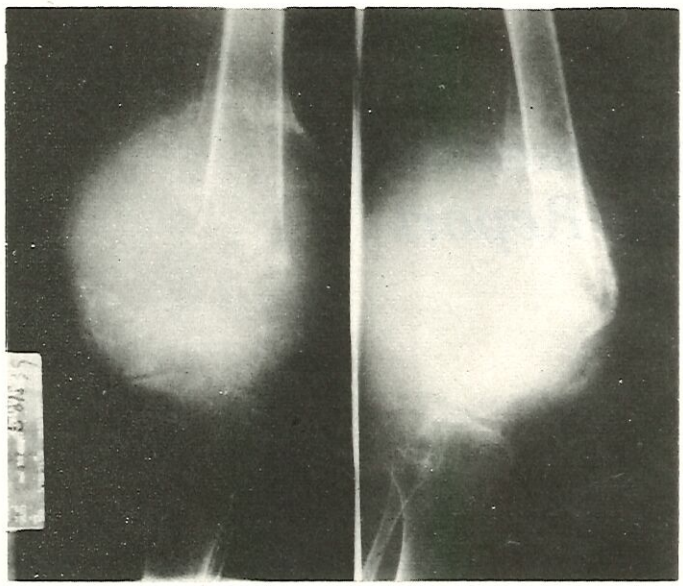

Fig. 2 Roentgenogram of right distal femur showing a soft tissue mass with bony destruction.

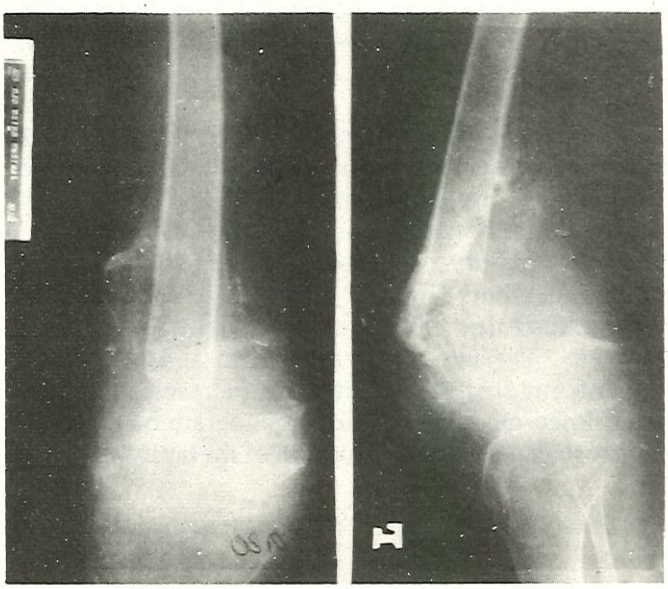

Fig. 3 Roentgenogram of the lesion before surgery.

distal femur. There was also a large soft tissue mass with osteoid formation which was suggestive of osteosarcoma of distal femur (Fig. 2). Diagnostic aspiration was performed under protection of cryoprecipitate but no pathology was obtained. Further diagnostic procedure was carried out by open biopsy and this showed no malignancy. Unfortunately, the biopsy site became infected and there was persistent drainage. It was treated by cryopricipitate and routine daily local wound dressing but no evidence of improvement was observed. On December 29, 1977 the patient underwent surgical exploration of the infected biopsy site through bilateral long vertical skin incisions of both sides of the right lower thigh. Infected granulation and osteoid-like material were curetted. The cavity between the bony parts was firmly packed with wet gauze. Local wound care with intermittent cryoprecipitate administration was provided for one month. The wound gradually covered with granulation tissue and closed without significant morbidity.

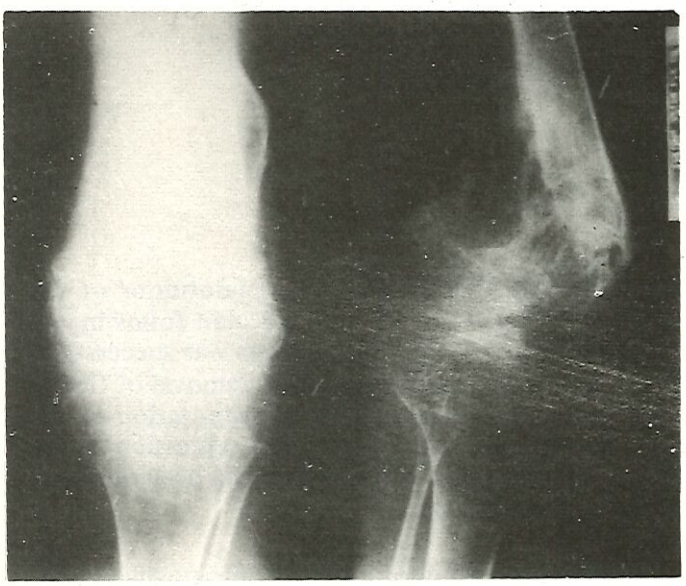

Fig. 4 Two months after surgery.

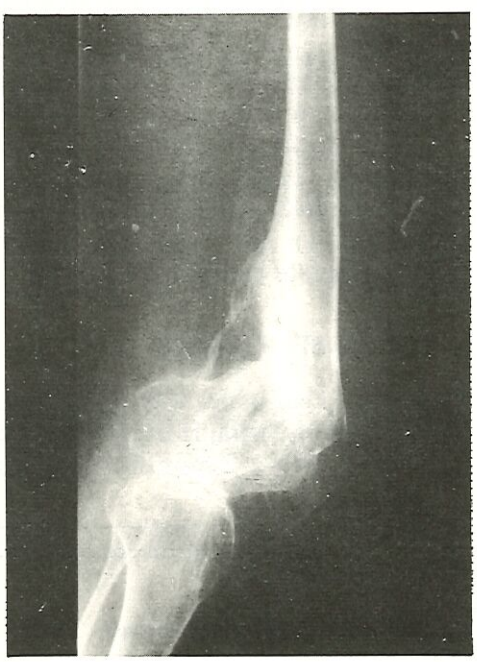

Fig. 5 Two years after surgery showing no evidence of recurrence.

Roentgenogram of the right distal femur for comparison before and after limited surgery at 2 months, and 2 years showed significant increasing calcified bone (Fig. 3,4,5) but no enlargement of soft tissue was noted. During the 2½ years of his convalescence, hematoma occurred underneath the previous scar on the medial side of right knee and required to be opened for evacuation of the clot. The patient has since returned to his work as a barber. His course has been quite favorable with considerable regression of the lesion.

DISCUSSION

Pseudotumor is an unusual complication of hemophilia, as mentioned earlier about 1 per cent of severe or moderate hemophilia A or B developed pseudotumor[3] and only some sixty cases were reported in the world literatures[4]. There is some difficulty in making a diagnosis particularly in those who have a solitary bone cyst or aneurysmal bone cyst. An early developed pseudotumor can be effectively treated by early and adequate substitution therapy[5].

Gilbert has classified Hemophilic Pseudotumor into 2 types; proximal and distal, the proximal type being situated in the pelvis and femur and the distal type localizing mainly in the hands and feet[6]. The former type appears mainly in adults and the latter type appears only in children and carries a better prognosis. Aspiration of pseudotumor usually results in the formation of a fistula, followed by infection and death from sepsis. Those who have had experience with this procedure have suggested that aspiration of pseudotumor is contraindicated[7]. Early pseudotumor could be theoretically prevented by drainage of the hematoma before pressure necrosis develops. However, this has often resulted in complications such as hemorrhage and infection. If pressure necrosis in a long bone ensues, with increasing swelling and pain, early amputation under strict hematological control may offer the best outcome. The indications for surgical intervention in pseudotumor in the past have been to exclude neoplasm and osteomyelitis, to prevent fatal rupture and to alleviate pain[8]. Conservative treatment has been successfully used in many cases, especially those in the small bones or distal type where there was resolution of the mass[6]. No case of a large pseudotumor at the ilium or femur has been proved to be successfully treated by the conservative method at the long term follow up. Radiation has also been successful when long-term adequate replacement therapy was unsuccessful, apparently because of the induced fibrosis[9]. Surgical intervention is indicated for a large pseudotumor. Complete removal of the cyst should be attempted and may be re-inforced by grafts, internal fixation or external immobilization. Amputation should be reserved for those cases in which limb is rendered useless by massive infection or extensive tissue destruction or neurovascular compromise[9]. In cases of local infection of pseudotumor with remaining function of the limb, the infected tumor part can be removed through the wide and adequate opening skin incisions either by curettage or ronguer. Adequate and careful local care of the wound can lead to successful healing without the sacrifice of the limb.

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