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CHRONIC HEMOPHILIC ARTHROPATHY
Case Report

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SUMMARY :

Introduction of factor VIII and IX concentrates in the early 1960's resulted in a significant change in the life expectancy of hemophiliac patients. This fact however, also brought a parallel rise in the incidence of chronic hemophilic arthropathy. In this paper, a 21 years old male patient, with a previous diagnosis of Hemophilia A, complaining of left shoulder and right elbow pain was presented and the results in the functional outcome after application of gentle stretching, progressive resistive and range of motion (ROM) exercises were reported.

Key Words: Hemophilia, arthropathy, physical therapy

ÖZET

KRONİK HEMOFİLİK ARTROPATİ
Olgu Sunumu

1960'lı yılların başlarında Faktör VIII ve IX preparatlarının kullanıma sunulması hemofilik hastaların yaşam beklentilerinde önemli değişimlere neden olmuştur. Ancak, bu gerçekle birlikte kronik hemofilik artropati insidansında da paralel bir artış gözlenmiştir. Bu yazıda sol omuz ve sağ dirsek ağrısından yakınan ve daha önceden hemofili A tanısı ile izlenmekte olan 21 yaşında bir erkek hasta sunulmuş ve hafif germe, progresif rezistif ve ROM (hareket açıklığı) egzersizleri uygulamasından sonra fonksiyonel açıdan elde edilen sonuçlar bildirilmiştir.

Anahtar Kelimeler: hemofili, artropati, fizik tedavi

INTRODUCTION

"Hemophilia" is a term used to designate a group of hereditary bleeding disorders characterized by either factor VIII or IX deficiency, representing Hemophilia A and B respectively. Both forms of the disease have identical clinical signs and laboratory abnormalities and both share the common genetic mode of transmission (1, 2, 3).

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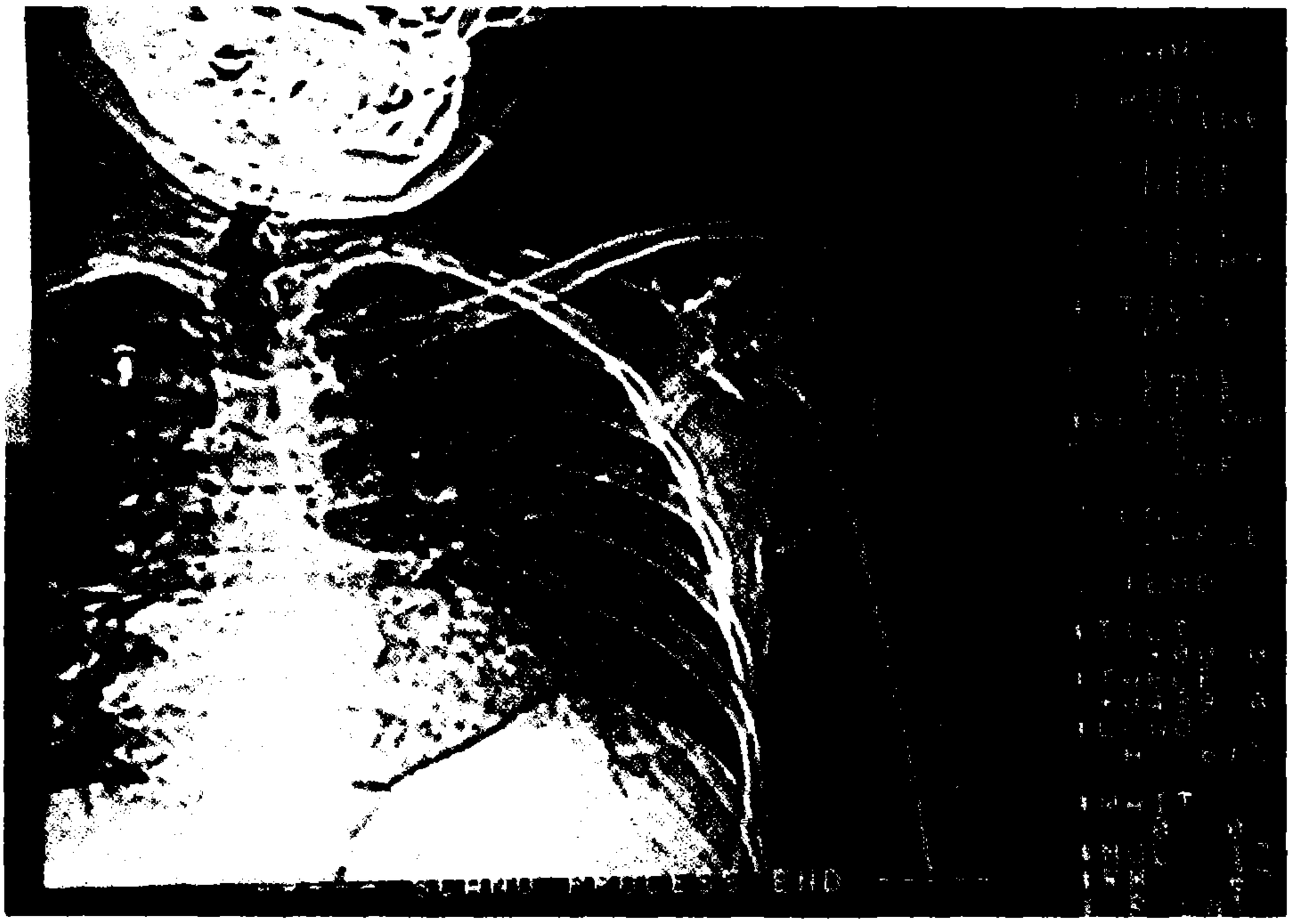

Figure I. CT image (left shoulder).

All the hemophiliacs are male due to X-linked recessive mode of transmission and about 80% of all hemophiliacs have hemophilia A (1, 2, 3, 4). The disease in both forms is characterized by recurrent bleeding episodes in many systems including the gastrointestinal tract, genitourinary tract and the musculoskeletal system. When the joints are involved, the initial lesion consists of synovial or subsynovial hemorrhage usually but not always following trauma. After repeated hemorrhage the synovial lining cells show hyperplasia and the resultant synovial fibrosis leads to the rigidity of the joint capsule. These changes together with the damage occurred in the cartilage and bone ultimately lead to limited range of motion in the involved joint. The joints most commonly effected by bleeding are the knees, ankles and elbows; shoulders, hips, wrists, fingers and toes are less frequently involved (1, 5).

A gradual failure of joint function ensues after repeated hemorrhage episodes. This picture is followed by muscle atrophy and various deformities, the most common of which are flexion contractures of the elbows and knees (1, 2, 3, 4, 5).

Figure II. CT image (left shoulder).

CASE REPORT

Patient F. B. 21 years old, male, admitted to our department with two epicrisis from two different centers with the diagnosis of Hemophilia A. He had received factor VIII concentrates for many times during his life. He had had many episodes of arthralgias and given Ibuprofen or Paracetamol but never had physical therapy before. He was admitted to our department because of pain and increasing limitation of motion in his left shoulder and right elbow interfering with some of his daily living activities.

The initial examination revealed an asthenic young male with a slight growth retardation and poor posture. His left shoulder girdle was atrophic and almost all the motions on this side either active or passive were signifisantly limited. There was a flexion contracture in his right elbow.

ROM degrees in the initial examination were:

Left Shoulder	Active	Passive
Flexion	70°	80°
Extension	25°	35°
Abduction	80°	90°
Adduction	0°	0°
Ext. Rotation	5°	15°
Int. Rotation	45°	45°
Right Elbow		
Flexion	125° (85° range)	125° (85° range)
Extension	0° limitation	40° limitation
Pronation	65°	
Supination	40°	

Laboratory examination revealed :

Hb: 16 g/dl	ESR: 2/9/20
WBC: 6000	RF: (–)
RBC: 4.500.000	CRP: (–)
PLT: 250.000	ASO: 200 TU

PTT was prolonged and Bleeding Time was in normal limits. Factor VIII level was less than 1%.

Plain radiograms of the left shoulder showed irregular obliteration of the joint space, huge spur formation and sclerosis; CT confirmed these findings and also showed cortex atrophy in the humeral head. Similar radiologic views were obtained from the right elbow (Figures I, II, III, IV). Scintigraphic examination with Tc 99 m-MDP revealed significant high uptake in the left shoulder, right knee, right ankle, right elbow and right humoral head. Uptake was also increased, although less significant, in the left ankle and in the first toe of the left foot (Figure V).

After examination of these results and repeated hematological consultation, the patient was diagnosed as chronic hemophilic arthropathy and a program consisting of gentle stretching, progressive resistive and ROM exercises were started. The patient was also given Ibuprofen 400 mg/day for pain control. After 30 sessions, the atrophy in the left shoulder girdle improved and ROM degrees were increased significantly. The patient reported neither pain nor striking disability in daily living activities at the end of 30 sessions.

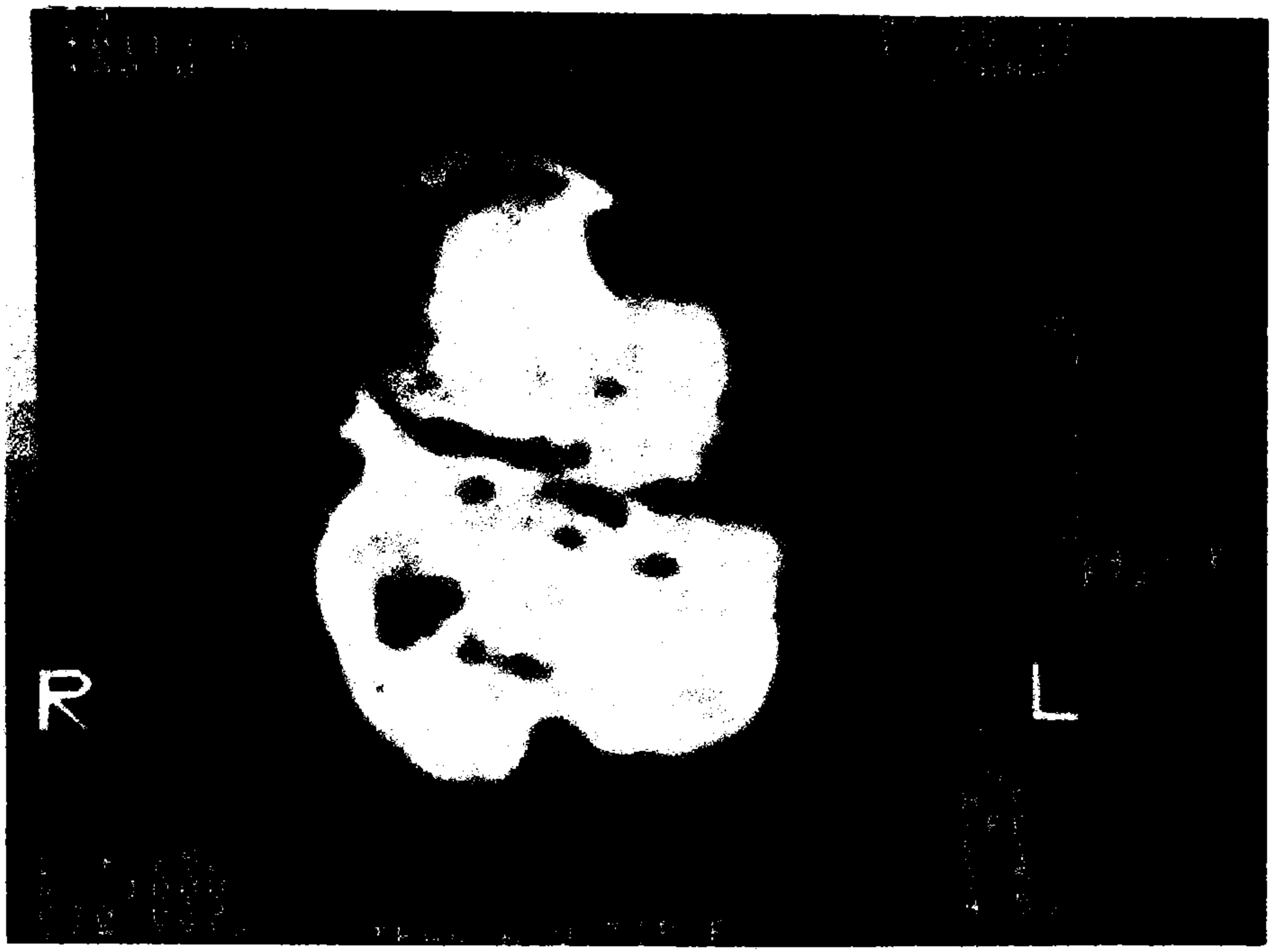

Figure III. CT image (left shoulder).

ROM degrees following exercises were

Left Shoulder	Active	Passive
Flexion	90°	140°
Extension	30°	45°
Abduction	90°	110°
Adduction	0°	0°
Ext. Rotation	20°	30°
Int. Rotation	55°	55°
Right Elbow		
Flexion	125° (105° range)	125° (105° range)
Extension	20° limitation	20° limitation
Pronation	90°	
Supination	40°	

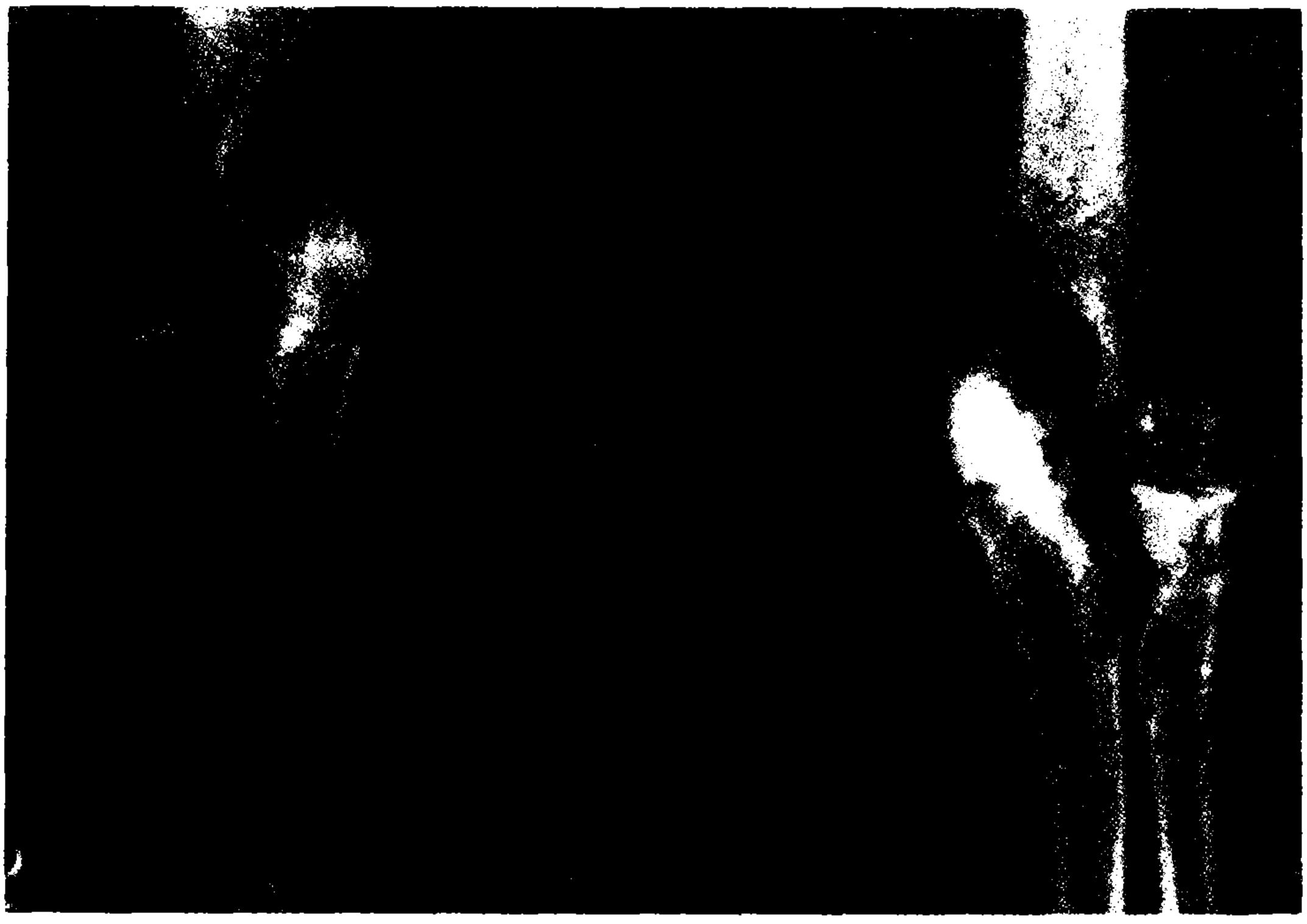

Figure IV. Plain radiograms (right elbow).

DISCUSSION

The incidence of Hemophilia has been reported to be 1 in 10.000 males (3, 4). Hemarthrosis constitutes the most common complication of the disease and accompanies the clinical picture in approximately 75-90% of cases (1, 4). As it was mentioned before, the most commonly effected joints by bleeding are knees, ankles and elbows; shoulders, hips, wrists, fingers and toes are less frequently involved (1, 2, 3, 4, 5). The scintigraphic image of the patient reported here revealed high uptake in the left shoulder, right knee, right ankle, right elbow, right femoral head, left ankle and the first toe of the left foot. The patient reported that he had received Factor VIII concentrates many times throughout his life, had not any surgical operation and had never been consulted by a physiatrist.

Our main concern in this report will be the striking improvement we have observed in range of motion degrees and in daily living activities of the patient after 30 sessions of an exercise program. Management of chronic hemophilic artrophathy includes aggressive factor VIII or IX replacement, surgical procedures and physical therapy measures involving strengthening and ROM exercises for joint stability and muscle building or stretching exercises and night splints to correct the flexion

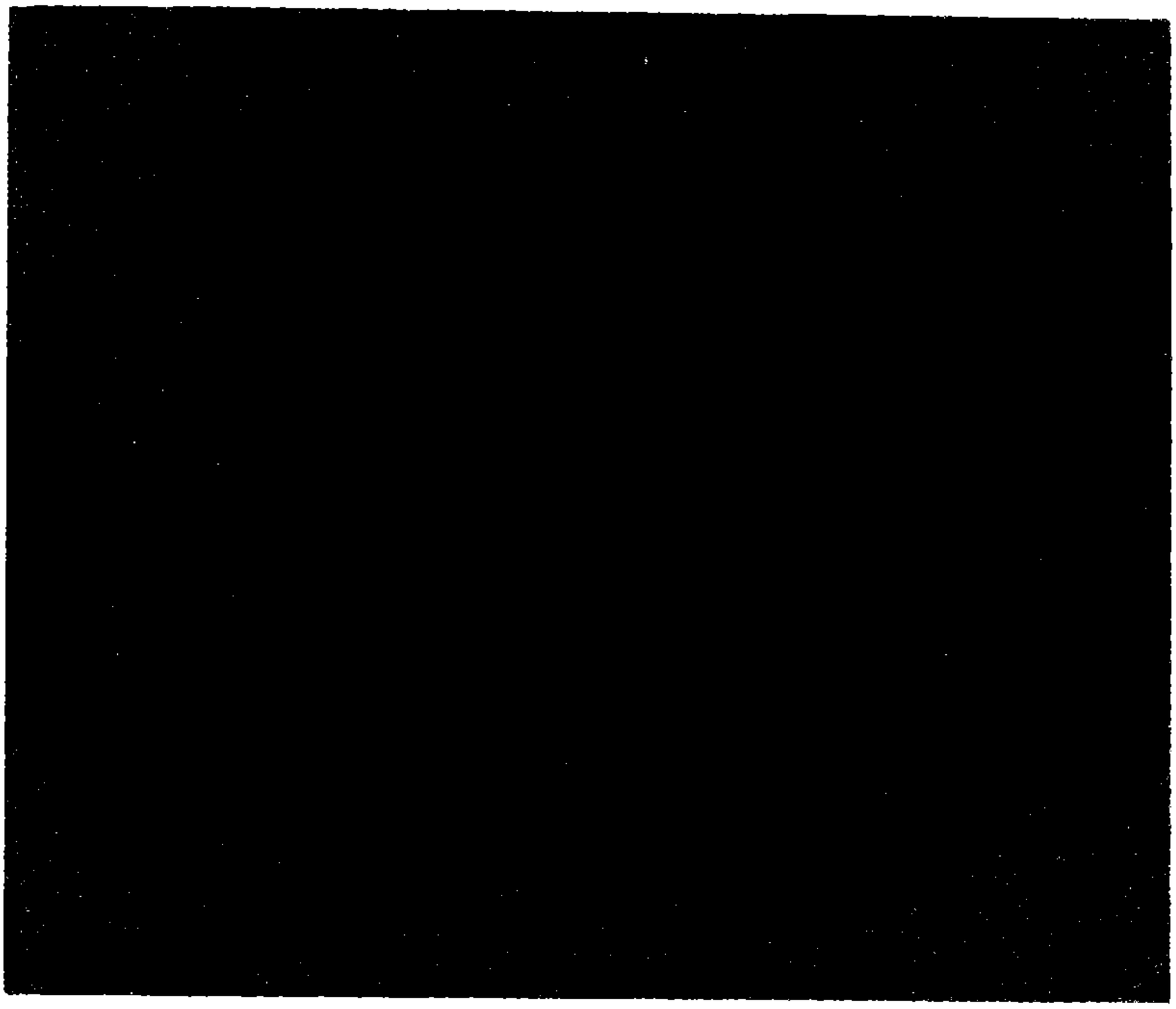

Figure V. Scintigraphic image.

contractures (1, 6). In our case after application of ROM and progressive resistive exercises to the involved joints and gentle stretching to correct the flexion contracture in the right elbow, the patient reported a striking improvement in his daily living activities.

Discussion of the management in chronic hemophilic arthropathy was beyond our scope instead our main concern was to underline the fact that treatment in Hemophilia should be multidisciplinary. During the course of each evaluation, the hemophiliac should be examined by a hematologist, an orthopedic surgeon and a physiatrist and also should be admitted to an oral surgeon, physical therapist and a genetic counselor (1).

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