

Retrospective Study of 14 cases of pseudotumours in hemophilia – From a Comprehensive Hemophilia Care Centre of Western India

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INTRODUCTION

Hemophilic pseudotumour is a rare complication occurring in 1–2% of patients with severe hemophilia. Results from repetitive bleeding resulting in an encapsulated mass of clotted blood and necrosed tissue. The radiographic findings of a soft tissue mass with areas of calcification and adjacent bone destruction in a patient with hemophilia is usually sufficient to make the diagnosis of a pseudotumour.^{1,2,3,4} The management of these is a clinical challenge, requires experience and it should be managed in collaboration of hematologists with surgeons, orthopedicians, laboratory personnel and physiotherapist. Here we report our clinical experience in the management of 14 cases of Pseudotumour in hemophilia patients from a Comprehensive Hemophilia Care Center (CHCC) treated over the period of last 10 yrs.

MATERIALS AND METHODS

Retrospective analysis of management of Pseudotumours, amount of factor used and complications. From 2004 to 2014, our CHCC has treated 14 Pseudotumours in 12 PWH with age from 5 - 67 yrs. Of these 7 had hemophilia A, 3 hemophilia B 2 hemophilia A with inhibitors.

RESULTS

Among 14 patients 11 were osseous, involving lower limb in 9 (foot -2, femur - 2, tibia - 3, thigh-1,) upper limb in one and one in mandible. Of the remaining one in scalp, one over medial malleolus and two had abdominal pseudotumour. They were symptomatic from 3 m - 4 yrs. Patients were managed either conservatively, surgically or with radiotherapy, depending on the condition of the patient, inhibitor status and availability of clotting factor concentrate.

CLINICAL PRESENTATIONS

Serial no	Factor level	Age (Yrs)	Inhibitor status	Site of pseudotumour	Duration of symptoms (Yrs)
1	VIII<1%	5	Negative	Over R Medial malleolus	¼
2	VIII<1%	10	Negative	Scalp	1
3	VIII<1%	18	Positive	L femur	2
4	VIII<1%	26	Negative	Intra abdominal	2
5	VIII<1%	27	Negative	R Tibia	1
6	VIII<1%	38	Positive	R Femur	4
7	VIII<1%	67	Negative	L Tibia	3
8	IX<1%	16	Negative	L thumb	1
9	IX<1%	19	Negative	L Foot	½
10	IX<1%	45	Negative	R thigh	3
11	VIII<1%	38	Negative	Retroperitoneal	3
12	VIII<1%	23	Negative	Great toe	1/2
13	VIII<1%	24	Negative	L tibia	2
14	VIII<1%	7	Negative	L mandible	1

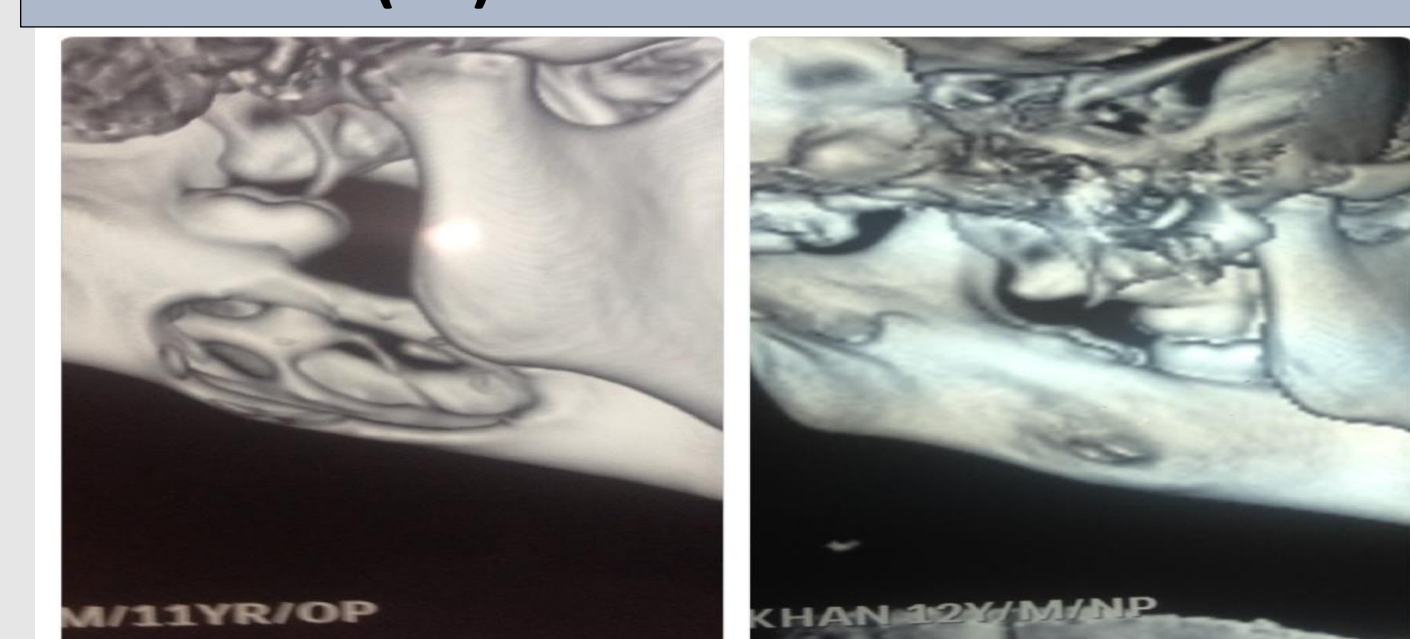
MANAGEMENT

Serial no	Weight of pt (KG)	Site of pseudotumour	Type of management/ Surgery	Total IU of AHF	Outcome
1	15	Over R Medial malleolus	Conservative	5500	Healed
2	40	Scalp	Evacuation and repair	25,000	Healed
3	45	L femur	Above knee Amputation	FEIBA 14500 aVII – 2o mg	Improved and able to walk
4	55	Intra abdominal	Excision	29,000	Recurrence and died
5	35	R Tibia	Excision and repair	22,000	Improved and able to walk
6	60	R Femur	Radiotherapy	-	Died
7	50	L Tibia	Conservative	5000	Lost to follow up
8	30	L thumb	Thumb amputation	14000	Healed
9	32	L Foot	Toe amputation and excision	20000	Healed
10	65	R thigh	Excision and repair	18,000	Healed
11	60	Retroperitoneal	Excision	35,000	Died after 1 yr
12	55	Great toe	Great toe amputation	13,000	Healed
13	45	L tibia	Excision and repair	14000	Healed
14	20	L mandible	Conservative with factor prophylaxis	12000	Healed

Pseudotumour of the left thumb X- ray of the pseudotumour and post surgery of patient no 8



CT scan of mandible Pre and post treatment (14)



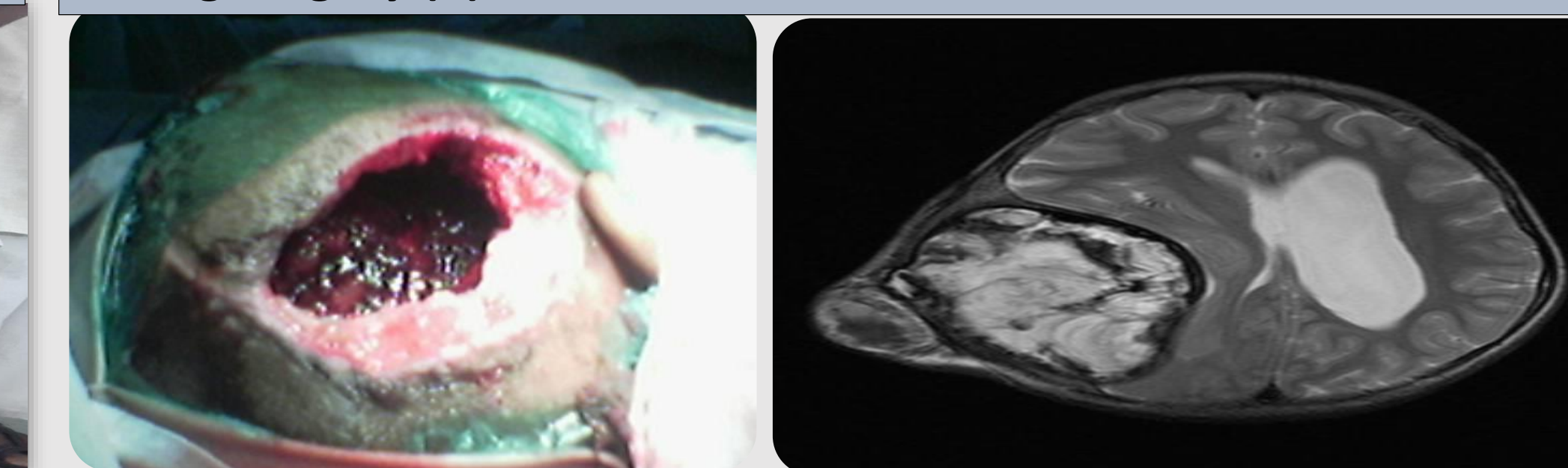
Pseudotumour of the left foot pre and post surgery (9)



Thigh Pseudo tumor excision MRI and clinical picture pre and post surgery (10)



CT scan of left parietal Interdiploic pseudotumour and picture during surgery (2)



OUTCOME

One of the pt with osseous pseudotumour of the lower end of femur with inhibitor of 128 BU was given local radiotherapy. The remaining 10 patients underwent surgery either excision repair or amputation of the involved site and 3 were managed conservatively. Average CFC with bony pseudotumour was 390 U/kg and it was 450U/kg in patients with intraabdominal pseudotumour. Among 3 patients died, had large pseudotumour and presented late to us with systemic complications. All other 10 patients are doing well.

CONCLUSION

The pseudotumours can remain asymptomatic for a long period of time without signs of growth. Timely suspicion of even minor musculoskeletal bleeding and appropriate factor replacement can prevent significant morbidity and mortality associated with this. The present case series clearly shows hemophilic pseudotumours are rare but serious complication of bleeding into musculoskeletal system.

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