SCIENTIFIC ARTICLE

Management of Hemophilic Cysts and Pseudotumors of the Hand in Bleeding Disorders: A Case Series

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Purpose Hemophilic cysts and pseudotumors (HCPTs) of the hand are rare and are secondary to bleeding disorders such as hemophilia A and B. This is a report of our experience in the management of this rare condition.

Patients and Methods Seven male patients with hemophilia A presenting with progressive swelling of the hand were treated between 2004 and 2013 at a tertiary referral hospital. All patients had clotting factor replacement based on our previously reported protocol. The age of the patients ranged from 3 to 49 years (median age, 19 years).

Results Four patients had soft tissue hemophilic cysts and 3 had bony hemophilic pseudotumours. Two patients had traumatic pseudoaneurysm of the ulnar artery in addition to the cysts. The soft tissue cysts required surgical excision in 3 patients under factor cover as per the protocol. The bony lesions were initially managed nonsurgically by factor replacement, but 2 patients failed to respond and required amputation of the fingers. The ulnar artery aneurysm was excised and artery ligated in 1 patient and the artery was vein grafted owing to poor hand perfusion in 1.

Conclusions Based on our observations in the management of HCPTs of the hand and the existing literature, we conclude that the soft tissue cysts require surgical excision along with factor replacement and distal bony lesions smaller than 3 cm respond to factor replacement. Larger bony lesions require surgical treatment. Treatment of hemophilic cysts and pseudotumours should be undertaken only in centers with a major hematology backup. *(J Hand Surg Am. 2017;■(■):1.e1-e9. Copyright © 2017 by the American Society for Surgery of the Hand. All rights reserved.)*

Type of study/level of evidence Therapeutic V.

Key words Hemophilia, bleeding disorders, hemophilic pseudotumors, hand pseudotumors, hemophilic cysts of hand.

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HEMOPHILIC CYSTS AND pseudotumors (HCPTs) are uncommon manifestations of bleeding disorders such as hemophilia A (Factor VIII deficiency), hemophilia B (Factor IX deficiency), and von Willebrand disease, with a reported incidence among these patients of 1.14%.[1] These have only rarely been reported in the hand.[2,3] Hemophilic pseudotumor was first described in 1918 by Starker as an encapsulated, slowly expanding hematoma in patients with severe coagulation disorders. The

TABLE 1. Details of Patients Including Age, Sex, Clinical Presentation, and Follow-Up

Patient	Age (y)/ Sex	Duration	Clinical Presentation	Radiology	Size (cm)	Type	Treatment	Healed (wk)	TAM (° at 12 mo)	DASH (at 12 mo)	Additional Features
A	19/M	6 mo	2 swellings in left palm, infected (Figs. 1–6)	NBI	5	HC	Excision of cyst Excision of pseudoaneurysm	2	270	97.5	Thrombosed ulnar artery pseudoaneurysm, ulnar and median nerve symptoms
B	49/M	3 mo	Large swelling i P3	Lysis P3–LF	3	PT	LF amputation owing to persistent infection and enlarging pseudotumour	2	–	97.5	Right thigh hematoma 2 y back
C	18/M	4 y	Left LF multiple bony swellings (Figs. 7–10)	Left PT, P1–LF, V MC; P1, P2–IF	2.5	PT	Nonsurgical, with factor replacement	15	23 0	97.5	Recurrent bleeds 3–4/y Ankylosed knees Nonambulatory
D	3/M	3 wk	Left palm swelling, infected, with discharge	NBI	1	HC	Nonsurgical, with factor replacement	4	260	97.5	–
E	35	5 d	Right palm swelling, cystic, about 6 cm in size	NBI	6	HC	Excision of cyst, local rotation flap	2	250	97.5	Preexisting small lesion with sudden increase in size -
F	40/M	3 mo	Left palm swelling, solid to cystic, about 3 cm in size, with ulnar artery aneurysm	NBI	3	HC	Excision of cyst and aneurysm, vein graft for artery	2	250	97.5	HCV + ulnar artery pseudoaneurysm, clawing and ulnar nerve paresthesia
G	14/M	2 y	Left little finger swelling, infected P2	Lysis of P2–LF	3.5	PT	LF amputation owing to persistent infection and enlarging pseudotumour	2	–	98.33	–

DASH, Disabilities of the Arm, Shoulder, and Hand; HC, hemophilic cyst; HCV, hepatitis C virus; IF, index finger; LF, little finger; MC, metacarpal; NBI, no bony involvement; P1, proximal phalanx; P2, middle proximal; P3, distal phalanx; PT, pseudotumor; TAM, total active motion; V MC, fifth metacarpal.