SURGICAL TREATMENT OF THE HEMOPHILIC PSEUDOTUMOR: A SINGLE CENTRE EXPERIENCE

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Type of Haemophilia	Virology	Year of Surgery	Age at surgey	Localization	Dimension (cm)	Symptoms	Surgery	Conservative	Remarks	Outcome
A <1%	not available	1967	40	thigh R	not available	not available	not available	not available	not available	no recurrence
A <1%	Hep C	1984	43	iliac L	20x11.5	growing mass over 1.5 years, no pain	resection	yes, not resolved	infection, fistula after 12y and 17y	no recurrence
A <1%	Hep C, B	2000	40	iliac L	17,5x10,1	growing mass over 1 year, pain	resection, cement, pin	yes, not resolved		no recurrence
		2007	48	thigh R	9,5x7,2	growing mass over 7 months, no pain	resection	yes, not resolved	died, liver failure after 3 years	no recurrence
A <1% inhibitor	Hep C	2000	41	ellbow L	3,2x2	paraesthesia over 3 years, pain over 6 months	resection	yes, not resolved	Inhibitor died, liver failure after 7.5 years	no recurrence
A <1%	Hep C, B	2004	61	iliac R	22,5x15	growing mass over 1.5 years, no pain	resection, cement, pin	yes, not resolved	infection after 4 weeks died, liver failure after 5 years	no recurrence
A <1%	Hep C, B, HIV	2000	48	tibia R	9,6x3,3	pain over 4 months	curretage, cement, LCDCP titanium	yes, not resolved	infection, fistula after 5 months	no recurrence

Table 1: Patients´ characteristics

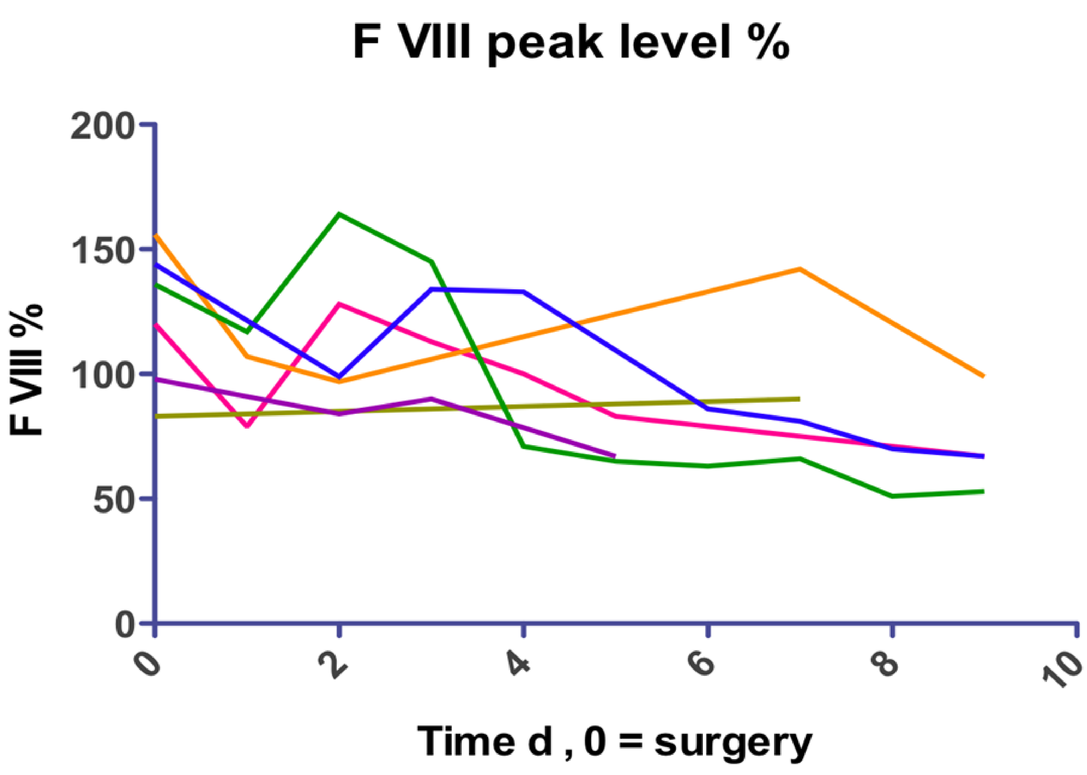
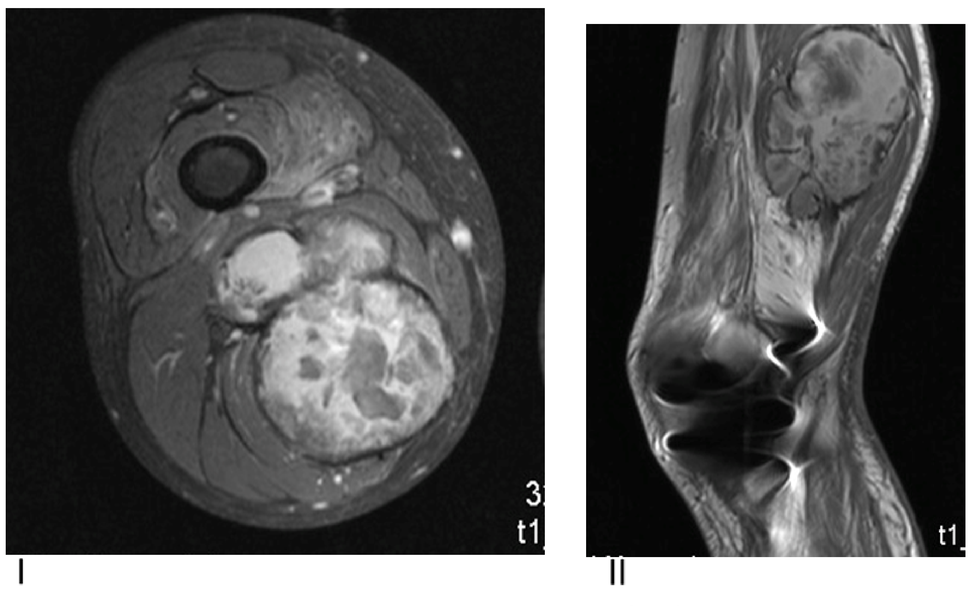
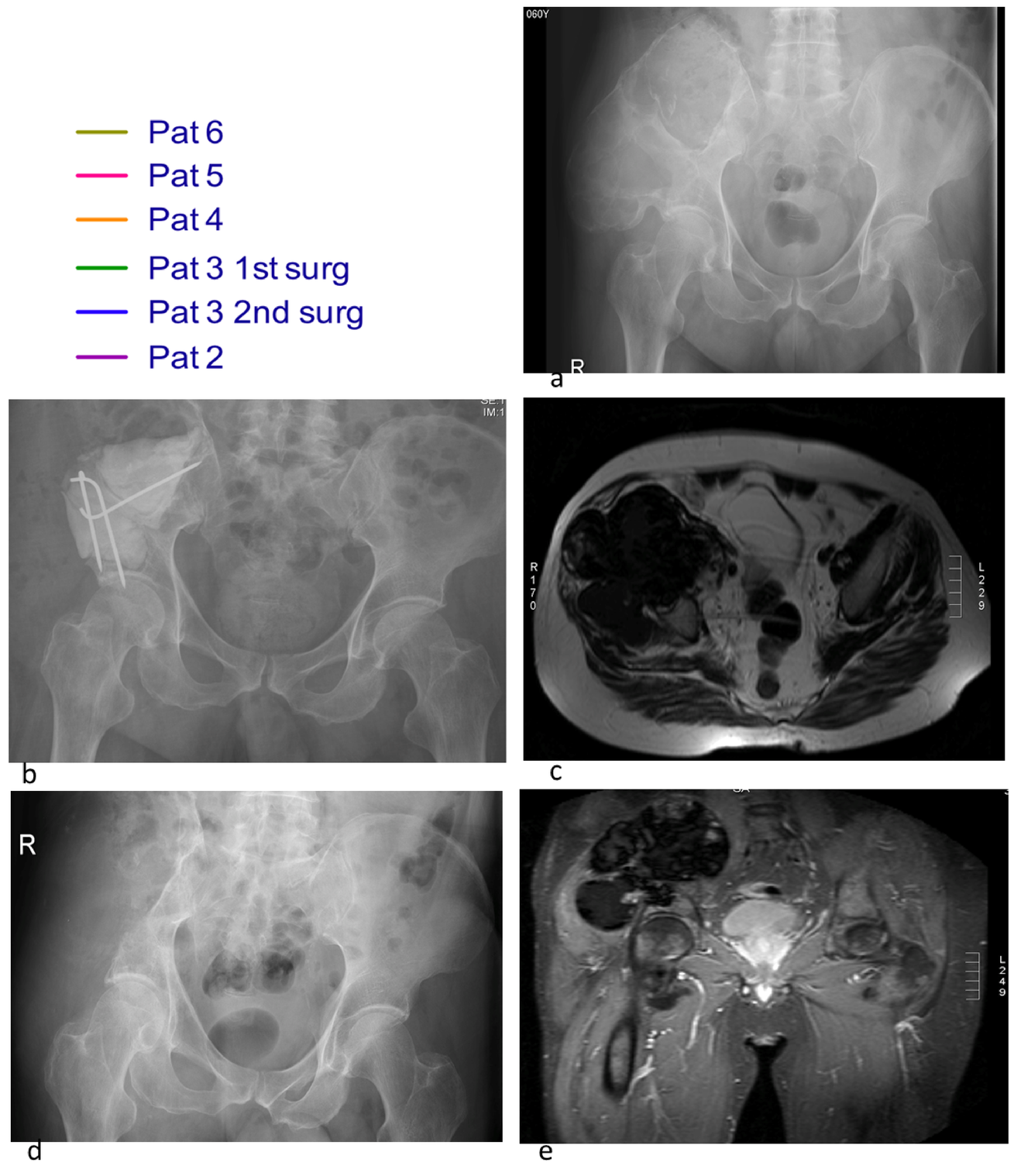


Figure 1: peak F VIII (%) during surgey



Pat 3: MRI right thigh before surgery (I,II)



Pat 5: MRI pelvis before surgery (c,e), x-ray before (a), 4 weks after surgery (b) and after removal bone cement and pins (d)

Purpose: Hemophilic pseudotumor was defined by Fernandez de Valderrama and Matthews as a progressive cystic swelling involving muscle, produced by recurrent hemorrhage and accompanied by roentgenographic evidence of bone involvement. The most common site for the hemophilic pseudotumor is the proximal skleleton around the femur and pelvis.

Methods: We retrospectively reviewed all clinical histories of 87 patients with bleeding disorders treated between 1967 to 2011 because of musculoskeletal affection due to congenital bleeding disorders. We identified 6 patients with a hemophilic pseudotumor who were treated at our department.

Results: The mean age at surgery was 45.9 (range: 40-61) years. The iliac bone was affected in 3 patients (one right, two left), the right tibia (distal diaphysis) in one, the right thigh in two and the right ulna (proximal part) in one patient. One patient had two pseudotumors. The perioperative course was easily controllable with adequate factor VIII substitution. At the latest follow up after 8.4 (4-24) years, normal healing with no recurrence was observed.

Discussion: The hemophilic pseudotumor is a rare but severe complication of hereditary bleeding disorders. In the international literature the resection and postoperative course are described as challenging and difficult and as requiring detailed preoperative planning. Operation done in specialised centres with close cooperation between surgeons and hematologists is advisable.

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