Surgical treatment for pelvic haemophilic pseudotumour: a retrospective analysis of 21 cases

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Summary

Background Due to the rarity of pelvic haemophilic pseudotumour (PHPT) and demanding surgical technique for PHPT excision, no study reports the mid-term follow-up outcomes of surgical treatment of PHPT in a relatively large cohort. PHPT with varying degrees of bony pelvic involvement and infection status necessitates different operative procedures, yet there is currently no classification system for PHPT based on surgical practice.

Methods The study was conducted between June 25, 2004 and July 18, 2023, in Peking Union Medical College Hospital and Nanfang Hospital in China. We performed a retrospective analysis involving 21 patients with 24 PHPTs with a mean follow-up period of 7.1 years. The demographic information, PHPT characteristics, surgical data, and perioperative complications were analysed.

Findings 21 consecutive male patients with 24 PHPTs (21 primary PHPTs and three recurrent PHPTs) that underwent surgical treatment were involved in the study. A classification system including four subtypes was introduced as (I) PHPT confined to soft tissue; (II) PHPT involving bony pelvic without pelvic discontinuity; (III) PHPT causing pelvic discontinuity; (IV) Infectious PHPT. Of the 24 PHPTs, 11 (45.8%) were identified as Type I, five (20.8%) as Type II, three (12.5%) as Type III, and five (20.8%) as Type IV. At the time of surgery, the patients had a mean age of 37.0 ± 9.5 years (Range, 24–52 years). The mean maximum diameter of PHPTs upon surgery was 17.0 ± 7.7 cm (Range, 4.3–40.0 cm). The mean surgical duration was 192 ± 77 min (Range, 60–330 min) and the median intraoperative blood loss was 400 mL (IQR, 225–950 mL, Range, 100–3000 mL). One patient (4.8%) underwent intraoperative cardiopulmonary arrest and expired the following week. Four PHPTs (16.7%) presented postoperative wound infections and poor wound healing. During the follow-up period, five PHPTs (20.8%) experienced pseudotumour recurrence.

Interpretation Our findings suggest that surgical treatment for PHPTs is feasible and relatively safe. Symptomatic and progressive PHPTs should undergo surgical intervention as early as possible to minimise the surgical risks. Intraoperative use of abundant gelatin sponges in PHPT excision draws attention to severe embolism complications.

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Keywords: Pelvic haemophilic pseudotumour; Haemophilia; Surgical treatment; Perioperative management of haemophilia

Introduction

Haemophilic pseudotumour (HPT) refers to a progressively enlarging encapsulated cystic mass caused by recurrent haemorrhaging in extra-articular musculoskeletal system that occurs in 1–2% of patients with haemophilia.[1–3] As a rare but serious complication of haemophilia, HPT can lead

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Translation: For the Chinese translation of the abstract see Supplementary materials section.

Research in context

Evidence before this study

We searched PubMed for relevant work published before Jan 20, 2024, with no language restrictions, using the terms ("Haemophilic pseudotumour" [All Fields] OR "Haemophilic cyst" [All Fields]) AND ("Surgery" [All Fields] OR "Surgical" [All Fields] OR "Operation" [All Fields] OR "Operative" [All Fields]), which yielded 73 results. The majority of resulting studies are case reports. Despite several case series, these often include PHPT analysed together with HPT at other sites, and no study includes more than five surgery-treated PHPTs. Furthermore, within PHPTs, obvious heterogeneity is present and different operative procedures are warranted, yet there is currently no classification system for PHPT. We aim to report the mid-term (7.1 years) outcomes of the largest PHPT cohort (21 patients and 24 PHPTs) so far and introduce a novel PHPT classification scheme.

Added value of this study

In this bicentric retrospective study, we reported the mid-term outcomes (7.1 years) of the largest PHPT cohort to date (21 patients and 24 PHPTs). Detailed information on patients, pseudotumours, surgery-related data, and perioperative complications was reported. Most PHPTs (75%) achieved and maintained favorable outcomes (defined as absence of severe postoperative complications or recurrence) during the follow-up. Intraoperative use of abundant gelatin sponges for cavity filling and haemostasis draws attention to severe embolism risks. Besides, the study introduces a novel classification scheme for PHPTs based on pseudotumour features and different surgical approaches required. The mid-term outcomes and detailed information are also reported separately by the PHPT classification, which can serve as a reference for further surgical treatment of PHPTs.

Implications of all the available evidence

Our findings suggested that surgical treatment for PHPTs is feasible and relatively safe convoyed by appropriate coagulation factor replacement strategies. Symptomatic and progressive PHPTs should undergo surgical intervention at the earliest opportunity to mitigate surgical risks. The introduced classification scheme for PHPTs is practical in determining surgical procedures and anticipating outcomes. Further prospective studies are required to verify the outcomes of surgical treatment for PHPT and validate the effectiveness of the classification scheme.

to fatal haemorrhaging, destructive bone erosion, as well as adjacent viscus and neurovascular compression.[4–6] The most frequent morbid sites for HPT are thigh (~56%) followed by pelvis (~30%).[7,8] Pelvic haemophilic pseudotumour (PHPT) is typically defined as HPT arising from pelvis, iliopsoas, and retroperitoneum, where the customary presenting complaint has been progressive augmenting lower abdominal mass with or without pain.[9–11] The compressive impact of PHPT on surrounding viscera such as ureter, peripheral nerve, colorectum, and blood vessel can frequently lead to hydronephrosis, nerve paralysis, intestinal obstruction, rectal irritation sign, and lower extremity edema.[7,10,12] In addition, PHPTs are commonly larger than other HPTs and pose an increased management complexity.[7,10]

The management of PHPT still lacks well-established protocols. Local aspiration, radiotherapy, interventional embolisation, and surgical excision have been reported for PHPT treatment, of which surgical excision of PHPT was believed to pose optimal outcomes.[2,13–19] However, the low incidence of PHPT and demanding surgical technique for PHPT excision results in few reports on the outcome of surgical treatment of PHPT, and no article reports mid-term follow-up outcomes of surgical treatment of PHPT in a relatively large cohort. Furthermore, there is still an absence of a classification system of PHPT to guide the rational treatment for such difficult cases. In this study, we report the 7.1-year outcome of 21 patients with 24 PHPTs that underwent surgical treatment. A novel PHPT classification based on pseudotumour feature and surgical treatment strategy is also introduced.

Methods

Study design and data collection

The study was conducted between June 25, 2004, and July 18, 2023, in Peking Union Medical College Hospital and Nanfang Hospital in China. PHPTs were diagnosed through clinical manifestation, physical examination, and radiological imaging, with a distinctive enlarging encapsulated cystic mass located in the pelvis, accompanied by a clear history of haemophilia. The inclusion criteria involved all patients who underwent PHPT surgical treatment. Demographic features, PHPT characteristics, surgery-related information, and perioperative complications were extracted from medical records. The follow-up data was collected through outpatient clinic and telephone interviews. CARE reporting guidelines were referred for the organising and writing of this article.[20]

Ethics

The study was approved by the institutional review board of Peking Union Medical College Hospital (S–K148) and Nanfang Hospital (NFEC-2023-460). Informed consent was obtained from all patients.

Surgical indications

The indications for PHPT excision encompassed four criteria: (1) Progressive enlarged PHPT that

unresponsive to conservative treatment such as coagulation factor replacement and physiotherapy; (2) Pelvic bone destruction with risk of pelvic discontinuity; (3) Symptomatic adjacent structure compression (viscera, nerve, or vessel); (4) Spontaneous PHPT rupture or perforation either with or without infection (either current or impending).

Surgery-based PHPT classification

Based on pseudotumour features and surgical requirements, we categorised PHPT into four types: (I) PHPT confined solely to soft tissue; (II) PHPT involving bony pelvic without pelvic discontinuity; (III) PHPT with pelvic discontinuity; (IV) PHPT complicated with infection resulting from spontaneous rupture or enteric fistulas development (Fig. 1).

Patients

21 consecutive male patients with 24 PHPTs (21 primary PHPTs and three recurrent PHPTs) that underwent surgical treatment were involved in the study. Of the 24 PHPTs, 11 (45.8%) were identified as Type I, five (20.8%) as Type II, three (12.5%) as Type III, and five (20.8%) as Type IV. The mean follow-up duration was 7.1 ± 4.9 years (Range, 0.1–19.3 years). The mean BMI of involved patients was 23.1 ± 3.5 (Range, 17.3–28.4). Of the involved patients, 19 (90.5%) were identified as haemophilia A and two (9.5%) as haemophilia B. The haemophilia severity was classified as mild (FVIII:C or FIX:C > 5%) in five patients (23.8%), moderate (1% < FVIII:C or FIX:C < 5%) in six patients (28.6%), and severe (FVIII:C or FIX:C < 1%) in ten patients (47.6%).[21] Notably, none of the patients in our study was detected positive for coagulation factor inhibitors. The blood type of involved patients was identified as A in six patients (28.6%), B in three (14.3%), AB in six (14.3%), and O in nine (42.9%). At the time of surgery, the patients had a mean age of 37.0 ± 9.5 years (Range, 24–52 years). Sixteen patients (76.2%) received varying duration of conservative treatment in the haematology department of our hospitals or other medical centres before undergoing surgical treatment. Eight patients (38.1%) were

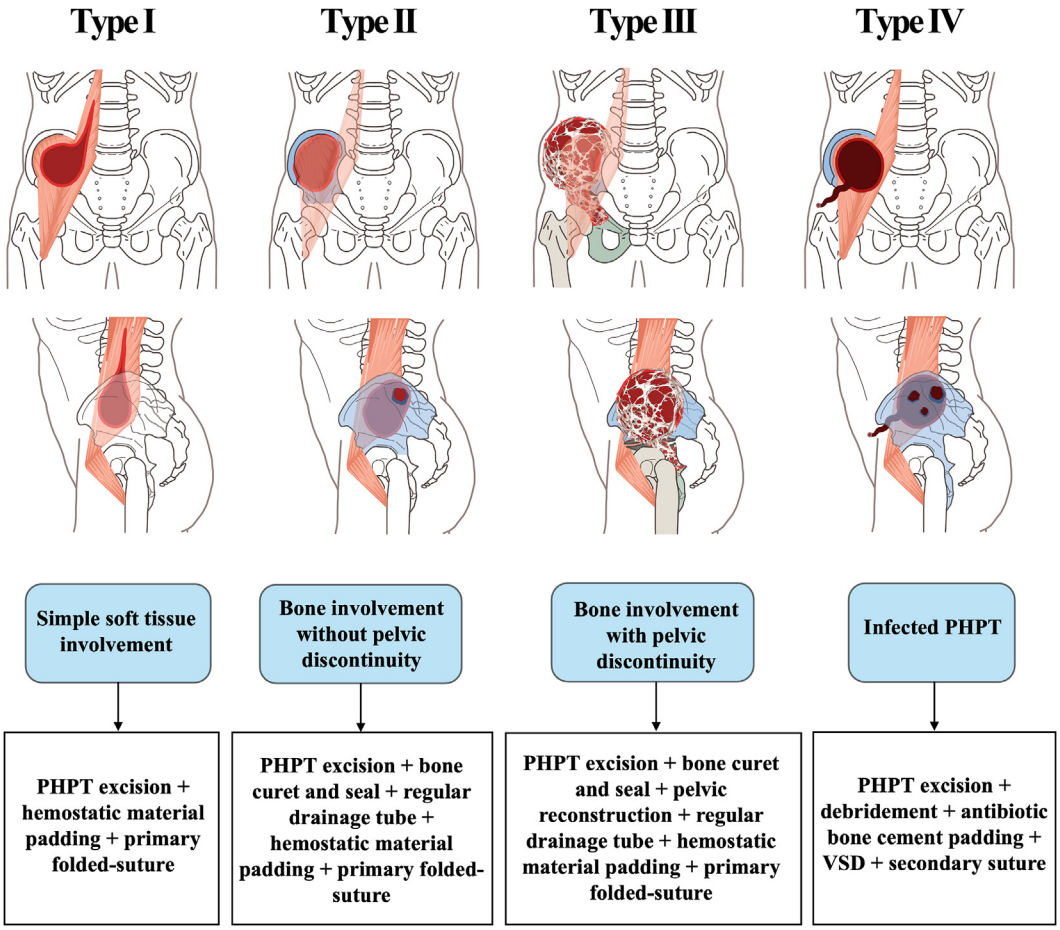

Fig. 1: **The classification scheme for PHPTs**. PHPTs with different characteristics require different surgical procedures. PHPT, pelvic haemophilic pseudotumour.

positive for hepatitis C and one (4.8%) was positive for hepatitis B. No patient was positive for human immunodeficiency virus (HIV). The demographic characteristics of involved patients and patients in each category are summarised in Table 1.

Information of PHPTs

The mean maximum diameter of PHPTs upon surgery was 17.0 ± 7.7 cm (Range, 4.3–40.0 cm). The median time from PHPT-related symptom onset to surgery was six years (IQR, 3.0–12.0 years, Range, 0.5–28.0 years). The location of PHPTs involved iliopsoas in 16 cases (66.7%), ilium in 13 cases (54.2%), sacrum in five cases (20.8%), hip joint in four cases (16.7%), pubis in four cases (16.7%), retroperitoneal space in three cases (12.5%), ischium in three cases (12.5%), gluteus maximus in one case (4.2%). The preoperative complications of PHPTs include penetrating skin sinus formation in two cases (8.3%), spontaneous pseudotumour rupture in two cases (8.3%), and ascending colon fistula development in one case (4.2%), all of which lead to preoperative pseudotumour infection (n = 5, 20.8%). Besides, the localised compression-related comorbidities of PHPTs include hydronephrosis resulting from ureteral obstruction in eight cases (33.3%), peripheral nerve paralysis-associated hypoesthesia and weakness of lower extremities in four cases (16.7%), lower limb edema caused by iliac vein compression in two cases (8.3%), intestinal obstruction in two cases (8.3%), and rectal irritation signs in one case (4.2%). Five patients had simultaneous HPT in other focus outside of pelvis, of which two were located in ipsilateral thigh, two in contralateral thigh, and one in contralateral gluteus maximus. The PHPT characteristics of involved patients and patients in each category are summarised in Table 2.

Coagulation factors replacement strategy

Patients with haemophilia A were supplemented with either plasma-derived or recombinant factor VIII (FVIII), whereas patients with haemophilia B were treated with prothrombin complex concentrate (PCC). All patients with haemophilia A underwent coagulation factor pharmacokinetic (PK) preliminary tests to tailor a customised perioperative coagulation factor supplement strategy. The coagulation factor dosage was titrated accordingly to maintain the peak factor concentration at 100% on the day of surgery, 80% on postoperative day (POD) one to three, 60% on POD four to seven, and 20%–30% thereafter, which was maintained for a minimum of six weeks after surgery. On the day of surgery, coagulation factors were administered as a single bolus infusion 1 h before the surgery, with an additional bolus given if the operation exceeded 6 h. In addition, a routine antifibrinolytic agent (tranexamic acid) was administered upon the accomplishment of general

	All PHPTs (n = 24, 21 patients)	Type I (n = 11, 10 patients)	Type II (n = 5, 5 patients)	Type III (n = 3, 3 patients)	Type IV (n = 5, 5 patients)
Age[a] (yr)	37.0 ± 9.5 (24–52)	35.1 ± 6.6 (24–45)	32.6 ± 11.0 (26–52)	36.0 ± 14.0 (26–52)	45.2 ± 7.7 (32–51)
Body mass index[a] (kg/m^2)	23.1 ± 3.5 (17.3–28.4)	23.9 ± 2.6 (19.5–26.8)	22.2 ± 4.9 (17.6–28.1)	22.1 ± 3.9 (17.6–24.5)	21.4 ± 4.2 (17.3–28.4)
Hospital stays[a] (d)	36.3 ± 31.9 (14–169)[c]	24.8 ± 8.9 (14–38)	29.8 ± 2.0 (27–32)	34.5 ± 3.5 (32–37)[c]	71.8 ± 60.6 (24–169)
Follow-up duration[a] (yr)	7.1 ± 4.9 (0.1–19.3)[c]	7.2 ± 4.1 (2.0–16.6)	9.8 ± 6.4 (3.8–19.3)	9.5 ± 3.9 (6.8–12.3)[c]	4.8 ± 4.6 (0.1–9.9)
Type of haemophilia[b]					
Haemophilia A	19 (90.5%)	10 (100.0%)	3 (60.0%)	3 (100.0%)	5 (100.0%)
Haemophilia B	2 (9.5%)	0 (0.0%)	2 (40.0%)	0 (0.0%)	0 (0.0%)
Haemophilia severity[b]					
Mild	5 (23.8%)	2 (20.0%)	1 (20.0%)	1 (33.3%)	3 (60.0%)
Moderate	6 (28.6%)	2 (20.0%)	2 (40.0%)	1 (33.3%)	1 (20.0%)
Severe	10 (47.6%)	6 (60.0%)	2 (40.0%)	1 (33.3%)	1 (20.0%)
Patients with inhibitor prior to treatment[b]	0 (0.0%)	0 (0.0%)	0 (0.0%)	0 (0.0%)	0 (0.0%)
Blood type[b]					
A	6 (28.6%)	2 (20.0%)	2 (40.0%)	1 (33.3%)	3 (60.0%)
B	3 (14.3%)	1 (10.0%)	2 (40.0%)	0 (0.0%)	0 (0.0%)
AB	3 (14.3%)	1 (10.0%)	0 (0.0%)	1 (33.3%)	1 (20.0%)
O	9 (42.9%)	6 (60.0%)	1 (20.0%)	1 (33.3%)	1 (20.0%)
Communicable disease[b]					
Hepatitis B	1 (4.8%)	1 (10.0%)	0 (0.0%)	0 (0.0%)	0 (0.0%)
Hepatitis C	8 (38.1%)	6 (60.0%)	1 (20.0%)	1 (33.3%)	0 (0.0%)
AIDS	0 (0.0%)	0 (0.0%)	0 (0.0%)	0 (0.0%)	0 (0.0%)

PHPT, pelvic haemophilic pseudotumour; AIDS, acquired immune deficiency syndrome. [a]The values are given as the mean and the standard deviation, with the range in parentheses. [b]The values are given as the number of patients, with the percentage in parentheses. [c]Deceased case excluded.

Table 1: **Demographic characteristics.**

	All PHPTs (n = 24, 21 patients)	Type I (n = 11, 10 patients)	Type II (n = 5, 5 patients)	Type III (n = 3, 3 patients)	Type IV (n = 5, 5 patients)
Maximum diameter[a] (*cm*)	17.0 ± 7.7 (4.3–40.0)	15.5 ± 6.5 (4.3–24.0)	15.6 ± 3.8 (10.0–20.0)	26.0 ± 15.0 (20.0–30.0)	17.1 ± 12.8 (10.5–40.0)
Duration of symptoms[b] (*yr*)	6.0 (3.0–12.0)	4.5 (3.3–7.3)	3.0 (2.0–9.0)	10.0	15.0 (9.0–22.0)
(Recurrent PHPT excluded)	(0.5–28.0)	(0.5–20.0)	(1.0–12.0)	(7.0–13.0)	(3.0–28.0)
Invaded structures[c]					
Iliopsoas	16 (66.7%)	9 (81.8%)	3 (60.0%)	1 (33.3%)	3 (60.0%)
Ilium	13 (54.2%)	0 (0.0%)	5 (100.0%)	3 (100.0%)	5 (100.0%)
Sacrum	5 (20.8%)	0 (0.0%)	3 (60.0%)	2 (66.7%)	0 (0.0%)
Hip joint	4 (16.7%)	0 (0.0%)	2 (40.0%)	1 (33.3%)	1 (20.0%)
Pubis	4 (16.7%)	0 (0.0%)	1 (20.0%)	2 (66.7%)	1 (20.0%)
Retroperitoneal space	3 (12.5%)	2 (18.2%)	1 (20.0%)	0 (0.0%)	0 (0.0%)
Ischium	3 (12.5%)	0 (0.0%)	1 (20.0%)	1 (33.3%)	1 (20.0%)
Gluteus maximus	1 (4.2%)	0 (0.0%)	1 (20.0%)	0 (0.0%)	0 (0.0%)
Preoperative pseudotumour complications[c]					
Pseudotumour infection	5 (20.8%)	0 (0.0%)	0 (0.0%)	0 (0.0%)	5 (100.0%)
Skin sinus	2 (8.3%)	0 (0.0%)	0 (0.0%)	0 (0.0%)	2 (40.0%)
Spontaneous pseudotumour rupture	2 (8.3%)	0 (0.0%)	0 (0.0%)	0 (0.0%)	2 (40.0%)
Ascending colon fistula	1 (4.2%)	0 (0.0%)	0 (0.0%)	0 (0.0%)	1 (20.0%)
Preoperative compression comorbidities[c]					
Hydronephrosis	8 (33.3%)	6 (54.5%)	0 (0.0%)	1 (33.3%)	1 (20.0%)
Peripheral nerve paralysis	4 (16.7%)	2 (18.2%)	1 (20.0%)	1 (33.3%)	0 (0.0%)
Lower limb edema	2 (8.3%)	1 (9.1%)	0 (0.0%)	1 (33.3%)	0 (0.0%)
Intestinal obstruction	2 (8.3%)	2 (18.2%)	0 (0.0%)	0 (0.0%)	0 (0.0%)
Rectal irritation signs	1 (4.2%)	0 (0.0%)	0 (0.0%)	0 (0.0%)	1 (20.0%)
Locations of concurrent HPTs[c]					
Ipsilateral thigh	2 (8.3%)	1 (9.1%)	0 (0.0%)	0 (0.0%)	1 (20.0%)
Contralateral thigh	2 (8.3%)	1 (9.1%)	1 (20.0%)	0 (0.0%)	0 (0.0%)
Contralateral gluteus maximus	1 (4.2%)	1 (9.1%)	0 (0.0%)	0 (0.0%)	0 (0.0%)

PHPT, pelvic haemophilic pseudotumour; HPT, haemophilic pseudotumour. [a]The values are given as the mean and the standard deviation, with the range in parentheses. [b]The values are given as the median, with the IQR and range in parentheses. [c]The values are given as the number of patients, with the percentage in parentheses.

Table 2: **Characteristics of PHPTs.**

anesthesia. During the postoperative day one to day six, the required coagulation factor dose is administered every 12 h. On POD seven and thereafter, the daily coagulation factor is supplemented as a single bolus. Patients with haemophilia B received empirical PCC supplements under the guidance of haematologists at our hospitals. Extra blood transfusion was administered as indicated based on perioperative haemoglobin levels, intraoperative bleeding, and postoperative drainage.

Imaging examination and arterial embolisation

Patients underwent preoperative plain X-ray, pelvic computed tomography (CT), or magnetic resonance imaging (MRI) assessment to elucidate the pelvic destruction and the anatomical correlation between PHPT and adjacent structures. When a major vessel is suspected to be involved, computed tomography angiography (CTA) should be performed, and preoperative embolisation may play a dominant role in minimising intraoperative bleeding. Regular follow-up CT or MRI examination was recommended to detect any potential recurrence of PHPT.

Surgical procedures

Before the surgery, the patient was positioned on lateral decubitus or supine with affected side of pelvis elevated on sandbags after anesthesia. The surgical site was thoroughly sterilised and draped followed by an oblique ilioinguinal skin incision made on the lateral aspect of the PHPT, extending proximally for 15–20 cm. The subcutaneous tissues, abdominal muscles, and transversalis fascia were incised layer-by-layer and separated from the superficial wall of the PHPT. The superficial wall of the PHPT was then opened, and the pseudotumour was resected with a philosophy of inside-out approach. After removal of the intracapsular contents completely, the deep layer of pseudotumour wall was excised as much as possible. However, the superficial capsule was preserved owing to the undistinguished boundary between the wall and surrounding normal tissue. For PHPTs confined in soft tissue (Type I), the cavity was then filled with haemostatic gelatin sponge after careful haemostasis and the wall was closed using the folded-suture technique to minimise the residual cavity space. For PHPTs with pelvic bone erosion but

without pelvic discontinuity (Type II), the eroded bone was completely curetted, and the surface was sealed with bone wax and gel-sponge. A regular drainage tube was then well placed. For PHPTs with pelvis destruction and discontinuity (Type III), the pelvis was further reconstructed. For PHPTs with an infected open wound (Type IV), debridement was performed and the space cavity after partial pelvis resection was filled with antibiotic-loaded bone cement. Vacuum sealing drainage (VSD) was placed for wound management and infection eradication, and the wound was closed at the second stage (Fig. 1).

Statistical analysis

Continuous variables are presented as the mean and standard deviation or as the median and interquartile range (IQR), depending on the normality of the distribution. Categorical data are reported as the number and percentage. All statistical analyses were conducted using SPSS (version 23.0; IBM).

Role of the funding source

All authors confirmed that they had full access to all the data in the study and accepted responsibility for the decision to submit for publication. There was no funding source for this study.

Results

21 consecutive male patients with 24 PHPTs (21 primary PHPTs and three recurrent PHPTs) were involved in the study. The median time from PHPT-related symptom onset to surgery was six years (IQR, 3.0–12.0 years, Range, 0.5–28.0 years) and the mean maximum diameter of PHPTs upon surgery averaged was 17.0 ± 7.7 cm (Range, 4.3–40.0 cm). At the time of surgery, the patients had a mean age of 37.0 ± 9.5 years (Range, 24–52 years). Of the 24 PHPTs, 11 (45.8%) were identified as Type I, five (20.8%) as Type II, three (12.5%) as Type III, and five (20.8%) as Type IV. The mean hospital stays were 36.3 days (Range, 14–169 days). Five PHPTs (including four Type I and one Type III) underwent arterial embolisation before surgery. The mean surgical duration was 192 ± 77 min (Range, 60–330 min) and the median intraoperative blood loss was 400 mL (IQR, 225–950 mL, Range, 100–3000 mL). The median transfused amounts included 0 units of red blood cells (IQR, 0–4 units, Range, 0–12 units), 0 mL of plasma (IQR, 0–400 mL, Range, 0–800 mL), and 0 whole blood (IQR, 0–0 mL, Range, 0–1500 mL). The median postoperative 24-h drainage was 230 mL (IQR, 110–450 mL, Range, 10–2250 mL).

As for the complications, all compression-associated complications demonstrated a significant resolve after

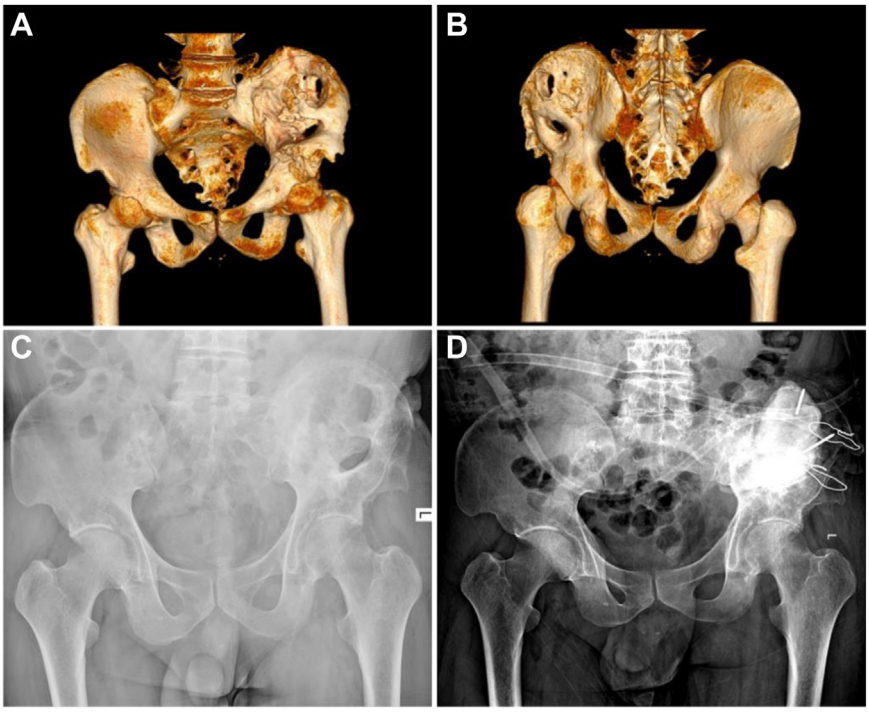

Fig. 2: **Surgical treatment of a patient with severely infected PHPT**. Preoperative three-dimensional reconstructed computed tomography images (A,B) and plane X-ray imaging (C) of the patient. Postoperative plane X-ray imaging of the patient after antibiotic-loaded bone cement padding, multiple debridement, and VSD treatment (D). Consent for the use of images has been obtained from the patient. PHPT, pelvic haemophilic pseudotumour; VSD, vacuum sealing drainage.

PHPT excision except one patient (Type III) experienced irreversible renal failure due to prolonged ureteral compression. Postoperative wound infection and poor wound healing were observed in four cases (16.7%), all of which occurred in Type IV PHPTs. After multiple debridement and VSD treatment, the infection was eradicated with successful wound healing in all cases (Fig. 2). One patient (Type II) developed a haematoma in the excised pseudotumour cyst, which was relieved after haematoma drainage. Notably, one patient with giant PHPT (Type III) developed cardiopulmonary arrest and irreversible respiratory circulatory failure in the operation room and died even after extracorporeal membrane oxygenation (ECMO) support two days later (Fig. 3). During the follow-up period, five PHPTs (20.8%, four Type I and one Type II) experienced recurrence. Three of these recurrent PHPTs were resected again, of which two were successfully treated without recurrence at the latest follow-up, while one recurred two years after the second excision and subsequently underwent interventional puncture drainage. One recurrent PHPT did not receive further treatment due to cost concerns. The surgical information and postoperative complication data are presented in Table 3.

Discussion

PHPT is a rare complication of haemophilia occurs in approximately 1–2% of patients with haemophilia,[2] of which about 30% were PHPT.[7] This study presented the largest PHPT cohort so far involving 21 patients with 24 PHPTs with a mean 7.1-year follow-up. Patients with PHPT can be asymptomatic or complaining of clinical pain or swelling. The swelling sometimes being massive and on occasion occur spontaneous rupture and ulceration leading to severe haemorrhage and secondary infection.[22,23] Five patients in our study developed spontaneous rupture or

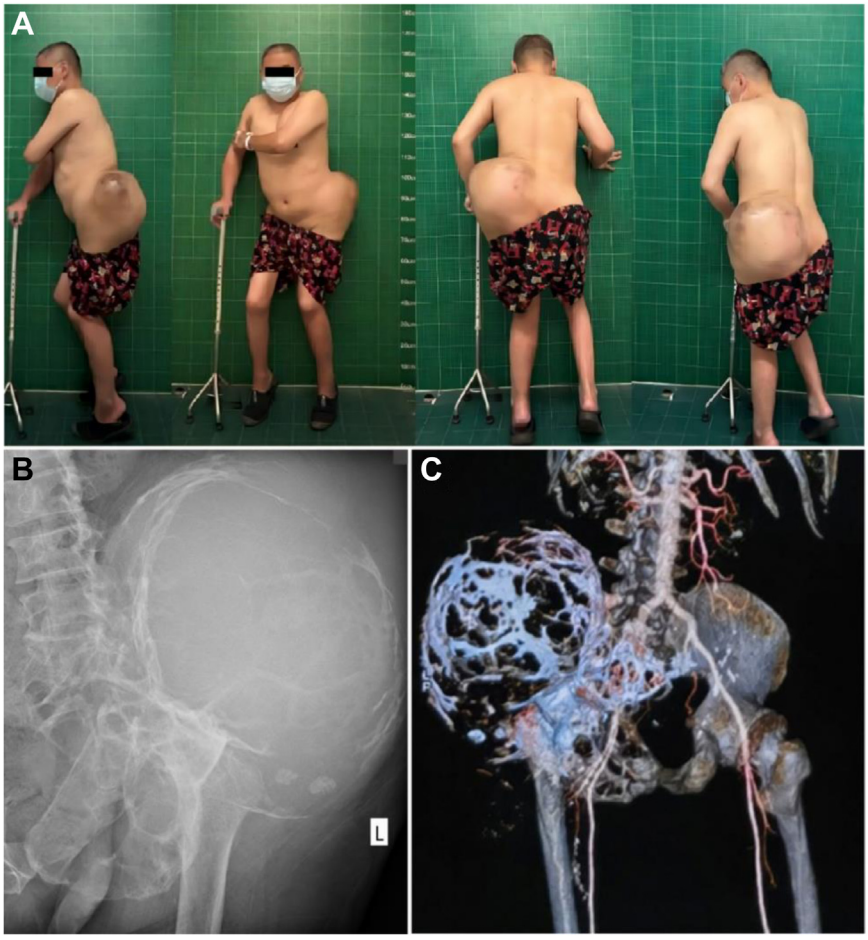

Fig. 3: **A patient with giant PHPT and pelvic discontinuity**. Digital photograph (A), plain X-ray imaging (B), and three-dimensional reconstructed computed tomography angiography image (C) of the patient. Consent for the use of images has been obtained from the patient. PHPT, pelvic haemophilic pseudotumour; ECMO, extracorporeal membrane oxygenation.

	All PHPTs (n = 24, 21 patients)	Type I (n = 11, 10 patients)	Type II (n = 5, 5 patients)	Type III (n = 3, 3 patients)	Type IV (n = 5, 5 patients)
Operative time[a] (*min*)	192 ± 77 (60–330)	173 ± 74 (60–295)	173 ± 88 (75–300)	262 ± 72 (186–330)	211 ± 66 (120–280)
Intraoperative bleeding[b] (*mL*)	400 (225–950) (100–3000)	300 (150–400) (100–3000)	300 (100–450) (100–600)	1600 (1300–2300) (1000–3000)	400 (400–1000) (300–2500)
Intraoperative transfusion[b]					
Red blood cells (*unit*)	0 (0–4) (0–12)	2 (1–4) (0–12)	0 (0–0) (0–4)	0 (0–6) (0–12)	4 (0–4) (0–6)
Plasma (*mL*)	0 (0–400) (0–800)	200 (0–400) (0–800)	0 (0–0) (0–0)	400 (200–600) (0–800)	400 (0–400) (0–400)
Whole blood (*mL*)	0 (0–0) (0–1500)	0 (0–0) (0–1500)	0 (0–0) (0–400)	320 (160–810) (0–1300)	0 (0–0) (0–0)
Postoperative 24-h drainage[b] (*mL*)	230 (110–450) (10–2250)[d]	180 (65–419) (10–600)	230 (200–400) (100–450)	1425 (600–2250)[d]	280 (100–400) (100–500)
Postoperative complications[c]					
Wound infection	4 (16.7%)	0 (0.0%)	0 (0.0%)	0 (0.0%)	4 (80.0%)
Poor wound healing	4 (16.7%)	0 (0.0%)	0 (0.0%)	0 (0.0%)	4 (80.0%)
Haematoma	1 (4.2%)	0 (0.0%)	1 (20.0%)	0 (0.0%)	0 (0.0%)
Death	1 (4.2%)	0 (0.0%)	0 (0%)	1 (33.3%)	0 (0.0%)
Recurrence of PHPT	5 (20.8%)	4 (36.4%)	1 (20.0%)	0 (0.0%)	0 (0.0%)

[a]The values are given as the mean and the standard deviation, with the range in parentheses. [b]The values are given as the median, with the IQR and range in parentheses. [c]The values are given as the number of patients, with the percentage in parentheses. [d]Deceased case excluded. PHPT, pelvic haemophilic pseudotumour.

Table 3: **Surgery information and postoperative complications.**

fistula leading to pseudotumour infection, which significantly complicates perioperative management and prolongs hospital stays (mean 27.4 vs 71.8 days). More commonly, the PHPT exerts compression on adjacent structures causing compression-associated symptoms. In our case series, the most frequent PHPT compression-associated complication was hydronephrosis (n = 8, 33.3%), followed by peripheral nerve paralysis (n = 4, 16.7%), lower limb edema (n = 2, 8.3%), intestinal obstruction (n = 2, 8.3%), and rectal irritation signs (n = 1, 4.2%). In these cases, surgical intervention of PHPTs may necessitate multidisciplinary cooperation involving urologists, vascular surgeons, and gastrointestinal surgeons. Notably, due to the insidious onset of PHPT, early diagnosis is challenging and often results in delayed surgical intervention.[6] In our cohort, patients typically experienced a median course of six years from self-awareness of PHPT to index surgery. During this period, all patients exhibited progressive pseudotumour enlargement, with a mean maximum diameter of 17.0 cm upon hospitalisation. In addition, the risk of compression-associated complications also increases over time. Thus, regular pelvic radiological examinations (semiannual) are recommended for patients with haemophilia.

There are no defined criteria for surgical indications of PHPT excision. We recommend the following surgical indications: (1) Progressively enlarged PHPT that unresponsive to conservative treatment such as coagulation factor replacement and physiotherapy; (2) Pelvic bone destruction with risk of pelvic discontinuity; (3) Symptomatic adjacent structure compression (viscera, nerve, or vessel); (4) Spontaneous PHPT rupture or perforation either with or without infection (either current or impending). Besides, it is worth noting that PHPTs rarely respond favorably to conservative treatment, thus early surgery upon diagnosis is recommended. Furthermore, we introduce a novel classification for PHPT, including (I) PHPT confined solely to soft tissue; (II) PHPT involving the bony pelvis without pelvic discontinuity; (III) PHPT causing pelvic discontinuity; (IV) Infectious PHPT. Different categories of PHPTs require different surgical strategies (Fig. 1). Given the frequently unclear boundary between PHPT and adjacent tissues, we emphasise intralesional excision as the primary surgical strategy, and the preservation of the superficial capsule of PHPT is recommended. In cases with pelvic bone involvement, efforts should be made to curette the affected bone. For cases with pelvic discontinuity, the goal of surgery was to reconstruct the pelvic ring. In our cohort, two cases (8.3%) underwent pelvic reconstruction, for which one case use allograft bone, while the other utilised screws and rods. During the follow-up period, no complications related to pelvic instability were observed in the two cases. Owing to the big cavity after the tumour resection, gelatin sponges were commonly used for the cavity filling and intraoperative haemostasis. However, the cancellous bone and blood vessel sinus can be opened after pseudotumour resection, which provides an entry for the exogenous clot to enter the blood circular system. In such condition, gelatin sponges may carry a risk of embolism.[24,25] In our study, one case (Type III) with a giant PHPT suffered a sudden cardiac arrest during surgery, and pulmonary embolism was suspected

according to transesophageal echo and computed tomographic pulmonary angiography. The gelatin of sponge was suspected to be the origin of the embolism, raising caution in using abundant sponge in such a situation. In addition, despite the median intraoperative blood loss for PHPT excision being 400 mL in our cohort, each category has cases with bleeding exceeding 1000 mL, all of which were ascribed to vessel injury due to unclear anatomy. Thus, meticulous intraoperative dissection and adequate preoperative blood preparation are emphasised. For infectious PHPTs, the antibiotic-loaded bone cement was employed to fill the cavity in conjunction with multiple debridement. VSD was introduced for postoperative management of infectious PHPTs, which was proved to possess high effectiveness and safety, as all postoperative infected wounds healed successfully, and no extra bleeding events related to VSD were observed. For patients with concurrent HPT in other locations (e.g., thighs), we recommend combined surgical resections of these HPTs at one stage, which avoids costs and coagulation factor inhibitor-generating risks associated with additional surgeries.

Compared with HPTs in other sites, PHPTs are more prone to recurrence. Complete HPT wall resection and cavity closure are of great importance for preventing HPT recurrence.[18,26] However, most PHPTs lack distinct boundaries closely interconnecting with surrounding viscera, blood vessels, and nerves, which made it difficult to get complete pseudotumour resection. Besides, the boundary of tumour always exceeds the lower abdomen, making it difficult to get enough extracorporeal pressure after surgery. All the above factors might contribute to the high recurrence rate of PHPT after operation. Thus, for PHPT, we recommend sufficient and prolonged use of coagulation factor for a minimum of six weeks postoperatively to get sufficient haemostasis. In our cohort, a majority of patients (n = 18, 85.7%) expressed general satisfaction with the surgical treatment during the follow-up.

The primary limitations of this study are the retrospective design and a relatively small number of cases, especially after categorisation. Despite achieving a mean follow-up time of 7.1 years, four patients were followed for less than two years. Furthermore, the rarity of PHPTs led to heterogeneous patient involvement. Research with larger sample sizes and longer follow-up periods is warranted to further standardise the surgical treatment of PHPT and verify the effectiveness of the PHPT classification system.

In summary, surgical treatment for PHPTs is feasible and relatively safe. Symptomatic and progressive PHPTs should undergo surgical intervention as early as possible to minimise the risks associated with PHPT enlargement, infection, and spontaneous rupture, all of which can significantly impact subsequent surgical procedures or even jeopardise patient's life. However, it is essential to conduct meticulous preoperative assessments and perioperative management. Intraoperative use of abundant gelatin sponges in PHPT excision draws attention to severe embolism complications.

Contributors

X.W., B.C., and B.F. designed and supervised the study. X.W. and Y. L. were involved in the project administration. Y.B., Y.W.X., and Y.M.X. were involved in data acquisition and curation. Z.L., W.Z., and X.Z. have accessed and verified the underlying data. Y.B. analysed the data and wrote the initial draft. X.W., B.F., and Z.L. critically revised the manuscript. All authors confirmed that they had full access to all the data in the study and accepted responsibility for the decision to submit for publication.

Data sharing statement

De-identified data from the study which is not found in this manuscript can be made available upon reasonable request by contacting the corresponding author.

Declaration of interests

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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Appendix A. Supplementary data

Supplementary data related to this article can be found at https://doi.org/10.1016/j.eclinm.2024.102497.

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