

Primary Renal Ewing Sarcoma with Level III IVC Tumour Thrombus and Acute Pulmonary Embolism

Surgical Challenges and Critical Decisions

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OBJECTIVE

Primary Ewing sarcoma of the kidney is rare and aggressive, and it can mimic renal cell carcinoma. With inferior vena cava (IVC) tumour thrombus it is a formidable surgical entity, and concurrent acute pulmonary embolism (PE) adds a critical dilemma: therapeutic anticoagulation has to be balanced against the high risk of haemorrhage during major oncological resection.

We present a rare case managed successfully by a multidisciplinary team.

CASE PRESENTATION

- **36-year-old man** (90 kg), no comorbidities
- Two-month history of progressive **left flank heaviness**
- Left renal mass on ultrasound, characterised on contrast-enhanced CT
- Locally advanced mass invading the **pancreatic tail, spleen and diaphragmatic crus**
- **Mayo Level III** thrombus extending into the retrohepatic IVC
- **Acute bilateral PE** with right ventricular strain

MANAGEMENT

ANTICOAGULATION PLAN

- **AT DIAGNOSIS**
Therapeutic enoxaparin started
- **48 H BEFORE SURGERY**
Switched to unfractionated heparin
- **12 H BEFORE SURGERY**
Heparin stopped
- **POSTOPERATIVELY**
IV heparin recommenced

SURGICAL PROCEDURE

After multidisciplinary tumour board review and high-risk consent:

- **Left radical nephrectomy** with open IVC tumour thrombectomy
- En bloc **distal pancreatectomy** and **splenectomy**
- **Access:** Mercedes-Benz incision with bilateral colonic mobilisation

VASCULAR CONTROL

- Distal IVC, right renal vein and proximal IVC controlled before thrombus extraction
- Lumbar vein control difficult: inferiorly displaced IVC, collateral flow and limited access

OPERATIVE CHALLENGES

- Tumour filled almost the entire left abdomen, invading the crus and obliterating normal planes
- Freed from the stomach, but inseparable from the pancreatic tail and spleen
- Extensive venous collaterals from IVC obstruction caused significant bleeding
- Meticulous dissection needed around the coeliac trunk and SMA

KEY DECISIONS

- 1 Operate despite acute PE, after tumour board review
- 2 Individualised bridging from LMWH to heparin
- 3 En bloc pancreatectomy–splenectomy for oncological clearance
- 4 Product-led resuscitation with planned surgical–anaesthetic pauses

RESULTS

12 h 1 min
Operative time

25 min
IVC cross-clamp time

3.3 L
Estimated blood loss

14 h
To extubation

The lumbar veins could not be safely clamped because of collateral flow and limited access, so thrombus extraction relied on meticulous IVC dissection.

Transfusion: 8 units PRBCs, 8 units FFP, 1 adult therapeutic dose of platelets

To protect the strained right ventricle, fluids were strictly restricted and resuscitation was product-led, avoiding haemodilution and volume overload.

He stayed haemodynamically stable and restarted IV heparin (1,700 IU/h) immediately after extubation. After 24 hours in ICU he moved to the high-dependency unit and made an **uncomplicated recovery**.

CONCLUSION

Primary renal Ewing sarcoma can present just like RCC with IVC tumour thrombus, so histology with immunohistochemistry is key to the diagnosis and to planning systemic therapy.

Acute venous thromboembolism does **not preclude definitive resection** in carefully selected patients with Level III IVC thrombus.

Tailored anticoagulation, precise vascular control and close surgical–anaesthetic coordination are central to a good outcome.

HISTOPATHOLOGY

FINAL DIAGNOSIS

Malignant round blue cell tumour, favouring Ewing sarcoma

- Superior pole, **17 cm** (170 × 155 mm)
- **High grade**, mitoses 10–15 / 10 HPF
- Necrosis present

pT4
> 15 cm

pN0
0 / 3 nodes

pMx
AJCC 8th

IMMUNOHISTOCHEMISTRY

CD99 +

Positive in tumour cells

NKX2.2 +

Positive in tumour cells

Desmin –

Excludes myogenic tumour

SS18-SSX –

Excludes synovial sarcoma

EWSR1 FISH recommended for molecular confirmation of this rare diagnosis.

EXTENT OF DISEASE

IVC thrombus	Tumour thrombus
Left renal vein	Involved
Renal sinus fat	Involved
Pancreatic tail	Focally involved
Spleen · Omentum	Free
Vascular & ureteric margins	Free

IMAGING & INTRA-OPERATIVE FINDINGS



Fig. 1 Preoperative ultrasound



Fig. 2 Preoperative ultrasound



Fig. 3 Preoperative ultrasound

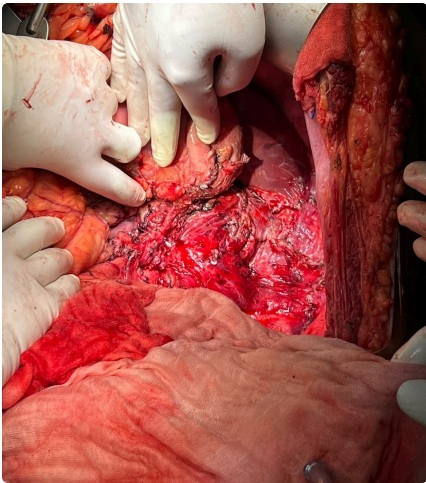


Fig. 4 Mobilisation of the tumour mass

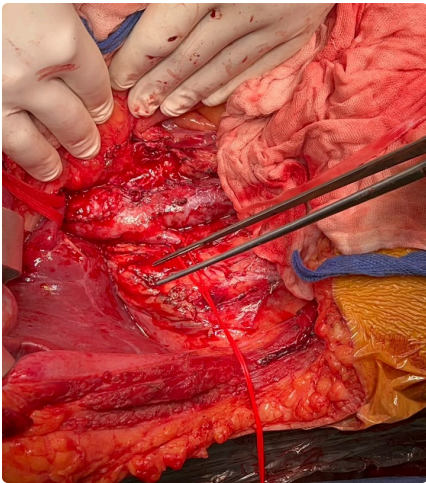


Fig. 5 Vascular control with vessel loop

