CASE REPORT

Open removal of a retained retrohepatic inferior vena cava filter with a residual primary neuroectodermal renal tumoral thrombus

Carlos A Hinojosa, Adriana Torres-Machorro, Rene Lizola, Javier E Anaya-Ayala

Department of Surgery, Section of Vascular Surgery and Endovascular Therapy, Instituto Nacional de Ciencias Medicas y Nutricion "Salvador Zubiran", Mexico, Mexico

Correspondence to
Professor Carlos A Hinojosa,
carlos.a.hinojosa@gmail.com

Accepted 15 October 2015

SUMMARY

Primary neuroectodermal renal tumours (PNET) are rare and aggressive neoplasms; thrombosis of the inferior vena cava (IVC) is associated with this entity. We report here the case of a 19-year-old man who experienced a new onset of abdominal pain. A CT scan revealed a large left renal mass, perirenal haematoma and IVC thrombosis. Owing to an acute drop in haemoglobin and subsegmentary pulmonary embolism, he underwent emergency selective renal artery angiography and embolisation of bleeding vessels and IVC filter (IVCF) placement. Once stable, he underwent a left radical nephrectomy and IVC thrombectomy; the pathology report confirmed PNET. 6 months later, imaging revealed a residual tumoral thrombus in the IVCF located in the retrohepatic IVC. The patient underwent removal of this device and the thrombus via a right thoracoabdominal approach. He recovered well and at 4 months, he continues his chemotherapy cycles.

BACKGROUND

Primary neuroectodermal renal tumours (PNET) are rare neoplasms characterised by aggressive behaviour, and thrombosis of the inferior vena cava (IVC) is a complication that was found in 50% of cases in a recent publication.¹ On the basis of the tumour thrombus extension, tumours are classified as infrahepatic (I), retrohepatic (II), supra hepatic (III) and intra-atrial (IV).¹ Radical nephrectomy, excision of PNET and thrombus removal are recommended in all cases and temporary IVC filters (IVCF) can be placed before surgery as a prophylactic measure to prevent pulmonary embolism (PE).² Despite the tumoral thrombus extending to the level of the right atrium, IVC thrombectomy combined with immunotherapy has demonstrated improved prognosis in patients with this condition.³ We present a case of open removal of a retained IVCF with residual PNET thrombus located in the retrohepatic IVC through a right thoracoabdominal surgical approach.

CASE PRESENTATION

A 19-year-old man was referred to our institution with acute onset of diffuse abdominal pain in July 2014. A contrast-enhanced CT scan that had been performed at an outside facility revealed a left renal mass measuring 13×12 cm, based on this imaging study the radiologists reported a perirenal haematoma as a possible aetiology. A thrombus involving the left renal vein and IVC was also visualised

(figure 1). A renal scintigraphy demonstrated non-functional left kidney; laboratory studies including blood urea nitrogen and creatinine were within normal limits and urine cultures were negative. Owing to an acute drop in his haemoglobin level and subsegmentary PE, he was taken to the catheterisation laboratory for selective renal artery angiography and successful embolisation of the middle segmentary renal artery. During this procedure, he underwent placement of an IVCF via the right internal jugular vein placement by the interventional radiology team. No anticoagulation could be initiated because of the continuous expansion of the perirenal haematoma. A hypercoagulable panel confirmed hyperhomocysteinaemia with positive homozygous C677T associated with methylenetetrahydrofolate reductase. He was taken to the operating room (OR) for a radical left nephrectomy with IVC thrombectomy by the urology service. A 14×10×8.3 cm mass (figure 2) was resected and the pathology report demonstrated a high grade primitive neuroectodermal tumour with lymphovascular invasion and positive CD99 on immunohistochemistry. Neither lymph node metastasis nor capsule involvement was found, and positive surgical margins on the renal vein thrombus were also documented. Four cycles of chemotherapy based on vincristine, doxorubicin and cyclophosphamide

Figure 1 Abdominal CT scan, coronal view revealing a 12×13 cm perirenal mass; the intracaval thrombus is also visualised (black hollow arrow).

To cite: Hinojosa CA, Torres-Machorro A, Lizola R, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2015-212190

Figure 2 Gross pathology demonstrating a lobulated 14×10×8.3 cm mass with perirenal haematoma.

followed by ifosfamide and etoposide were administered as adjuvant therapy.

When the patient returned 6 months later, a repeat abdominal CT scan demonstrated a new partially occluding tumoral thrombus from the right renal vein fairly below the supra hepatic veins, where the IVCF was previously placed (figure 3). Owing to the positive surgical margins in the thrombus and the retained IVCF perforating the vessel wall, we decided to re-intervene. The patient was taken to the OR for removal of the device and the tumoral thrombus; once the patient was positioned, a right thoracoabdominal approach was pursued to facilitate proximal control of the suprahepatic IVC and suprahepatic veins. Following the complete mobilisation of the liver, a

Figure 3 Abdominal CT scan after the left nephrectomy and tumour resection demonstrating an intracaval residual tumoral thrombus (black arrow) and extrusion of the inferior vena cava (IVC) filter in the retrohepatic IVC.

careful dissection of the retrohepatic and infrarenal IVC was performed and a filter strut protruding from the retrohepatic vena cava was evident. Selective control of the IVC, right renal vein and suprahepatic veins was achieved with vessel loops and a Rumel tourniquet (figure 4A). Independent vascular control of the portal vein was also obtained. A retrohepatic cava vein was clamped just below the supra hepatic veins to avoid occlusion of portal flow and subsequent hypotension and bowel congestion. After the infrarenal segment of the abdominal aorta was exposed and encircled with a vessel loop, we cross-clamped the infrarenal IVC; subsequently, a longitudinal cavotomy revealed a tumoral thrombus. Thombectomy with IVCF extraction was successfully completed without compromising the caval diameter (figure 4B); then the cavotomy was closed primarily by restoring the caval flow. The filter was carefully inspected for missing elements. Finally, a 32 Fr chest tube was placed during thoracoabdominal closure (figure 5A,B).

The specimen reported CD99+ cells confirming a recurrent primitive neuroectodermal tumoral thrombus suspicion (figure 6). The patient recovered well from the surgery and was discharged home in stable condition. At 4 months, he continues to follow-up with the oncology service currently on anticoagulation therapy with rivaroxaban.

DIFFERENTIAL DIAGNOSIS

- ▶ Wilms' tumour
- ▶ Neuroblastoma
- ▶ Carcinoid tumour
- ▶ Renal cell carcinoma

OUTCOME AND FOLLOW-UP

The patient recovered well from the surgery and was discharged home and at 4 months he continues to follow-up with the oncology service.

DISCUSSION

Spontaneous non-traumatic kidney haemorrhage, known as the Wunderlich syndrome, is an uncommon cause of abdominal pain; well recognised entities associated with this manifestation included tumours, vascular malformations, renal artery aneurysms and portal hypertension.⁴ PNET are rare tumours occurring predominantly between the second and fourth decades (85% of cases) showing a male predominance. These tumours have an aggressive behaviour with a high rate of local recurrence and a tendency to metastasise to lymph nodes, lung, bone and bone marrow.⁵ To better address the diagnosis, an immunohistochemical analysis is necessary; CD99 staining greatly aids in recognising the ES (Ewing Sarcoma)/PNET family of tumours.⁶ Many PNET also stain for neural markers, including NSE, Leu-7, S-100 protein, synaptophysin and PGP9.5.⁷ The combination of surgical resection, postoperative radiotherapy and chemotherapy is the standard of treatment. When the presence of a tumoral thrombus complicates this entity, complex surgical approaches are necessary and these are based on the level and extension of the thrombus, the IVC patency and laterality of tumour.⁸ A thoracoabdominal surgical approach allows better visualisation of the retrohepatic IVC and facilitates dissection of the suprahepatic cava vein in comparison to a transabdominal approach. A profound knowledge of the anatomy of the retrohepatic segment of the IVC is critical in order to avoid vessel injury during this operation. In a recent anatomic observational study in cadaveric adult human livers by Gosh and Paul, the authors observed that the median length of the retrohepatic segment of the IVC measured on average 7.3 cm and was

Figure 4 Intraoperative view. (A) Through a right thoracoabdominal approach, the retrohepatic inferior vena cava (IVC) is exposed, after proximal IVC control (1), middle hepatic vein (2), right renal vein (3) and distal IVC (4) control; (B) a longitudinal cavotomy is performed, the IVC filter is visualized (black hollow arrow).

directed obliquely with respect to the vertical axis of the liver in 92.5% of cases. A total of 1376 ostia venae hepaticae were observed, and the median number of openings per liver was 7. The right hepatic vein had a single opening in 156 (97.5%) cases and the left and middle hepatic veins had a common opening in 144 (90%) cases. A longitudinal area on the anterior wall of the retrohepatic IVC to the right side of the midline was relatively avascular with 10.1% of the venous openings, of which 70% were a single opening of the right dorsal vein with a small diameter.⁹ The authors of this study concluded that during the liver hanging manoeuvre, a rightward direction of the dissecting forceps would avoid injury to the caudate vein and allow access to the safe avascular space in the retrohepatic IVC.⁹ IVCF removal is recommended to avoid complications associated with prolonged indwell times such as adjacent vessel perforation, filter fracture, embolisation, migration, repeat deep vein thrombosis, penetration of adjacent viscera or solid organ (eg, duodenum, liver parenchyma, pancreas) or embedding within adjacent bony structures.^{10 11} In some patients with levels II, III or IV disease with a patent IVC, exfoliation of the tumour near the renal vein may be so difficult that both tumour thrombectomy and wedge resection of the IVC should be performed to completely resect the tumour that has invaded the IVC wall around the renal vein orifice. Radical nephrectomy and excision of the tumour extending into the IVC, with IVC resection, are advisable in patients in whom the IVC is occluded by a tumour thrombus. In the present case, peeling off the thrombus from IVC did not compromise the vessel diameter nor required resection to achieve negative surgical margins.

The survival rate of renal PNET is poor despite the multidisciplinary approach for treatment and most patients die within 1 year of the diagnosis.¹² Depending on the thrombus extension level and IVC patency, surgical techniques should be selected, as they are, along with adjuvant chemotherapy and radiotherapy, the only reasonable chance for survival, and prognosis is poor despite multimodal treatment.¹³ Preoperative placement of temporary IVCF is desirable to prevent PE during the surgery. In this case, IVCF was placed after PE occurred in association with active tumoral bleeding that contraindicated anticoagulation and also to prevent recurrent PE during radical nephrectomy. Interestingly, even when left renal vein tumoral thrombectomy was performed initially, the recurrent extensive tumoral thrombus involving the IVC filter (located beneath the suprahepatic veins) warranted contralateral surgical intervention in order to achieve complete thrombus resection and IVC filter extraction. To the best of our knowledge, only one thoracoabdominal approach has been reported in a patient with an IVCF and tumoral thrombus, and this reported case was associated with renal cell carcinoma.¹⁴

In conclusion, PNET are rare neoplasms with aggressive behaviour and poor prognosis. Radical nephrectomy, renal vein and IVC thrombectomies are indicated, followed by chemotherapy. IVCF is recommended in patients with a history of PE prior to the surgery and this requires retrieval after surgical management to avoid complications related to this device as vessel perforation, migration and recurrent thrombotic events. In this particular case, a right thoracoabdominal surgical approach allowed successful removal of the IVCF and tumoral thrombus.

Figure 5 Following the removal of the filter and the tumoral thrombus, the inferior vena cava is closed primarily maintaining vessel patency (A). The extracted filter was carefully inspected for missing elements (B).

Figure 6 Microscopic view of the removed thrombus demonstrating positive CD99 on immunohistochemistry confirming the recurrent primary neuroectodermal renal tumour.

Learning points

- ▶ Primary neuroectodermal renal tumours (PNET) are rare neoplasms with aggressive behaviour and poor prognosis.
- ▶ Radical nephrectomy and inferior vena cava (IVC) thrombectomy are indicated followed by chemotherapy and radiotherapy.
- ▶ IVC filters are recommended prior to surgical resection in patients with a history of pulmonary embolism. Device retrieval is necessary after the procedure when it is safe to start anticoagulation if indicated.
- ▶ The right thoracoabdominal surgical approach allows optimal exposure of the retrohepatic IVC for thrombus removal.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Huang J, Zhang J, Dong B, *et al.* Primitive neuroectodermal tumor of the kidney: a single-institute series of six patients from China. *Urol Int* 2013;90:174–8.
- 2 Veroux M, Tallarita T, Pennisi M, *et al.* Late complication from a retrievable inferior vena cava filter with associated caval, aortic, and duodenal perforation: a case report. *J Vasc Surg* 2008;48:223–5.
- 3 Jibiki M, Iwai T, Inoue Y, *et al.* Surgical strategy for treating renal cell carcinoma with thrombus extending into the inferior vena cava. *J Vasc Surg* 2004;39:829–35.
- 4 Connolly PH, Balachandran VP, Trost D, *et al.* Open surgical inferior vena cava filter retrieval for caval perforation and a novel technique for minimal cavotomy filter extraction. *J Vasc Surg* 2012;56:256–9; discussion 259.
- 5 Aghili M, Rafiei E, Mojahed M, *et al.* Renal primitive neuroectodermal tumor: does age at diagnosis impact outcomes? *Rare Tumors* 2012;4:e15.
- 6 Folpe AL, Goldblum JR, Rubin BP, *et al.* Morphologic and immunophenotypic diversity in Ewing family tumors: a study of 66 genetically confirmed cases. *Am J Surg Pathol* 2005;29:1025–33.
- 7 Carter RL, al-Sams SZ, Corbett RP, *et al.* A comparative study of immunohistochemical staining for neuron-specific enolase, protein gene product 9.5 and S-100 protein in neuroblastoma, Ewing's sarcoma and other round cell tumours in children. *Histopathology* 1990;16:461–7.
- 8 Patel HJ, Williams DM, Upchurch GR Jr, *et al.* The challenge of associated intramural hematoma with endovascular repair for penetrating ulcers of the descending thoracic aorta. *J Vasc Surg* 2010;51:829–35.
- 9 Ghosh SK, Paul S, Anatomy of the retrohepatic segment of the inferior vena cava and the ostia venae hepaticae with its clinical significance. *Surg Radiol Anat* 2012;34:347–55.
- 10 Kearon C, Akl EA, Comerota AJ, *et al.* Antithrombotic therapy for VTE disease: Antithrombotic Therapy and Prevention of Thrombosis, 9th ed: American College of Chest Physicians Evidence-Based Clinical Practice Guidelines. *Chest* 2012;141(2 Suppl):e419S–94S.
- 11 Iliescu B, Haskal ZJ. Advanced techniques for removal of retrievable inferior vena cava filters. *Cardiovasc Intervent Radiol* 2012;35:741–50.
- 12 Patnaik N, Mishra K, Saini P, *et al.* Primitive neuroectodermal tumor of the kidney in a young male: case report and review of literature. *Urol Ann* 2015;7:236–9.
- 13 Ciancio G, Shirodkar SP, Soloway MS, *et al.* Renal carcinoma with supradiaphragmatic tumor thrombus: avoiding sternotomy and cardiopulmonary bypass. *Ann Thorac Surg* 2010;89:505–10.
- 14 Escudero JU, de Campos MR, López SC, *et al.* [Surgical treatment of the renal carcinoma with inferior vena cava thrombus and filter]. *Arch Esp Urol* 2008;61:730–3.

Copyright 2015 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow