

เนื้องอก Leiomyosarcoma ของเส้นเลือดดำใหญ่ในช่องท้อง : กรณีศึกษาการผ่าตัด เส้นเลือดดำใหญ่ในช่องท้องทดแทนด้วยเส้นเลือดเทียม Dacron

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Primary Leiomyosarcoma of the Inferior Vena Cava : A Case Report of Radical Surgery with Dacron Reconstruction

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หลักการและวัตถุประสงค์: ผู้ป่วยหญิงอายุ 33 ปี มีอาการปวดท้องซีกขวาเรื้อรังโดยไม่มีก้อนในช่องท้องหรือขาบวมเลยเป็นเวลา 1 ปี เอกซเรย์คอมพิวเตอร์ส่งสัยมะเร็งระดับ 2.9 ซม. ได้ทำการตรวจคลื่นแม่เหล็กไฟฟ้าเพิ่มเติม พบเนื้องอกที่ผนังของเส้นเลือดดำใหญ่

วิธีการรักษา: แผนการรักษาประกอบด้วย การผ่าตัดเอาเนื้องอกในเส้นเลือดดำใหญ่ในช่องท้องและตับส่วนที่ 1 ออก มีความจำเป็นต้องทดแทนเส้นเลือดดำใหญ่ด้วยเส้นเลือดเทียม Dacron ขนาด 20 มม. และเส้นเลือดดำของไตซีกซ้าย 12 มม.

ผลการศึกษา: ผลจากการใส่เส้นเลือดเทียม Dacron มีภาวะคงเปิดของเส้นเลือดเทียมดี

สรุป: การใช้เส้นเลือดเทียม Dacron เพื่อทดแทนเส้นเลือดดำใหญ่ในช่องท้องเป็นอีกทางเลือกหนึ่งที่ดี

คำสำคัญ: Leiomyosarcoma, เส้นเลือดดำใหญ่ในช่องท้อง, Dacron

Background and Objective: A 33 year-old woman present with one year right-sided abdominal pain without abdominal mass or leg edema, investigated with CT was suspected hepatocellular carcinoma 2.9 cm but MRI suspect leiomyosarcoma of IVC .

Methods: Treatment plans included radical resection with caudate lobe resection need 20 mm Dacron graft for IVC reconstruction and 12 mm for left renal vein re-implantation.

Results: The treatment resulted good patency of Dacron graft

Conclusion: Dacron graft vena caval replacement is considered as a good alternative graft for IVC reconstruction.

Keywords: Leiomyosarcoma, Inferior vena cava, Dacron

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Introduction

Primary tumors of the inferior vena cava (IVC) are rare, less than 300 cases reported in the literature, that leiomyosarcoma (LMS) representing the vast majority (95%)^{1,2}, usually present in late middle age women with abdominal discomfort due to an abdominal mass³. The

mainstay of treatment is surgical resection, if there is no evidence of metastasis, other treatment outcome is not quite clear⁴. We report pararenal vena cava leiomyosarcoma with emphasis on the surgical procedure and reconstruction of caval continuity with Dacron graft.

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Case report

A 33 year-old woman presented with 1 year right-sided abdominal pain without abdominal mass or leg edema was investigated with Computed Tomography revealed a 2.9 cm rim enhancing hypodense mass with rapid wash out in segment 1 of liver compress on IVC and portal vein, as shown in Fig 1. Another investigation using Magnetic Resonance Imaging (MRI) revealed a 3.1 cm lobulated intraluminal filling defect attach to medial wall of upper IVC compromised of lumen, as shown in Fig 2, Leiomyosarcoma of IVC is diagnosis.

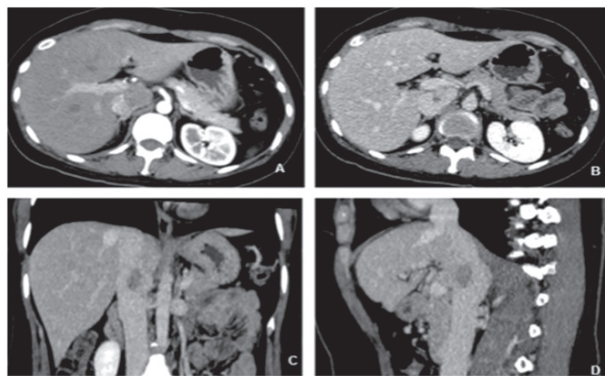


Fig 1 CT shows 2.7x2.9 cm rim enhancing hypodense mass with rapid wash out in segment 1 of the liver with compression on IVC and portal vein.

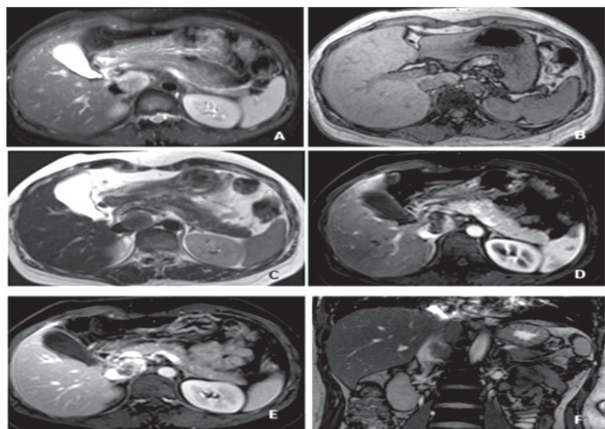


Fig 2 MRI shows 2.8x2.8x3.1 cm, lobulated intraluminal filling defect attach to medial wall of upper IVC.

The patient was scheduled for surgical resection, after mobilized left and right lobe liver, tumor was identified as Fig 3. Radical resection was performed, IVC reconstruction was needed with 20 mm Dacron, left

renal vein re-implanted with 12 mm Dacron as shown in Fig 4.

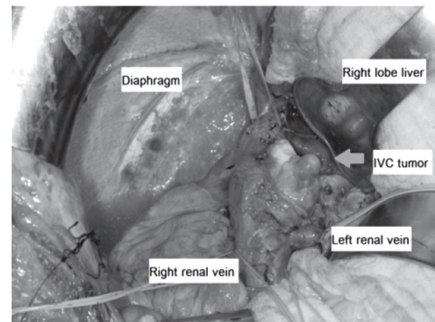


Fig 3 Intra-operative finding shows tumor at IVC after mobilized left and right lobe liver.

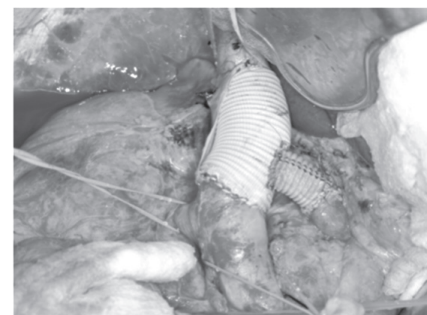


Fig 4 Intra-operative finding show IVC was reconstructed with 20 mm Dacron, left renal vein was re-implanted with 12 mm Dacron.

A well-circumscribed rubbery mass, measuring 4x3x3 cm, showing homogeneous cut surfaces in Fig 5 with microscopic finding show low grade leiomyosarcoma in Fig 6.

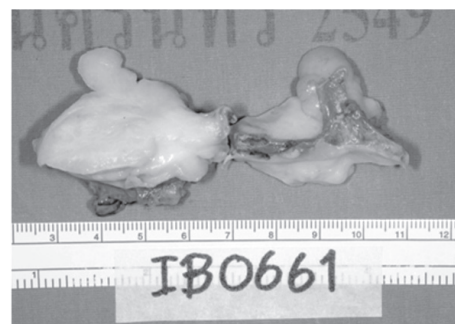


Fig 5 Gross pathology shows the tumor excision specimen was a well-circumscribed rubbery mass, measuring 4x3x3 cm, and homogenous cut surface.

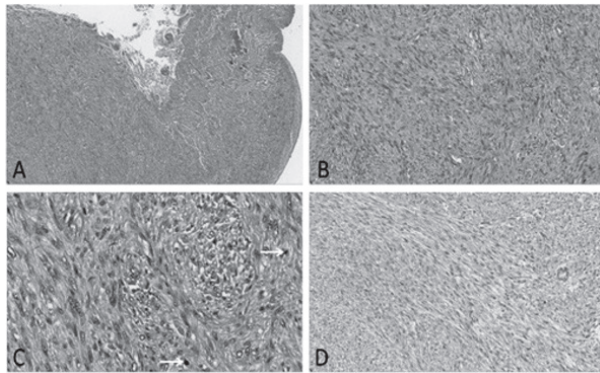


Fig 6 Microscopic finding shows low grade leiomyosarcoma, an indurated and extraluminal expansile mass at vascular wall (A), comprised of cellular spindle proliferation in fascicles (B) and area of pleomorphism with few mitosis (C). Tumor cells expressed membranous and cytoplasmic staining for smooth muscle actin antibody (D).

After an eventful recovery, the patient was discharged. Recurrent tumor at surgical bed after 18 months was followed up with good patency of Dacron graft as in Fig 7, afterward the patient passed away 30 months after surgery.

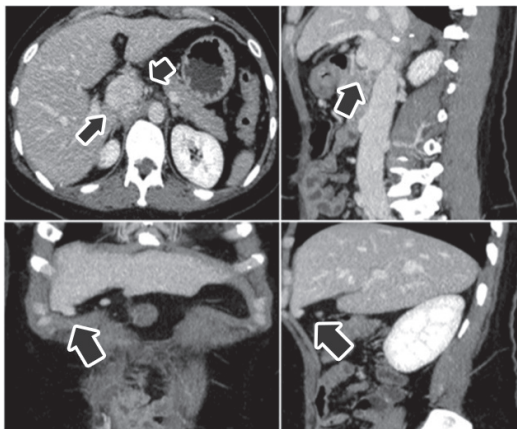


Fig 7 CT shows recurrent of tumor at surgical bed in 18 months follow up with patent of Dacron graft.

Discussion

Leiomyosarcoma is the most common primary malignancy of blood vessels, arise anywhere in the vascular system, most recorded cases are in the IVC⁵.

Perl first described it in 1871 and the first surgical resection was in 1928 by Mechior^{6,7}. Tumor growth within the IVC is intra-luminal in 5%, extra-luminal in 62%, and both intra- and extra-luminal in 33%⁸. Histopathological features are identical to those found in LMS of other sites. There is no standardized histological grading system for IVC tumors, some authors have recommended using the criteria for retroperitoneal LMSs, which is based on mitotic count⁵.

The presentation of LMS depends on its location along the IVC, which can be divided into three segments. Segment I (lower) is below the renal veins, involved in 36% of cases, presenting symptoms may include lower extremity edema, deep venous thrombosis, abdominal pain, and palpable mass. Segment II (middle) is from the hepatic veins to the renal veins, 44% of cases, symptoms may include abdominal pain, nephrotic syndrome, and renal hypertension. Segment III (upper) is from the right atrium to the hepatic veins, 20% of cases, present with weight loss, nausea, Budd-Chiari syndrome, and cardiac arrhythmias⁹⁻¹². Tumor of this patient was located in segment II which make operation more difficult and can cause many symptoms, but the patient got only abdominal pain and operation was performed without any complication.

The best modality to get tissue diagnosis is endoscopic biopsy under ultrasound guidance¹³, the principal aim of preoperative imaging assessment is to determine the local and distant extent of the disease using a combination of CT or MRI scanning, bone, and positron-emission tomography (PET) scans¹⁴⁻¹⁶. Ascending or retrograde cavography may be used to evaluate involvement of major braches¹⁷.

Aggressive radical resection remains the current treatment of choice, palliative resections may temporarily improve symptoms but do not offer long-term survival^{18,19}. Recurrence due to incomplete removal of tumor is common⁴. The surgical management of partial resections of the IVC is a matter of current debate and includes ligation, primary repair/cavoplasty, or replacement with a graft. Reconstruction of the IVC is not always required, because gradual occlusion of the IVC allows the development of venous collaterals.

However, when pararenal leiomyosarcoma of the IVC is present, reconstruction of the IVC and the renal vein is necessary to prevent transient or permanent renal dysfunction^{20,21}.

Historically, the prognosis of primary IVC leiomyosarcoma has been poor. In those with metastatic disease at presentation, survival is usually 3 - 6 months. In those who have an attempted curative resection, 5- and 10-year survival rates are 30- 53% and 7.4%, respectively^{3,4,10} with the principle determinant of survival being surgical resectability with tumor-free margins; the post resection recurrence rate is around 50%²².

Several technical considerations, when PTFE is used, some suggest that externally supported PTFE prevents collapse from respiratory compression and from surrounding structures, while others believe that it incorporates poorly and predisposes to entero-prosthetic fistula formation with the duodenum²³. Under-sizing of the PTFE has been recommended with the rationale that the resulting increase in blood flow velocity within the graft might reduce thrombotic risk²⁴. Others recommend a larger diameter graft, as PTFE tends to form a thick pseudo-intima that may result in obstruction²⁵. We used 20 mm Dacron graft without external support in IVC reconstruction and 12 mm Dacron graft for left renal re-implantation, after follow up for 5 years there is no compression, thrombosis or entero-prosthetic fistula formation. Compared with previous reports, this patient had comparable of survival and patency even different of graft.

Conclusion

In this report, the feasibility and safety of Dacron graft vena caval replacement is documented. Therefore, it can be considered a good alternative graft to reconstruction of IVC in the case of a rare primary leiomyosarcoma.

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