Primary leiomyosarcoma of inferior vena cava: Case report

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Abstract

An intramural venous leiomyosarcoma is a rare, malignant tumour arising from the smooth muscle cells of the vessel wall and the inferior vena cava (IVC) is the most common location. The middle part of IVC is most often affected, often involving the kidneys. There is a strong prediction for women. Clinical symptoms depend upon the size and location of the tumour. Diagnosis is often not made until advanced stage, as the symptoms are non-specific and they present late in the disease course. (Ind J Thorac Cardiovasc Surg 2008; 24: 261-263)

Key words: Magnetic resonance angiography, Kidney, Thoracotomy

Introduction

Tumours of the inferior vena cava (IVC) are rare and often malignant. Leiomyosarcoma is the most common primary vascular tumor, which originates from within the wall of the IVC. The true prevalence of these tumours is unknown because the slow growth of these tumour delays their detection. Early detection and surgical resection and reconstruction improves life expectancy.

Case report

A 32-years-old female presented with vague upper abdominal pain and magnetic resonance angiography showed a large mass arising from the intra-luminal part of IVC. Laparotomy was performed and the mass was found to arise from middle 1/3 of IVC encasing both the renal veins. Following frozen section diagnosis of primary leiomyosarcoma of IVC, the tumor was resected en-block including part of both renal veins. Poly tetra fluoro ethylene (PTFE) tube graft was used to re-establish IVC continuity with reimplantation of renal veins into the graft. Patient is healthy and asymptomatic even after 2 years.

A 45-year-old male presented with severe upper abdominal pain and on investigation for the same, Ultrasoundography (USG) abdomen revealed a mass arising from the IVC. Magnetic resonance angiography (MRA) revealed a mass arising from the lower 1/3 of IVC not involving the renal veins. Following frozen section diagnosis, the tumour was resected en-block and re-established IVC continuity using PTFE graft. Patient is doing well at 1-year post operatively.

A 35-years-old female presented with vague upper abdominal pain with worsening pain of 2 weeks duration. USG and computed tomography (CT) abdomen and MRA revealed a very large mass arising from infra hepatic portion of IVC extending below and involving both the renal veins. The tumour was resected after extensive dissection as the tumour was adhered to the liver, gall bladder and most of the bowel. Massive blood transfusion was required due to the highly vascular nature of the tumour. Both the renal veins were ligated as the tumour involved it. Since the segment of the IVC from which the tumour arose was thrombosed, proximal and distal ends of the IVC was ligated and over sewn without re-establishing IVC continuity. Postoperatively the patient deteriorated with massive fluid retention, hypotension and disseminated intravascular coagulation, irreversible brain damage and died 1 week postoperatively.

A 33-years-old female presented with history of vague upper abdominal pain of 2 months duration with...
history of vomiting since 15 days and abdominal distension and worsening abdominal pain since 3 days. Liver function tests were deranged. MRA of abdomen showed a vague mass arising from the suprarenal inferior vena cava and extending up to the superior vena cava with hepatic vein thrombosis probably inferior vena caval tumour and budd chiari syndrome. Through a midline laparotomy and right thoracotomy incision the tumour was freed from the liver bed with transection of the right hepatic vein and dividing the tumour from the superior vena cava. PTFE graft was used as a conduit to create the right hepatic vein from the opening in the liver bed and anastomosed to the cut end of the superior vena cava and flow re-established. The distal end of inferior vena cava was suture ligated. Patient is asymptomatic on follow up.

Discussion

Tumours of the inferior vena cava (IVC) are rare and often malignant. Leiomyosarcoma is the most common primary vascular tumor, which originates from within the wall of the IVC. The true prevalence of these tumours is unknown because the slow growth of these tumour delays their detection. Primary caval tumours may arise from any segment of the vena cava. Leiomyosarcoma affects the middle segment (from the renal to hepatic veins) in 42%, the lower segment (infra renal IVC) in 34% and the upper segment (from hepatic veins to right atrium) in 24%. Symptoms vary according to the portion of the IVC involved.

Pathology

These tumors are usually polypoid or nodular in pathologic appearances. They are firmly attached to the vessel of origin and exhibit less intra tumour haemorrhage and necrosis than other retroperitoneal sarcomas. The most common growth pattern is intra luminal, but tumour can extend through the adventitial surface of the vein wall and involve adjacent structures because the vein wall is thin. This characteristic makes them difficult to differentiate from other retroperitoneal sarcomas. Such extra luminal growth may occur early in the course of the disease, which would account for the presence of distant metastasis in almost half of patients at the time of diagnosis. Metastasis to the lung, liver, kidney, bone, pleura or chest wall occur hematogenously or by lymphatic spread. If the lesion remains untreated, survival is measured in months, with the patient dying of complications from loco regional growth or metastatic disease.

Microscopically, grading of the malignancy can be difficult and extensive sampling of the tumour is necessary. Of the criteria used to determine malignancy the most important determinant of malignancy in leiomyosarcoma is mitotic activity.

Clinical presentation

Primary leiomyosarcoma of the vena cava is much more prevalent in woman than men in the ratio of 1:5 (M: F). Clinical presentation of patients with primary caval tumors is related to symptoms or sings from metastatic disease or from obstruction of the vena cava. Rarely these tumors are detected at an early stage as incidental findings during evaluation of non-specific symptoms such as back or abdominal pain. Symptoms and resectibility depend on the location and extent of the tumour. Patients with leiomyosarcoma of the IVC usually present with non-specific complaints such as malaise, weight loss, backache or abdominal pain.

Leiomyosarcoma of IVC was usually diagnosed during laparotomy or autopsy. With the advent of radiologic imaging studies like USG, CT, magnetic resonance imaging (MRI) and venocavography it is possible to diagnose these tumours preoperatively and at an earlier stage (Fig. 1)

The final diagnosis can be confirmed by fine needle aspiration or true cut needle biopsy at laparotomy (Fig. 4). Except for germ cell tumors most primary and secondary tumors of IVC do not respond to chemotherapy or radiotherapy and hence preparative tissue sampling has little role in treatment (Fig. 2).

Surgical resectibility is highly dependent on the location of the tumour. Complete resection of the