A 53-year-old Thai female presented with progressive dyspnea for 10 days. According to The New York Heart Association (NYHA), her status had changed from class I to class II–III. She had no chest pain, fever, orthopnea, or paroxysmal nocturnal dyspnea. The physical examination was normal. There was no hepatosplenomegaly or signs of chronic liver stigmata. The laboratory test showed abnormal liver enzymes. The patient was referred for further investigation by ultrasound and computed tomography (CT) scan of abdomen.

The ultrasound of abdomen revealed heterogeneous hyperechoic lesion occupying in enlarged inferior vena cava (IVC) (Figure 1 and 2). There was no internal vascularity within this lesion. The abdominal CT showed heterogeneous isodensity lesion in intrahepatic IVC (Figure 3). After contrast medium injection this lesion had inhomogeneous enhancement (Figure 4 and 5). There was a small non-enhancing thrombus inferior to aforementioned lesion (Figure 5). The liver also had inhomogeneous enhancement as a result of hepatic venous occlusion (Figure 4 and 5).

The biopsy was subsequently performed. The pathology revealed leiomyosarcoma of the IVC.

Primary tumor of IVC is rare. Leiomyosarcoma is the most common primary malignant tumor of the IVC which the majority affecting women in 5th to 6th decades of life (1). In 1996, The International Registry of Inferior Vena Cava Leiomyosarcoma published the largest series of
IVC leiomyosarcoma in 218 patients, most of the patients had insidious onset of nonspecific symptoms; for example, abdominal pain, weight loss, anorexia, fever, nausea, vomiting, and superficial abdominal vein distension (2, 3). Thus, the diagnosis of IVC leiomyosarcoma is often made by imaging findings such as ultrasound, CT scan, or magnetic resonance imaging (MRI).

Leiomyosarcoma of IVC arises from smooth muscle tissue within the vascular wall. The tumor shows polypoid or nodular masses abutting with vascular wall. Three major growth patterns were reported: completely intraluminal (6%), completely extraluminal (62%), and intra- and extraluminal components (32%) (4). Most of the patients have tumor located in middle (between hepatic to renal veins: 50.8%) and lower segments (infrarenal portion: 44.2%) of the IVC. Only 4.2% of the patients have tumor in upper segment of IVC (between hepatic veins to right atrium) (1). The risk of death was increased in patient with tumor involving upper segment, lower limb edema, hepatic venous occlusion syndrome, intraluminal tumor growth, and IVC occlusion (2).

The ultrasound findings of IVC leiomyosarcoma are dilatation of IVC associated with echogenic mass. Color Doppler ultrasound can demonstrate filling defect in IVC or intratumoral vascularity. However, the limitation of ultrasound is operator dependent. CT scan shows hypodensity mass with intra- or extraluminal components and inhomogeneous enhancement (5). Necrosis or cystic degeneration is not uncommon. MRI reveals intermediate signal intensity on T1 weighted image (WI) and hypersignal intensity on T2WI with enhancement after gadolinium injection (5). In intraluminal type, the intraluminal thrombus can occur. Differential diagnosis between tumor and thrombus is made by using contrast medium injection. The tumor shows enhancement whereas thrombus has no enhancement (6).
In conclusion, IVC leiomyosarcoma is the most common primary malignant tumor of the IVC. This tumor can involve intraluminal, extraluminal, or intra- and extraluminal compartments. Radiologic imaging plays the major role in making the diagnosis.

References