Interrupted inferior vena cava: high-risk anatomy for right thoracotomy

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Abstract

Interrupted inferior vena cava (IVC) is a rare developmental defect characterised by azygos continuation following failure of fusion of one or more of the component parts of the embryological IVC and occurring in approximately one in 5000 of the general population. It is usually an isolated finding and generally asymptomatic. We present a case of non-small cell lung cancer requiring right pneumonectomy in a patient with an interrupted IVC with azygos continuation.

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1. Case report

A 69-year-old female non-smoker presented with a three-months’ history of persistent cough. Physical examination was unremarkable. Chest radiograph showed a large right upper zone mass extending to the hilum. Computerized tomography (CT) of the chest confirmed the presence of a 5-cm right upper lobe mass in close proximity and possibly invasion into a large azygos vein (AZV) (Fig. 1a).

Pulmonary function testing revealed a forced expiratory volume in 1s (FEV₁) of 1.6 (96% of predicted) and a carbon monoxide transfer factor (TLCO) of 89% of predicted. Positron emission tomography (PET) scanning showed high avidity in the right upper lobe mass and increased avidity in a hilar lymph node. Clinical staging was denoted cT₂N₀M₀.

At staging CT, the patient was found incidentally to have abnormal venous anatomy, consisting of interrupted inferior vena cava (IVC) with azygos continuation. The IVC was in continuity with and drained into a grossly dilated azygos system which in turn drained into the superior vena cava (SVC). The hepatic venous system drained separately and directly into the right atrium (Fig. 1b). Venography confirmed IVC continuity with a large AZV (Fig. 2a).

The patient underwent rigid bronchoscopy, right thoracotomy (Fig. 2b) and pneumonectomy. At rigid bronchoscopy tumour was seen abutting the right upper lobe bronchus. Following right posterior thoracotomy the tumour was noted to be crossing the fissure into the middle and lower lobes.

2. Discussion

Interruption of the IVC is rare, with an incidence of approximately 1:5000 of the population, based on prenatal ultrasonic screening. In 90% of cases, it occurs as an isolated anomaly, although it may be associated with cardiac or splenic abnormalities [1]. When occurring in isolation it is usually asymptomatic with no clinical signs. However, haematochezia has been reported in a case of infrarenal interrupted IVC associated with diffuse systemic and caval-portal venous collateralization [2].

Interrupted IVC results from failure of fusion of the component parts of the embryological IVC and may occur at any level. The IVC is composed of four segments: hepatic, prerenal, renal and postrenal. These segments occur from the formation, fusion and regression of paired cardinal veins. The hepatic segment is formed from the hepatic veins, the prerenal segment from the subcardinal vein, the postrenal segment from the supracardinal vein whilst the renal segment is formed from the subcardinal and supracardinal anastomosis. The azygos system is formed from the supracardinal veins. The hepatic segment is sub-
the normal IVC casts a linear shadow representing it is posterior border, which when absent is also a useful sign indicating azygos continuation of the IVC [4]. However, in the modern imaging era it is probably most commonly noted at CT imaging.

In the presence of normal venous anatomy, sacrifice of the azygos is safe and well-tolerated when required for complete resection due to tumour invasion or necessitated by procedures, such as carinal resection. However, in the present case, where the AZV drains the lower half of the body, invasion of tumour into the AZV would preclude such sacrifice. Indeed, in the preCT era Effler et al. described ligation of the azygos in the presence of interrupted IVC during attempted resection for bronchogenic carcinoma with fatal consequences [5]. Since the cranial extension of the hepatic segment still drains directly into the right atrium in the presence of interrupted IVC, this may be mistaken for the IVC and there may be failure to recognise the condition. Even in the modern era of CT scanning, interrupted IVC may be misdiagnosed as paraoesophageal lymph nodes [6], retroperitoneal neoplasm [7] or aortic dissection [8].

In the present case, preoperative CT suggested invasion of the AZV by tumour which would have precluded curative resection. In this event, exploratory thoracotomy alone, followed by chemoradiotherapy was planned. This case demonstrates that the potentially fatal consequences of what is normally a safe manoeuvre may be avoided by preoperative recognition of interrupted IVC.

References

