# Abdominal neuroblastoma in a child with inferior vena cava anomaly

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**ABSTRACT** Congenital anomalies of the inferior vena cava are rare. Such anomalies pose great challenges to the surgeon in neuroblastoma surgery, especially when unrecognised preoperatively. We report the first case of an abdominal neuroblastoma detected in a child with a developmental anomaly of the left-sided inferior vena cava. The patient underwent surgical resection after good response to preoperative chemotherapy.

Keywords: child, left inferior vena cava, neuroblastoma, surgery

## INTRODUCTION

Congenital anomalies of the inferior vena cava (IVC) are rare.[1] Such anomalies pose great challenges to the surgeon during neuroblastoma surgery, especially when unrecognised preoperatively. Other than their similarity to para-aortic lymphadenopathy on non-contrast cross-sectional computed tomography (CT) images, these anomalies often pose difficulties during retroperitoneal tumour dissection and increase the risk of vascular injury. Herein, we report the first case of an abdominal neuroblastoma detected in a child with a developmental anomaly of a left-sided IVC.

## CASE REPORT

A 3-year-old Chinese boy was referred to our centre for surgical resection of an abdominal neuroblastoma after initial diagnosis and preoperative chemotherapy at a foreign facility. He was diagnosed with stage III abdominal neuroblastoma three months earlier, when he presented with abdominal pain and fever. At diagnosis, CT with intravenous contrast showed a large retroperitoneal tumour that encased the IVC, aorta and its branches (Fig. 1). Anomaly of the IVC was initially unrecognised because of the tumour's compression of the vessels and the venous enhancement's resemblance to tumour calcifications. An ultrasonography-guided needle biopsy was performed, and it confirmed the presence of a neuroblastoma with absence of _MYCN_ amplification. There was no evidence of metastases. After three cycles of chemotherapy, preoperative CT showed a marked reduction in the size of the abdominal tumour, with features consistent with a left-sided IVC (Fig. 2).

During laparotomy, a right adrenal tumour was found adjacent to a normal-sized suprarenal IVC and right renal vein. The suprarenal IVC tapered inferiorly into a 3-mm diameter vein that was completely encased by a para-aortic mass on the right side of the aorta. Further distal dissection confirmed it to be the right testicular vein. A robust left IVC of 10-mm
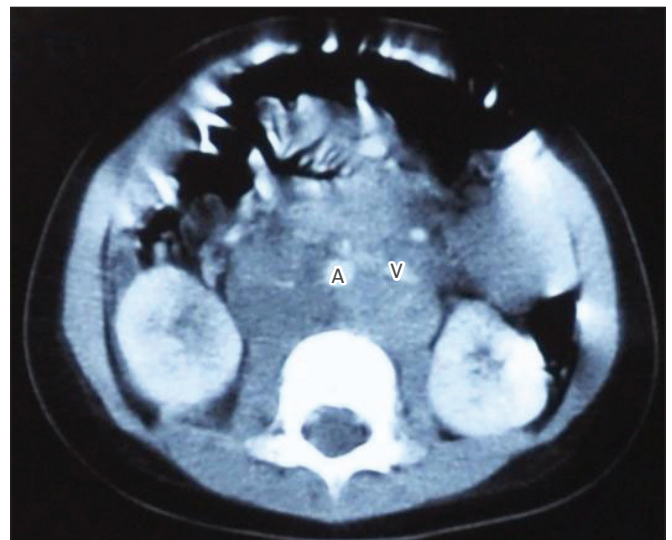


**Fig. 1** Cross-sectional CT taken at diagnosis. CT image shows the aorta (A) and left inferior vena cava (V) encased by the large para-aortic tumour.
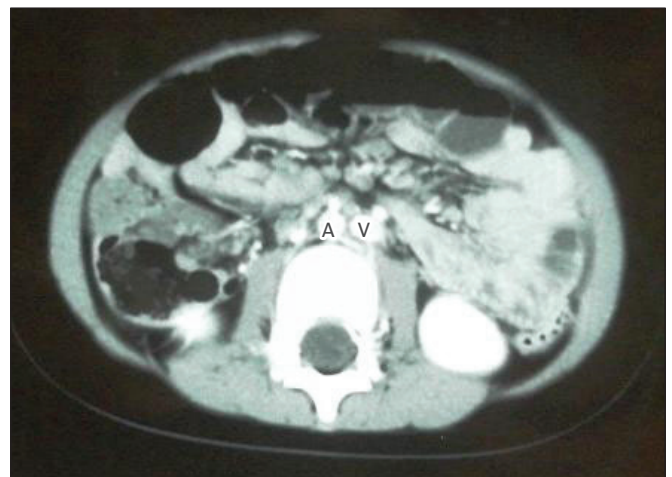


**Fig. 2** Cross-sectional CT taken after preoperative chemotherapy. CT image shows a significantly reduced tumour size, with the diameter of the left inferior vena cava (V) comparable to that of the aorta (A). Enhancement on the right of the aorta represents tumour calcification.

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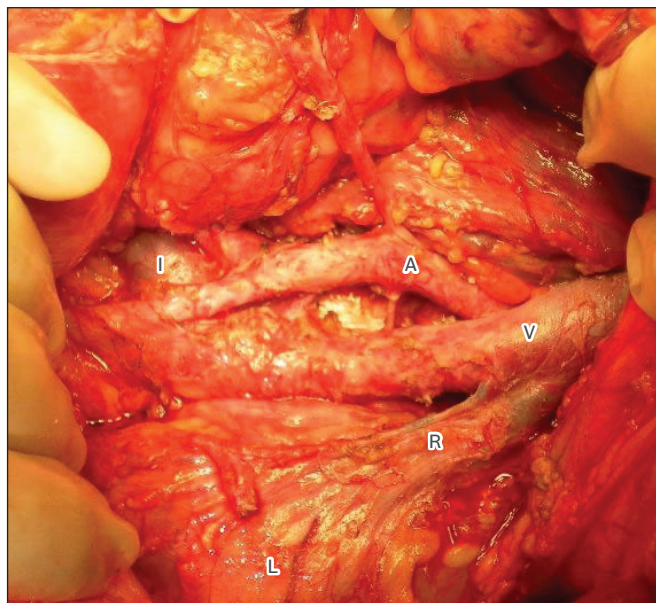
**Fig. 3** Intraoperative photograph of the left side of the abdomen shows the left inferior vena cava (V) receiving the right iliac vein (I) posterior to the aorta (A). The left renal vein (R) is draining the left kidney (L), and the inferior mesenteric artery branches ventrally from the aorta (A).

diameter was identified on the left side of the aorta, receiving venous drainage from both the lower extremities and draining into the left renal vein (Fig. 3). The left renal vein crossed anterior to the aorta before draining into the suprarenal IVC. A diagnosis of congenital anomaly of the left-sided IVC was made. Gross total resection of the neuroblastoma was successfully completed without injury to the blood vessels.

Histopathology confirmed poorly differentiated neuroblastoma without *MYCN* amplification and chromosome 1p deletion, although chromosome 17q was gained. Postoperative recovery was uneventful and the patient returned to his referring oncologist to continue with chemotherapy.

## DISCUSSION

The principles of neuroblastoma surgery include recognition of the normal vascular anatomy, preservation of vital structures and safe tumour resection. These principles enable good tumour clearance while minimising the morbidity of surgery. Variations in vascular anatomy may be unexpected and challenging. Previous case studies have reported the association of IVC anomalies with Wilms' tumour[2] and ganglioneuroma[3] in children. However, this is the first case report of an abdominal neuroblastoma found in a child with a left IVC anomaly.

Developmental anomaly of the IVC is rare. The embryogenic process of the IVC begins from the sixth week of gestation and is completed by the tenth week.[1] The IVC is formed from the continuous appearance and regression of three pairs of embryonic veins. The posterior cardinal veins appear first on the posterior aspect of the embryo and then regress (except for their distal aspects, which become the iliac bifurcation). The subcardinal veins then appear anterior and medial to the posterior cardinal veins. The right subcardinal vein remains to form the suprarenal IVC, while the left subcardinal vein completely regresses. Subsequently, the supracardinal veins appear dorsal to the subcardinal veins. Under normal circumstances, the left supracardinal vein then regresses, while the right supracardinal vein forms the infrarenal IVC.

Anomalies of the IVC occur when the normal process of appearance and regression of these embryonic veins is disrupted. Although the pathogenesis of anomalies of IVC is unclear, some postulations regarding the mechanism of venous regression include embryological venous dysgenesis,[4] intrauterine venous thrombosis that leads to vascular atrophy,[4] and coalescence of multiple venous anastomoses.[1] Interestingly, our patient's neuroblastoma, a common foetal malignancy,[5] was found at the site of the regressed right supracardinal vein. This was most likely a coincidental association rather than the aetiology of right supracardinal venous regression. The developmental process of the IVC would have completed prior to the end of the first trimester of pregnancy, before the development of the neuroblastoma. Thus far, there has been no evidence of neuroblastoma development in the first trimester of pregnancy.[6-8]

Traditionally, left IVC and double IVC have been described as two separate anatomical entities. The left IVC entity results from the regression of the right supracardinal vein with the persistence of the left supracardinal vein.[1] Its prevalence is reported to be 0.2%–0.5%.[1] The left IVC joins the left renal vein, which crosses anterior to the aorta, uniting with the right renal vein to form a right-sided prerenal IVC. Typically, the drainage of the right lower extremity would converge into the left IVC. Meanwhile, the double IVC, which has a prevalence of 0.2%–3%,[1] results from the failure of regression of the left supracardinal vein.[1,9] The coexisting left IVC typically ends at the left renal vein, which crosses anterior to the aorta in a normal fashion to join the right IVC. Drainage of the lower extremities is usually via the IVC of the respective sides. A distinction between the aforementioned entities in preoperative imaging is important so as to avoid unexpected vascular injuries.

CT and magnetic resonance imaging are common preoperative imaging techniques used for the planning of neuroblastoma surgery. The former is usually preferred in most centres due to its accessibility, surgeons' familiarity and the obviated need for anaesthesia. However, neuroblastoma often develops calcifications, which may confound the interpretation of intravascular contrast on cross-sectional CT images. In addition, the neoplasm's ability to compress, distort and displace critical vascular structures should also be appreciated. Therefore, precontrast studies, and sagittal and coronal sections on CT or magnetic resonance imaging should be obtained so as to better illustrate the vascular anatomy.

In conclusion, major vascular anomalies in the retroperitoneum should be borne in mind while performing

neuroblastoma surgery, so as to avoid debilitating vascular injuries. Appropriate preoperative imaging techniques are crucial for surgical planning.

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