

Available online at [www.sciencedirect.com](http://www.sciencedirect.com)

journal homepage: [www.elsevier.com/locate/radcr](http://www.elsevier.com/locate/radcr)

## Case Report

# Circumaortic right renal vein with multiple vascular anomalies

Marjeta Tanka<sup>a,\*</sup>, Fjorda Tuka<sup>b</sup>, Erjona Abazaj<sup>c</sup>

<sup>a</sup>Radiologist at University Hospital Center "Mother Teresa", Tirana 1001, Albania

<sup>b</sup>Radiologist at American Hospital, Tirana 1001, Albania

<sup>c</sup>Parasitologist at Institute of Public Health, Tirana 1001, Albania

### ARTICLE INFO

#### Article history:

Received 5 March 2018

Revised 27 April 2018

Accepted 3 May 2018

#### Keywords:

Vascular anomaly

Embryology

Cardiac catheterization

### ABSTRACT

Circumaortic right renal vein is an extremely rare finding and to our knowledge only 1 case has been reported in the literature so far. Its rareness, in contrast to left renal vein anomalies, is thought to be due to a relatively simple embryologic development of right renal vein compared with left renal vein. On the other hand, association of Circumaortic right renal vein with inferior vena cava agenesis and aortic coarctation is an extremely rare occurrence. Our aim is to introduce a case of Circumaortic right renal vein in a 3-month-old child with inferior vena cava agenesis and aortic coarctation. Discussion on the underlying embryology of Circumaortic right renal vein, its clinical importance and the association with other vascular anomalies, will be on our focus as well. Precise understanding of renal vein anomalies is important when planning retroperitoneal surgery or interventional vascular procedures. Awareness of such anomaly implies crucial knowledge for radiologists who should include it in the medical reports to aid future patient's management.

© 2018 The Authors. Published by Elsevier Inc. on behalf of University of Washington.

This is an open access article under the CC BY-NC-ND license.

(<http://creativecommons.org/licenses/by-nc-nd/4.0/>)

## Case report

A 3-month-old girl, with no clinical symptoms, presented at our Radiology department to perform a chest contrast-enhanced computed tomography (CECT) scan, referred by the cardiac pediatrician.

Previous medical history included the diagnoses of aortic coarctation immediately after birth with a subsequent surgical repair on her sixth day of life.

A detailed evaluation of the thorax great vessels was requested to exclude any possible late complication. The examination was performed through a 4-detector row CT scanner (Siemens Somatom Emotion Duo) with administration of 12 ml contrast medium (ultravist with a concentration of 300 mg/dl) at an injection speed of 2.5 ml/s.

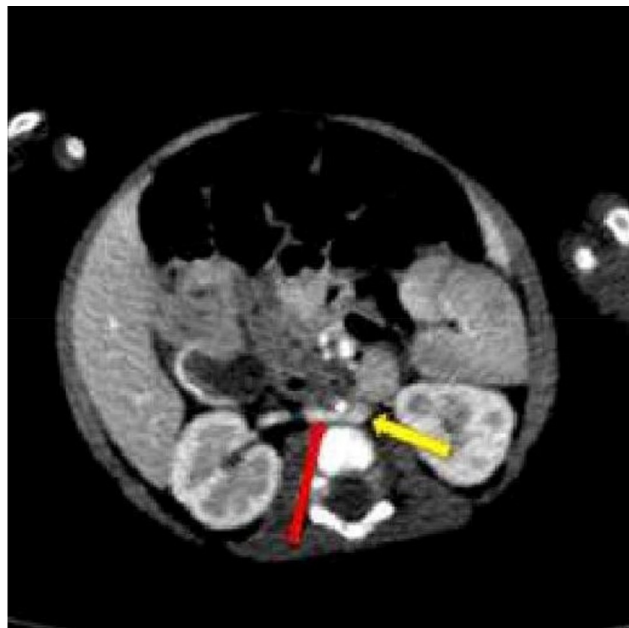
Two-millimeter thick sections and 3-dimensional multiplanar reconstructions were obtained.

Contrast-enhanced CT examination revealed a postductal short stenotic segment of the aorta.

Later cardiac catheterization-confirmed recoarctation with the presence of a residual stenotic segment at the level of anastomosis and subsequent balloon dilatation was performed.

\* Corresponding author.

E-mail address: [marjeta.tanka@yahoo.com](mailto:marjeta.tanka@yahoo.com) (M. Tanka).



**Fig. 1** – Axial computed tomography-image shows retroaortic branch (red arrow) of Circumaortic right renal vein passing posterior to the aorta and draining into hemiazygos vein (yellow arrow). (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

To better evaluate the intraabdominal segment of the aorta, a few slices of the upper abdomen were obtained up to the inferior pole of both kidneys.

Incidental finding of Circumaortic right renal vein (CaRRV) was noted (which had no relation with the previous coarctation treatment or recurrence). On the other hand, a vascular structure positioned on the left of the aorta was observed following its course upward in the thorax cavity and passing on the left of the thorax spine suggesting the presence of a dilated hemiazygos vein (HV) emptying into the left brachiocephalic vein, which after receiving the left subclavian and left jugular veins drained into the normal positioned superior vena cava.

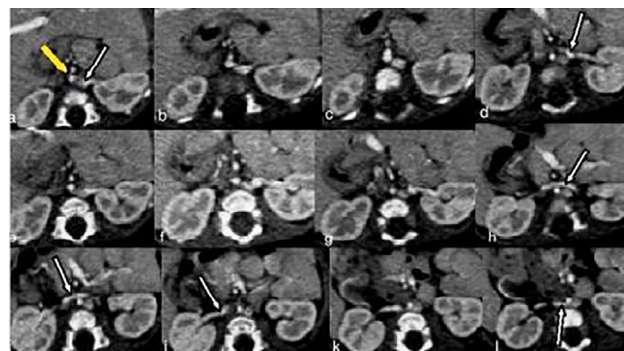
The right renal vein (RRV) at the parahilar region was divided into a longer anterior branch, which passed anterior to the aorta and HV, joined the normal left renal vein (LRV) following upward in a short 5 mm common trunk to drain on the lateral aspect of HV. While the posterior branch passed between the aorta and the vertebral body draining into the posterior part of the HV. The anterior branch drained at a more superior level than the posterior branch. Agenesis of the hepatic, suprarenal and renal segments of inferior vena cava (IVC) were also evident, with the suprahepatic veins draining directly into the right atrium. Evaluation through few slices obtained inferiorly to the renal veins showed absence of an infrarenal segment of IVC suggesting an IVC atresia (Figs. 1–3).

## Discussion

A number of anomalies relating the IVC and renal veins are well recognized and can only be explained through embry-



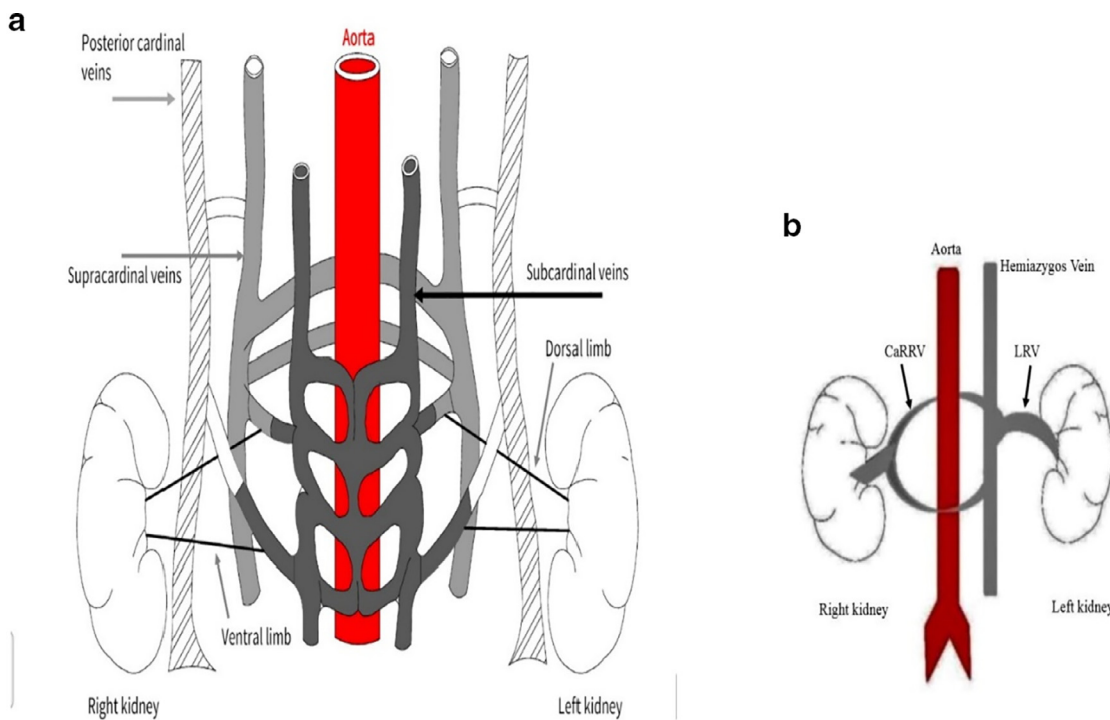
**Fig. 2** – Image showing anterior branch of Circumaortic right renal vein passing anterior to the aorta.



**Fig. 3** – Contiguous 2-mm thick computed tomography sections presented from cranial to caudal show the vessels anomaly (j–l). (a) White arrow—hemiazygos vein, orange arrow—aorta, (d) arrow points to the level where the anterior branch of Circumaortic right renal vein joins the left renal vein. (h–l) Contiguous images show the main right renal vein and its anterior and posterior branches composing Circumaortic right renal vein. (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

ologic basis [2,3]. Coexistence of more than 1 anomaly may appear. Two congenital vein anomalies are observed in our patient: agenesis of IVC and CaRRV.

The embryologic development of renal veins is part of the complex process of IVC embryogenesis, which involves an extensive network of anastomoses between 3 pairs of cardinal veins undergoing a regulated process of appearance and regression until the normal IVC and renal veins are formed [1].



**Fig. 4.** – (a) Schematic drawing shows the position of the 3 pairs of cardinal veins relative to the aorta during inferior vena cava and renal veins embryogenesis. (b) Schematic drawing of our patient's renal vein anomaly with anterior branch of Circumaortic right renal vein joining the left renal vein before draining at the lateral part of hemiazygos vein. The posterior branch of Circumaortic right renal vein drains at the posterior part of Hemiazygos vein at a lower level relative to the anterior branch.

Anomalies of the IVC and renal veins derive from an aberration of regression or abnormal persistence of these embryonic veins. The normal renal veins develop in a complex vascular environment composed by the so-called renal collar. Specifically this collar is composed ventrally by the inter-subcardinal anastomoses and small parts of the right and left subposterior anastomoses, laterally by anastomoses between supra and subcardinal veins while the inter-supracardinal anastomoses complete the renal collar dorsally (Fig. 4). Initially each kidney is connected with the renal collar through a pair of veins composed of a dorsal and ventral limb. Later on, regression of the dorsal limb and posterior branch of the renal collar will give rise to the normal renal vein [4].

Imaging findings in our patient showed a single RRV that left the hilum and was further divided into 2 branches, which encircled the aorta before emptying into a left sided HV. Furthermore, the suprarenal and renal segments of IVC were absent and no vascular structure was seen inferior to the RRV suggesting the atresia of IVC.

Considering the embryologic mechanism, we assume that in our patient circumaortic course of RRV might be explained by persistence of the inter-subcardinal anastomoses, inter-supracardinal anastomoses, and the right sub-supracardinal anastomoses, even though, we have found no actual report that gives an explanation of the embry-

ologic mechanism concerning our patient's whole vascular anomaly.

To our best knowledge, CaRRV has been reported only once in the literature [5]; however, a CaRRV draining into HV is an extremely rare finding which has not been reported so far [2].

Further studies should be performed to explain the exact embryologic mechanism of such anomaly.

In general, renal vein anomalies are clinically silent and most commonly discovered as incidental finding after routine examinations for other reasons or in autopsy.

When present, these anomalies hold an important clinical, surgical, and therapeutic implication. CaRRV has significant clinical importance when planning a nephrectomy especially in this era of renal transplantations we are experiencing recently.

Careful evaluation and awareness of renal vein anatomy is crucial prior to planning a retroperitoneal surgery especially when using laparoscopic approach since the narrow window of view may make it difficult to appreciate aberrant vessels. Familiarity with such anomalies remains of specific significance even to radiologists when staging for tumor lesions as abnormal renal veins may be misdiagnosed as lymphadenopathies [6].

Rarely, the posterior limb of CaRRV may be compressed by the aorta giving rise to the nutcracker syndrome.

On the other hand, a dilated thoracic segment of HV may be misdiagnosed as a mediastinal mass, which may lead to disastrous complications if transthoracic biopsy is performed.

In summary, CaRRV is an uncommon developmental defect in contrast to LRV anomalies probably due to a relatively simple embryologic development of RRV compared with LRV. Association of CaRRV with IVC agenesis and aortic coarctation is an extremely rare occurrence. Precise knowledge of renal vein anomalies gains significant importance especially when planning retroperitoneal surgery or interventional vascular procedures [7]. Awareness of this anomaly is crucial to the radiologist and should be included in the medical reports to aid future patient management.

---

### Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:[10.1016/j.radcr.2018.05.001](https://doi.org/10.1016/j.radcr.2018.05.001).

### REFERENCES

---

- [1] Sadler TW. *Langman's medical embryology*. 9th ed. Philadelphia: Lippincott Williams & Wilkins; 2004. p. 261–2.
- [2] Mayo J, Gray R, St Louis E, Grosman H, McLoughlin M, Wise D. Anomalies of the inferior vena cava. *AJR Am J Roentgenol* 1983;140:339–45.
- [3] Bass JE, Redwine MD, Kramer LA, Huynh PT, Harris JH Jr. Spectrum of congenital anomalies of the inferior vena cava: cross-sectional imaging findings. *Radiographics* 2000. doi:[10.1148/radiographics.20.3.g00ma09639](https://doi.org/10.1148/radiographics.20.3.g00ma09639).
- [4] Moore KL, Persaud TV. *The developing human. clinically oriented embryology*. 8th ed. London: Saunders Elsevier; 2008. 285–236.
- [5] Kim M-K, Ku YM, Chun CW, Lee SL. MDCT findings of right circum-aortic renal vein with ectopic kidney. *Korean J Radiol* 2013. doi:[10.3348/kjr.2013.14.5.786](https://doi.org/10.3348/kjr.2013.14.5.786).
- [6] Kleberg I, Wajsman Z. Duplicated inferior vena cava simulating retroperitoneal lymphadenopathy in a patient with embryonal cell carcinoma of the testicle. *J Urol* 1986;136:678–9.
- [7] Babaian RJ, Johnson DE. Major venous anomalies complicating retroperitoneal surgery. *South Med J* 1979;72:1254–8.