ANNALI ITALIANI DI CHIRURGIA

Digital Edition
e-publish on-line
ISSN 2239-253X
Direttore Nicola Picardi

Multimodality imaging of epithelioid haemangioendothelioma of the inferior vena cava

Ann Ital Chir, 2023; 12 - Sept. 4
pii: S2239253X23038604
Online Epub

Emanuele Casciani*, Roberto Meniconi**, Elisabetta Polettini*, Giuseppe Maria Ettore**, Gianfranco Gualdi*

**Reparto di Diagnostica per immagini, Clinica PIO XI, Roma, Italia*
***Dipartimento Interaziendale Trapianti A.O. San Camillo-Forlanini, Roma, Italia*

Multimodality imaging of epithelioid haemangiothelioma of the inferior vena cava.

Primary epithelioid haemangioendotheiomas (EHE) of the inferior vena cava (IVC) are extremely rare malignant tumours of vascular origin. Only about 40 case of vascular EHEs are described in the literature, and only fourorigin by IVC (1-4) and two by superior cava vein (5,6). Only surgical therapy for EHE has proven beneficial.It is often difficult to make the right diagnosis with imaging before histhological examination. Imaging, usually, help to surgical planning, but in our case helps to make the right diagnosis. We present a rare case of a patient with an IVC EHE, studied with Computed Tomography Angiography (CTA), Magnetic Resonance (MR), and Positron EmissionTomography Computed Tomography with fluorine 18 fluorodeoxyglucose (18F-FDG PET/CT).

KEY WORDS: CT, Diagnosis, Retroperitoneal Tumor

Case report

A 46-year-old woman presented to our hospital due to back pain and abdominal discomfort. Her medical and family history revealed no other major medical problems. On examination, she appeared healthy and her abdomen was soft and nontender; only moderate swelling in her feet was present.
A no contrast MR scan of the abdomen originally performed at an outside institution demonstrated a filling defect in the ICV just superior to the insertion of the renal veins, reported as thrombosis without common iliac vein thrombosis. It was hyperintense on T2-weighetd, hypointense on T1-weghted sequences, without remarkable signal intensity on diffusion weighted imaging (DWI).

A triple-phase Multidetector CTA of the abdomen was performed. Para-sagittal multiplanar reconstruction (MPR) from arterial and venous phase CTA (Figs. 1a, 1b) showed an intraluminal non-enhancing tumor (20x22x25 mm) with peduncolated hypervascular structure (22x3 mm) attached to the wall of the IVC.
A 18F-FDG PET/CT revealed no uptake by the mass.
A midline laparotomy was performed. The right colon and the liver were totally mobilized exposing the retrohepatic inferior vena cava up to the ilicac vein confluence. The suprarenal IVC was dissected and controlled. The renal veins were controlled in a similar fashion and taped bilaterally, and the tumor was clearly palpable in the IVC superiorly to the confluence of the renal veins (Fig. 2). After clamping the IVC and the renal veins anterior cavotomy was performed at the level of the renal veins. The tumor was identified and resected with its pedicle, and the incision of the IVC was closed by a single layer suture. No extracorporeal circulation was needed. The abdomen was closed leaving one tubular drainage in the retroperitoneal space and the patient transferred to the intensive care unit. The postoperative course was uneventful and the patient was discharged on postoperative day 6.

Pervenuto in Redazione Giugno 2022. Accettato per la pubblicazione Ottobre 2022
Correspondence to: Emanuele Casciani (e-mail: emanuelecasciani @gmail.com)

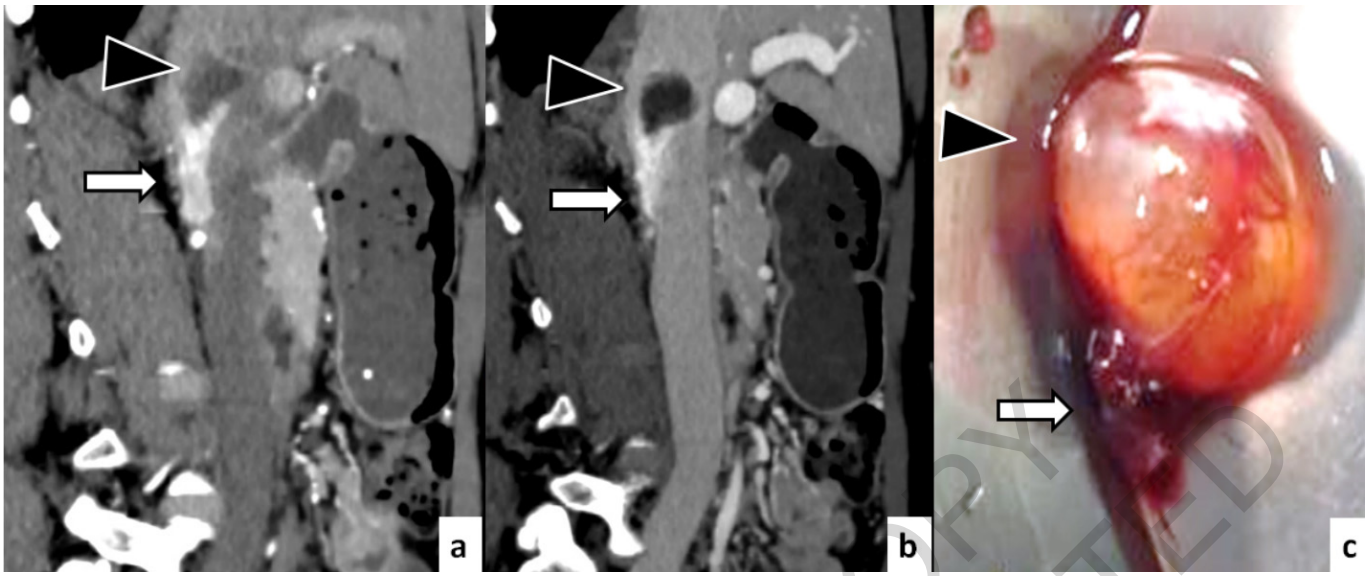

Fig. 1: Para-sagittal multiplanar reconstruction (MPR) from arterial. A) and venous, B) phases of CTA and macroscopic appearance, C) showed an intraluminal non-enhancing tumor (arrowhead) with peduncolated hypervascular structure (arrow) attached to the wall of the IVC.

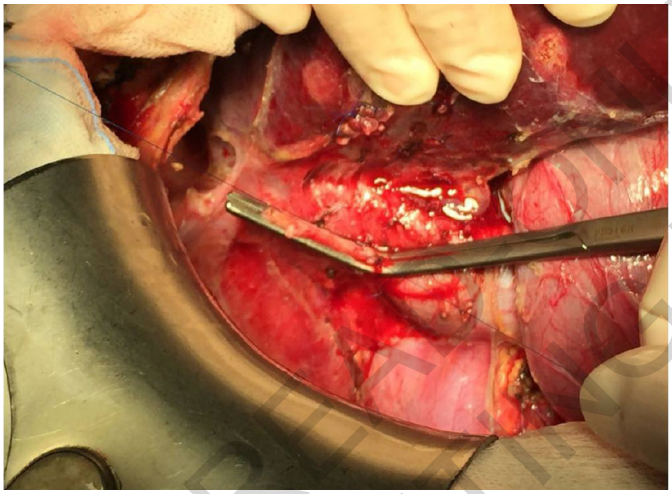

Fig. 2: Operative field, showing the IVC and the renal veins isolated and taped. The tumor was found superiorly to the renal vein confluence (note that there were two right renal veins).

A macroscopic examination of the resection specimen found a pseudocyst formation with thin walls (2,5 cm) of reddish color with vascularized pedicle within the lumen of the IVC (Fig. 1c). The tumor demonstrated typical histological features of EHE, including positive immunohistochemical staining for the endothelial markers cluster of differentiation CD 31, CD 34, cytokeratin AE1/AE3 and actyne smooth-muscle. The tumor did not have areas of necrosis or cytological atypia with a mitotic rate of < 1/10 high-powered fields; moreover the cell proliferation index assessed with proliferation marker Ki-67 was found < 1%. These findings suggested a low risk of metastasis.

One year after surgery, a follow up CTA scan demonstrated no tumor recurrence.

Discussion

EHE is a rare disease that is often misdiagnosed and for which there is little understanding of the etiology, management or prognosis. Only surgical therapy for EHE has proven beneficial, with an ambiguous role for adjuvant chemotherapy and none for radiation therapy.
Clinical presentation was variable, depending on the size, the location of the tumor, the entity of occlusion of the vessel, and on the involvement of other organs.
It's important to diagnose correctly the mass before determined the appropriate treatment plan.
In the case report of Kabbani L, et al [3] thrombus without tumor was reported on the initial CT and mechanical thrombectomy was performed with an endovascular thrombectomy device, followed by stent placement in the inferior vena cava. The initial pathology on the retrieved specimen revealed an undifferentiated neoplasm, and a second time of surgical resection of the retroperitoneal soft tissue tumor of the IVC has been needed and documented a rare case of EHE.
Up to now, cross sectional imaging has been used to evaluate wall infiltration of the cava vein, extra parietal extension, infiltration of adjacent organs, the presence of adenopathies and distant metastases [4].
A vascular pedicle has not been described in the previous ICV EHE studied with CT [1-3,5-6].
We firstly described the possibility of identifying the vascular pedicle of the endoluminal mass in the ICV with arterial and venous phases of CTA. CTA performed with

MPR has been extremely accurate in the preoperative evaluation of ICV mass. MPR can be performed, not just in sagittal and coronal planes, but in any defined anatomic plane, thereby providing a novel perspective that is customized to the unique anatomy of the patient. This feature can be useful for the characterization of the endoluminal mass.

Surveillance by CT is recommended since even after surgical excision wide, the HEE can recur or metastasize. The rate of recidivism is 13%. About 30% of tumors will present metastases, most of the time of late onset, basically in regional lymph nodes and lungs [7], but this does not seem to influence survival. Less than half of patients who develop metastases die and for those who survive the mortality is less than 20% at five years old. Metastases in the lymph nodes locoregionals are available for surgical excision and this allows a longer survival of patients [6]. Currently it is believed that whatever the treatment, the survival at age five varies between 43% and 71.3% [4]. Yet a prognosis less favorable had been proposed in the publication initial Weiss and Enzinger where the average survival without recurrence of 20 out of 31 cases followed by four months to 18 years was 18 months old [8].

In conclusion, multidetector CTA with MPR was found to be an effective method to demonstrate the presence of a pedunculated mass in the IVC, providing useful indications for surgical planning.

Riassunto

Gli emangioendoteiomi epitelioidi primari (EHE) della vena cava inferiore (IVC) sono tumori maligni estremamente rari di origine vascolare. In letteratura sono descritti solo circa 40 casi di EHE vascolari e solo quattro originati da IVC (1-4) e due da vena cava superiore (5, 6). Solo la terapia chirurgica per l'EHE si è rivelata utile. Spesso è difficile fare la diagnosi corretta con l'imaging prima dell'esame istologico. L'imaging, di solito, aiuta la pianificazione chirurgica, ma nel nostro caso ha aiuto a fare anche la diagnosi corretta. Presentiamo un raro caso di un paziente con un IVC EHE, studiato con angiografia con tomografia computerizzata (CTA), risonanza magnetica (MR) e tomografia computerizzata con tomografia a emissione di positroni con fluoro 18 fluorodeossiglucosio (18F-FDG PET/CT).

References

1. Gundara JS, Gill AJ, Neale M, et al: *Inferior vena cava epithelioid hemangioendothelioma*. J Vasc Surg, Venous and Lym Dis, 2013; 1:75.

2. Henriquez CR, Cazes A, Fabiani JN, Bruneval P: *Epithelioid hemangioendothelioma of the inferior vena cava*. Ann Pathol, 2011; 31:218-21.

3. Kabbani L, Balraj P, Mathews J: *Epithelioid hemangioendothelioma presenting as inferior vena cava obstruction diagnosed using an endovascular thrombectomy device*. Ann Vasc Surg, 2017; 40:294.

4. Casciani E, Polettini E, Sollaku S, et al.: *Imaging of primary retroperitoneal neoplasms.* Ann Ital Chir, 2022; 93, 5: 489-503.

5. Scordi-Bello IA, Snyder A, Schwartz M, Fallon JT: *Intravascular epithelioid hemangioendothelioma of the inferior vena cava: Case report of an unusual and unpredictable vascular tumor.* Cardiovascular Pathology, 2009; 18:243-46.

6. Lahon B, Fabre D, De Montpreville V, Dartevelle P: *Epithelioid haemangioendothelioma of the superior vena cava*. Interactive Cardio Vascular and Thoracic Surgery, 2012; 15:186-87.

7. Ferretti GR, Chiles C, Woodruff RD, Choplin RH: *Epithelioid hemangioendothelioma of the superior vena cava: Computed tomography demonstration and review of the literature*. J Thorac Imaging, 1998; 13:45-8.

8. Palsson B: *Epitheloid hemangioendothelioma.* Acta Oncol 1999; 38: 659-61.

9. Weiss SW, Enzinger FM: *Epithelioid hemangioendothelioma: A vascular tumor often mistaken for a carcinoma*. Cancer, 1982; 50:970-81.