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CASE REPORT Open Access

Rare presentation of recurrent ovarian carcinoma with secondary Budd–Chiari syndrome: a case report

S. Damini[1*], S. H. Chandrashekhara[1] and M. D. Ray[2]

Abstract

Background Budd–Chiari syndrome (BCS) is a rare condition, usually associated with hematological disorders such as thrombotic diathesis and hypercoagulability. Serum CA-125 level is an established tumor marker of ovarian malignancy; however, cases of primary BCS may also show raised CA-125 levels. BCS in a case of ovarian carcinoma is usually primary in nature due to hypercoagulable state, and raised CA-125 levels with tender hepatomegaly in a treated case of ovarian carcinoma usually imply metastatic recurrence in the liver. However, our case demonstrates an atypical secondary cause of BCS in such a patient caused by extrinsic compression of IVC due to recurrent disease.

Case presentation We report an unusual case of a 69-year-old female who presented with nausea and abdominal pain. She had a 7-year-old history of endometrioid carcinoma of the right ovary for which she underwent total abdominal hysterectomy, bilateral salpingo-oophorectomy, omentectomy and pelvic lymph node dissection along with adjuvant chemotherapy. Currently, she had right hypochondrium tenderness, deranged liver function tests (LFT) and raised CA-125 levels, which raised suspicion of hepatic metastasis. However, CECT abdomen revealed peripheral mottled enhancement of liver with multifocal extrahepatic tumor deposits, one of them causing compression of inferior vena cava (IVC) implying a diagnosis of secondary Budd–Chiari syndrome.

Conclusions In a background of treated ovarian malignancy with raised CA-125 levels and deranged LFT, primary suspicion is of hepatic tumor recurrence. However, in our case, radiological investigation revealed diagnosis of secondary Budd–Chiari syndrome due to perihepatic metastatic recurrence with the absence of frank intrahepatic lesions.

Keywords Budd–Chiari syndrome, Ovarian carcinoma, Computed tomography, Case report

Background

Budd–Chiari syndrome (BCS) is differentiated into primary or secondary depending on the mechanism of obstruction of hepatic veins or inferior vena cava (IVC). Primary BCS is initiated from an obstruction within the vein, and secondary BCS is due to external obstruction of the vein [1].

Hematological diseases or systemic conditions leading to thrombotic diathesis and hypercoagulability are the most common causes of primary BCS [2, 3]. Underlying malignancy like ovarian carcinoma is one such cause of prothrombotic state [2].

Serum CA-125 level is an established tumor marker of ovarian malignancy; however, cases of primary BCS may also show raised CA-125 levels [1, 4]. Shah et al. [5] reported a case of primary Budd–Chiari syndrome with significantly raised CA-125 levels without any underlying ovarian neoplasm.

*Correspondence:
S. Damini
damy2804@gmail.com
[1] Department of Radiodiagnosis, All India Institute of Medical Sciences, New Delhi 110029, India
[2] Surgical Oncology, IRCH, All India Institute of Medical Sciences, New Delhi 110029, India

Raised CA-125 levels with tender hepatomegaly in a treated case of ovarian carcinoma raised initial suspicion of metastatic recurrence in the liver. BCS in a case of ovarian carcinoma is usually primary in nature due to hypercoagulable state caused by underlying malignancy. However, our case demonstrates an atypical secondary cause of BCS in such a patient caused by extrinsic compression of IVC due to recurrent disease.

Case presentation

A 69-year-old female presented to our hospital with complaints of nausea and pain in the abdomen for 2 weeks. She was diagnosed with endometrioid carcinoma of the right ovary—stage IIIc 7 years ago, for which she underwent a total abdominal hysterectomy, bilateral salpingo-oophorectomy, omentectomy and pelvic lymph node dissection. She had also received six cycles of adjuvant chemotherapy (Paclitaxel + Carboplatin), following which she was lost to follow-up.

Currently, the patient denied any history of fever, cough, dyspnea or jaundice; however, she reported a 2.5 kg weight loss over the past year. She was neither hypertensive nor diabetic and was a lifetime non-smoker with no significant family history of any major diseases. General physical examination revealed mild tenderness in right hypochondrium. Laboratory investigations revealed anemia (hemoglobin 6.8 mg/dl) with a normal total white blood cell count (10.4×10^9/L), platelet count (2.4×10^5/uL) and coagulation profile (PT 12.5s, INR 1.0). The liver function tests were mildly deranged (alkaline phosphate 256 IU, total protein 7.2 g/dL and serum albumin 3.4 g/dL). The kidney function tests (blood urea 22 mg/dL and serum creatinine 0.22 mg/dL) were within normal limits. Moreover, serum CA-125 level of the patient was significantly raised to 936 U/mL.

In view of recently developed symptoms and signs, the patient was referred for contrast-enhanced CT (CECT) scan of chest and abdomen to look for recurrence. CECT was done on a 128-slice multidetector CT scanner after injecting 1.5 ml/kg Iohexol contrast agent and 1 mm thin sections were acquired. One liter of diluted oral iodine-based contrast (5% dilution) was also administered over an hour before the scan. The scan was interpreted by a senior consultant radiologist with 20 years of experience in the field of gastrointestinal radiology. The images revealed multiple heterogeneously enhancing pleural and peritoneal deposits (Figs. 1, 2, 3). The largest deposit was located at the right posterior cardio-phrenic region, which was compressing the supra-hepatic IVC anteriorly, causing its significant luminal attenuation and obstruction (Fig. 1). The liver was of normal size and showed peripheral mottled heterogeneous enhancement (Figs. 1, 3) with normal central enhancement and sparing of caudate lobe (Fig. 3). No other focal lesion was seen in the liver. Hepatic veins and its confluence with IVC (Fig. 3) were normal. Diagnosis of secondary Budd–Chiari syndrome was made on the basis of radiological imaging alone without the need for liver biopsy.

The patient was subjected to positron emission tomography-CT (PET-CT) which showed high metabolic uptake in the masses detected on CT scan, confirming recurrence. There were no established treatment protocols to address this specific case due to its unique nature. Thus, the counseling and treatment options were based primarily on anecdotal experience. Surgical decompression with metastatectomy would have been the ideal choice of treatment in this case. However, after a multi-disciplinary team discussion, in view of advanced cancer state and poor health status indicating poor prognosis, IVC stenting was planned after initiation of palliative chemotherapy and

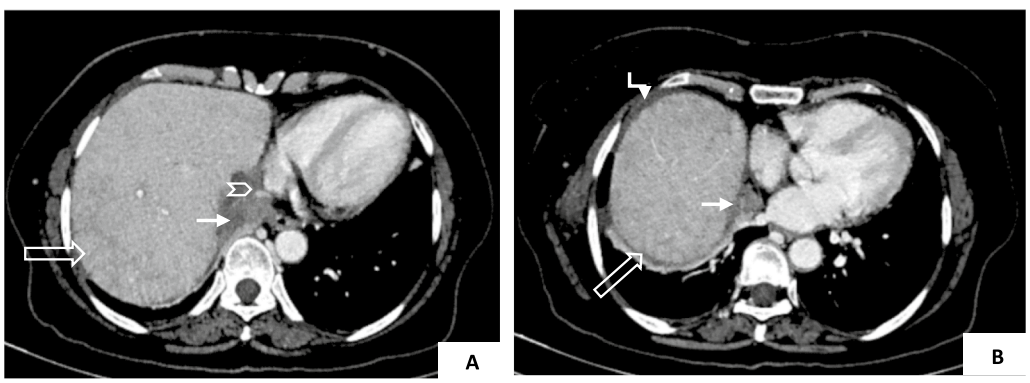

Fig. 1 **A**, **B** Axial CECT at the level of right atrium shows heterogeneously enhancing deposit (white arrow) in right posterior cardio-phrenic angle, causing slit-like compression of IVC (arrowhead). Liver shows resultant peripheral mottled enhancement pattern (open arrow). Similar deposits noted along the subphrenic surface of liver (elbow arrow)

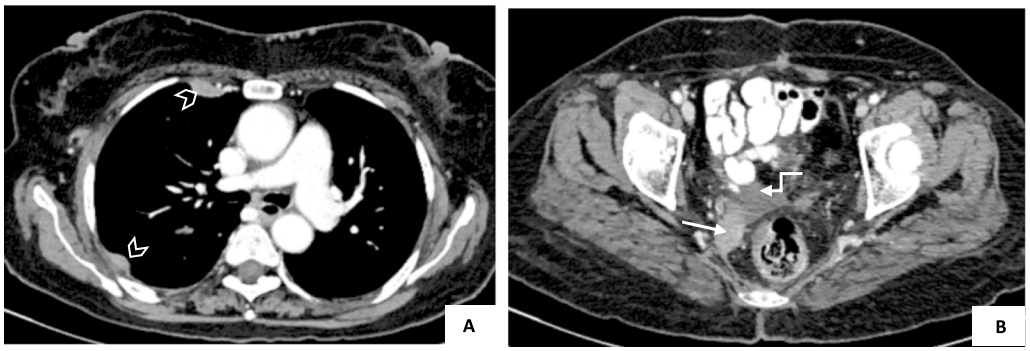

Fig. 2 **A**, **B** Axial CECT at the level of thorax and pelvis, respectively, depicts heterogeneously enhancing deposits along right pleura (arrowheads) and right adnexa (white arrow). Mild ascites is also noted (elbow arrow)

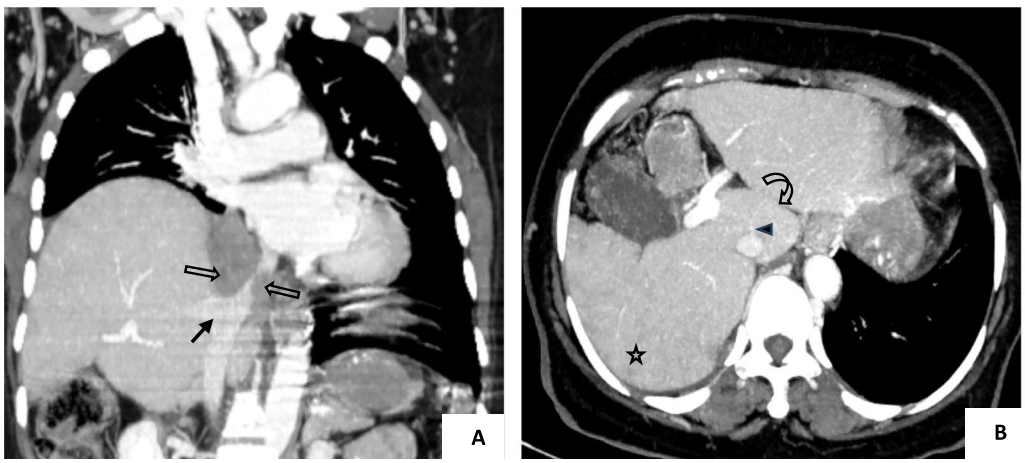

Fig. 3 **A**, **B** are coronal and axial maximum intensity projection (MIP) images, respectively; **A** depicts gross compression of supra-hepatic inferior vena cava (IVC) by the deposit (black open arrows) in right posterior cardio-phrenic angle. Black solid arrow shows the confluence of hepatic vein into IVC. **B** Relative sparing of caudate lobe (curved arrow) and mottled enhancement of periphery of liver (star). Caudate vein draining directly into IVC is also seen (black arrowhead)

radiotherapy to treat metastases. However, the patient succumbed to the initial cycles of medical treatment.

Discussion

BCS is a rare entity and by definition is blockade of the IVC or hepatic venous outflow tract [6]. Venous thrombosis is the most common cause of BCS, and this is termed as primary BCS [6]. This can be due to underlying diseases leading to hypercoagulability or idiopathic in nature. Secondary BCS is much less common. Secondary Budd–Chiari syndrome occurs when a lesion outside the veins, typically a malignancy, compresses or invades the hepatic veins and/or the inferior vena cava [2].

Idiopathic BCS was found in up to 25% of the patients; however, recently it has been established that multifactorial or specific detectable cause can be elicited in 90% of the primary BCS patients due to advanced technology in gene testing and imaging modalities [2, 3].

It has been found that many patients initially labeled as idiopathic BCS tend to have underlying undetected myeloproliferative disease. In a study described by Hitawala et al. [2], only 20 percent of BCS patients had no diagnosable cause and were termed idiopathic BCS, while 80 percent of the cases had an underlying cause leading to hypercoagulable state resulting in development of Budd–Chiari syndrome. Around 50% of the them were associated with myeloproliferative disorders, and only around 10% were associated with other malignancies. Malignancies cause BCS by either leading to hypercoagulability and venous thrombosis or due to direct tumor invasion of vessels [2]. The most common malignancies associated

with BCS are hepatocellular carcinoma (HCC), followed by adrenal tumors, renal cell carcinoma (RCC), leiomyosarcoma, right atrial myxoma and Wilms tumor. Thus, it is important to look for the cause along with diagnosis of Budd–Chiari syndrome for prompt treatment [2].

The key clinical features of BCS are hepatomegaly, ascites and abdominal pain [7].

Clinically, BCS has been classified depending upon the presentation and duration of disease [3] (Table 1).

Earlier, invasive investigations were the main stay of BCS diagnosis. Nowadays, noninvasive cross-sectional imaging such as CT/MRI and ultrasonography is found to be sufficient for diagnosis of BCS [8].

CT scans play a vital role in confirming BCS. It demonstrates vascular abnormalities, evaluates the hepatic parenchymal changes as well as allows vascular mapping for guiding surgery or endovascular interventions such as trans-jugular intrahepatic portosystemic shunt (TIPS) procedure [3, 8] (Table 2).

In the acute stages of BCS (Fig. 1), liver may be normal or enlarged in size with mottled appearance on CECT abdomen, related to inhomogeneous perfusion of the liver, as seen in our case. In late arterial phase, liver displays decreased patchy peripheral enhancement due to portal and sinusoidal stasis, with higher central enhancement [8] (Fig. 3), followed by reversal of enhancement pattern in portal venous phase which is referred to as "flip-flop pattern". Ascites and caudate lobe enlargement may also be seen. Thrombosis or compression of hepatic veins/IVC may also be seen [3, 8].

Table 1 The clinical classification of Budd–Chiari syndrome

Type	Presentation
Acute	< 1 Month. Ascites, initiation of hepatic necrosis, without venous collaterals
Sub-acute	1–6 Months. Ascites, minimal hepatic necrosis and porto-hepatic venous collaterals
Chronic	> 6 Months. Additional features of cirrhosis

The sub-acute type of BCS is characterized by volume redistribution and development of porto-hepatic collaterals in addition to the heterogenous enhancement of peripheral liver in late arterial phase with homogenous attenuation in delayed phases [3].

Chronic BCS is characterized by predominant features of chronic liver disease such as nodular outline with asymmetric atrophy of liver, differential attenuation of liver parenchyma and possible additional features of portal hypertension. However, the differential enhancement pattern between the peripheral and central zones of liver is less conspicuous as compared to acute/sub-acute variants. Additional development of arterial phase hyper-enhancing regenerative nodules may also be seen which show persistent enhancement in later phases. Chronic BCS also predisposes to development of HCC [3].

The complications associated with Budd–Chiari syndrome (BCS) often depend on pre-existing condition and the extent of hepatic damage. If left untreated, BCS can lead to a serious cascade of events, including hepatic encephalopathy, variceal hemorrhage, hepatorenal syndrome, portal hypertension and bacterial peritonitis in the presence of ascites and HCC [9].

Secondary BCS due to direct tumoral or metastatic invasion of hepatic veins, IVC or right atrium is less frequently found scenario [4]. Horiguchi et al. [10] reported a case of intrahepatic metastasis from thymoma causing IVC obstruction with resultant BCS. Shih et al. [11] reported a case of left renal tumor directly infiltrating left renal vein, extending to the inferior vena cava and hepatic veins causing BCS. Kropf et al. [12] described a patient of breast carcinoma presenting with liver metastasis leading to BCS. We have not found any reported case of secondary BCS due to ovarian carcinoma in the literature as was seen in our case.

The treatment of BCS necessitates an algorithmic approach after a complete interdisciplinary team discussion. The aim of the treatment is to relieve obstruction, treat underlying conditions, limit liver deterioration and manage complications [2, 6] (Table 3).

Table 2 The utility of cross-sectional imaging for diagnosing Budd–Chiari syndrome

Role and advantage of cross-sectional imaging in BCS diagnosis [2, 8]:
1. Noninvasive modality with high sensitivity and specificity of 89 and 72% for CT
2. To evaluate suspicion of underlying mechanical obstruction as the cause
3. To identify rare causes of venous obstruction such as membranous web in IVC
4. To confirm intrahepatic collateral veins which appear due to HV obstruction (classified into four types: large collaterals draining into the IVC, subcapsular veins, collateral cobwebs and veno-venous shunts)
5. Modality of choice to characterize nodules associated with BCS

Table 3 The treatment options for Budd–Chiari syndrome [2, 13]

Treatment options for Budd–Chiari syndrome
Anticoagulants are the first line therapy—starting with low-molecular-weight heparin and then transitioned to warfarin, which continued lifelong. PT/INR monitoring is done and kept in therapeutic range for patients on warfarin
Radiologic intervention local thrombolysis and stenting, balloon angioplasty or a trans-jugular intrahepatic portacaval shunt (TIPS) in acute BCS, which are unresponsive
Surgical decompression can be attempted in acute forms of BCS, when other options fail
Liver transplantation offered in cases progressed to decompensated liver cirrhosis or as salvage treatment in the setting of failed portal derivative techniques

Conclusions

In a patient with history of ovarian malignancy, raised CA-125 levels and deranged LFT generally raise a suspicion of hepatic tumor recurrence. However, secondary BCS due to juxta-caval metastatic recurrence can be a rare cause for similar clinical presentation. Radiological imaging helps in such scenarios for achieving a comprehensive diagnosis and to guide further management.

Abbreviations

ALP Alkaline phosphatase
BCS Budd–Chiari syndrome
CECT Contrast-enhanced computed tomography
CT Computed tomography
IVC Inferior vena cava
MRI Magnetic resonance imaging

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Not applicable.

Consent for publication

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Competing interests

The authors have no conflicts of interest.

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