

Massive ovarian edema mimicking an ovarian neoplasm in a patient with IVC web

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ABSTRACT

Massive ovarian edema is an uncommon benign condition affecting young females predominantly those in the child bearing age group and preadolescent girls. Its clinical and radiologic overlap with ovarian neoplasms and torsion which require surgical intervention makes it imperative for the radiologist to consider this entity preoperatively as preserving fertility is vital in this young age group. We report a case of massive ovarian edema, a rare presentation in a patient with inferior vena cava web and consequent Budd Chiari Syndrome, an association previously unreported in literature.

CASE REPORT

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A 28-year-old nulliparous female was referred from the Department of Gynaecology for right lower abdominal pain and metrorrhagia for 3 months. She had a history of pulmonary tuberculosis for which she was treated. Her laboratory parameters including ovarian tumor markers (carcinoembryonic antigen, cancer antigen 125 and 19-9, alpha-fetoprotein and lactate dehydrogenase) were within normal limits. A provisional diagnosis of tubercular pelvic inflammatory disease was considered for which the patient was being worked up. She had undergone repeated ultrasound and Computed Tomography (CT) scans for her pelvic complaints outside our hospital which either revealed right ovarian mucinous cystadenoma or torsion. However, patient's ovarian tumor markers were within normal limits and her pain was vague and long-standing unlike that seen in torsion.

She was referred to us for Contrast-enhanced CT (CECT) of the abdomen to look for any evidence of tuberculosis. On CECT, right adnexa showed a heterogeneously enhancing lesion (mean attenuation 25 HU) with mild free fluid in pelvis. In addition, complete occlusion of the intrahepatic inferior vena cava (IVC at T10-T11 vertebral level) with irregular contour of the liver and caudate lobe hypertrophy was noted. Multiple lumbovertebral, mesenteric, retroperitoneal and abdominal wall collaterals were seen along with mesenteric and retroperitoneal fat engorgement. No imaging features of tuberculosis were seen (Figure 1). A diagnosis of IVC occlusion with Budd Chiari syndrome and multiple venous collaterals was made and ultrasonography (USG) of the pelvis was performed for better characterization of the adnexal lesion.

USG revealed enlarged right ovary with hypochoic hypovascular stroma and peripherally displaced follicles. Mild free fluid was noted in pelvis. Uterus was unremarkable. Upper

abdomen examination showed occlusion of a segment of intrahepatic IVC with echogenic contents within (Figure 2).

Magnetic Resonance Imaging (MRI) Pelvis was performed for further evaluation of the adnexa. MRI showed enlarged right ovary (measuring ~3.4 (anteroposterior) x 5.4 (transverse) x 3.7 (craniocaudal) cm) with T2 hyperintense stroma and peripherally arranged follicles. No discrete mass lesion was seen in the ovary (Figure 3).

A final diagnosis of IVC occlusion with Budd Chiari Syndrome and massive ovarian edema was made. To the best of our knowledge, this association has not been previously reported.

The patient is currently on follow-up and is being considered for angioplasty with stenting of the IVC.

DISCUSSION

Massive ovarian edema is a rare tumor-like condition affecting young women [1]. WHO describes it as 'the accumulation of edema fluid within the ovarian stroma separating normal follicular structures' [1]. The condition was first described by Kalstone et al in 1969 [2]. Its clinical presentation and imaging findings are often non-specific and overlap with ovarian neoplasms which leads to unnecessary oophorectomy.

Budd Chiari Syndrome is a rare liver disease with a prevalence of 5 per million in Asia [3]. IVC web, though an uncommon vascular anomaly resulting from a congenital cause or as a sequelae to thrombosis, is the most frequent cause of Budd Chiari syndrome in Asia [3,4].

Etiology & Demographics:

Massive ovarian edema most often affects females in the second and third decades, although it can occur at any age [5]. It is hypothesized that partial recurrent torsion of the mesovarium impedes ovarian venous and lymphatic drainage resulting in stromal edema while the arterial supply of the ovary is preserved, maintaining viability [5]. However, cases without torsion have also been reported [6]. Right ovarian vein drains directly into the IVC while the left ovarian vein drains into the left renal vein. The lymphatic drainage of the ovary is to the internal iliac, paraaortic, paracaval and inguinal group of lymph nodes [7].

Most cases are unilateral with right-sided predominance because of higher pressure in the right ovarian vein due to its drainage pathway [5].

Clinical & Imaging Findings:

The common presenting symptoms include intermittent abdominal pain, palpable tuboovarian mass, menstrual disturbances, nausea, vomiting and infertility. Rarely, patients present with virilization and precocious puberty. Occasionally, it may be incidentally picked up during surgery for other causes [5,6].

Ultrasound is the first-line imaging modality for adnexal pathologies due to its easy availability, affordability, lack of ionizing radiation and acoustic window provided by the distended urinary bladder. In addition, transvaginal USG provides excellent resolution. Massive ovarian edema may have variable appearance on USG ranging from solid tumor-like enlargement of the ovary or a complex solid-cystic lesion or an enlarged hypoechoic ovary with peripherally displaced follicles and preserved architecture. In a case series by Dahmouh et al, two of three cases, did not show reliable colour flow signal on colour/spectral doppler. Ovarian stromal vascularity could be demonstrated in one of their cases which showed increased echogenicity of the stroma. Teardrop configuration of the ovary was also seen in their study. Ipsilateral deviation of the uterus, endometrial hyperplasia and ascites are other associated findings [5].

MRI is the imaging investigation of choice for evaluating ovarian lesions because of its exquisite soft tissue contrast resolution. The enlarged ovary shows a homogeneous low signal intensity on T1 and a heterogeneous high signal intensity with peripherally located follicles on T2 weighted image which are better appreciated on post-contrast images [5]. This is explained by the peripheral displacement of follicles by edema fluid accumulated in the stroma [8]. There may be homogeneous or heterogeneous enhancement of the ovarian stroma [5].

CT is not routinely advocated for imaging ovarian pathologies. However, reported CT findings include an enlarged hypodense ovary, ipsilateral deviation of the uterus and presence of a twisted edematous ipsilateral fallopian tube [5].

Treatment & Prognosis:

Most published cases have been treated with salpingo-oophorectomy because of intra and post-operative concern for malignancy [5].

IVC web can be treated by medical, endovascular, surgical approach or a combination of these. Medical therapy includes systemic thrombolysis. Endovascular procedures include catheter-directed thrombolysis, angioplasty and stent placement. Surgical procedures such as thrombectomy, reconstruction and ligation are used as last resort [9].

Differential Diagnoses:

Ovarian neoplasms

Most solid ovarian tumors are hypointense on T2 weighted images and lack the presence of follicles, unlike massive ovarian edema. The lack of a discrete mass or enhancing nodule favours the diagnosis of massive ovarian edema. The marked T2 hyperintensity on MRI may simulate a cystic ovarian mass, however, this pitfall can be avoided with ultrasound correlation as cystic ovarian mass appears anechoic with posterior acoustic enhancement while edematous ovary is hypoechoic on USG [5].

Ovarian torsion

The presence of ovarian enlargement with peripherally displaced follicles in massive ovarian edema may mimic torsion. However, patients with torsion usually have a more acute presentation and twisted vascular pedicle and lack of contrast enhancement is usually seen on imaging. Intact ovarian vascularity and enhancement point towards a diagnosis of ovarian edema [5].

Polycystic ovaries

Polycystic ovaries closely mimic massive ovarian edema as both show enlarged ovaries with increased ovarian volume and peripherally displaced small follicles, but the latter is mostly unilateral with right-sided predominance as opposed to polycystic ovaries which are usually bilateral. Stromal T2 hyperintensity also favours massive ovarian edema [5].

TEACHING POINT

Massive ovarian edema is a close mimicker of ovarian neoplasms and torsion, both of which require surgical intervention. The radiologist plays a key role in suspecting/diagnosing massive ovarian edema preoperatively to avoid inadvertent oophorectomies as it is important to preserve fertility in this young age group. The clinician and radiologist must be aware of this uncommon, yet plausible presentation of IVC web syndrome.

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FIGURES

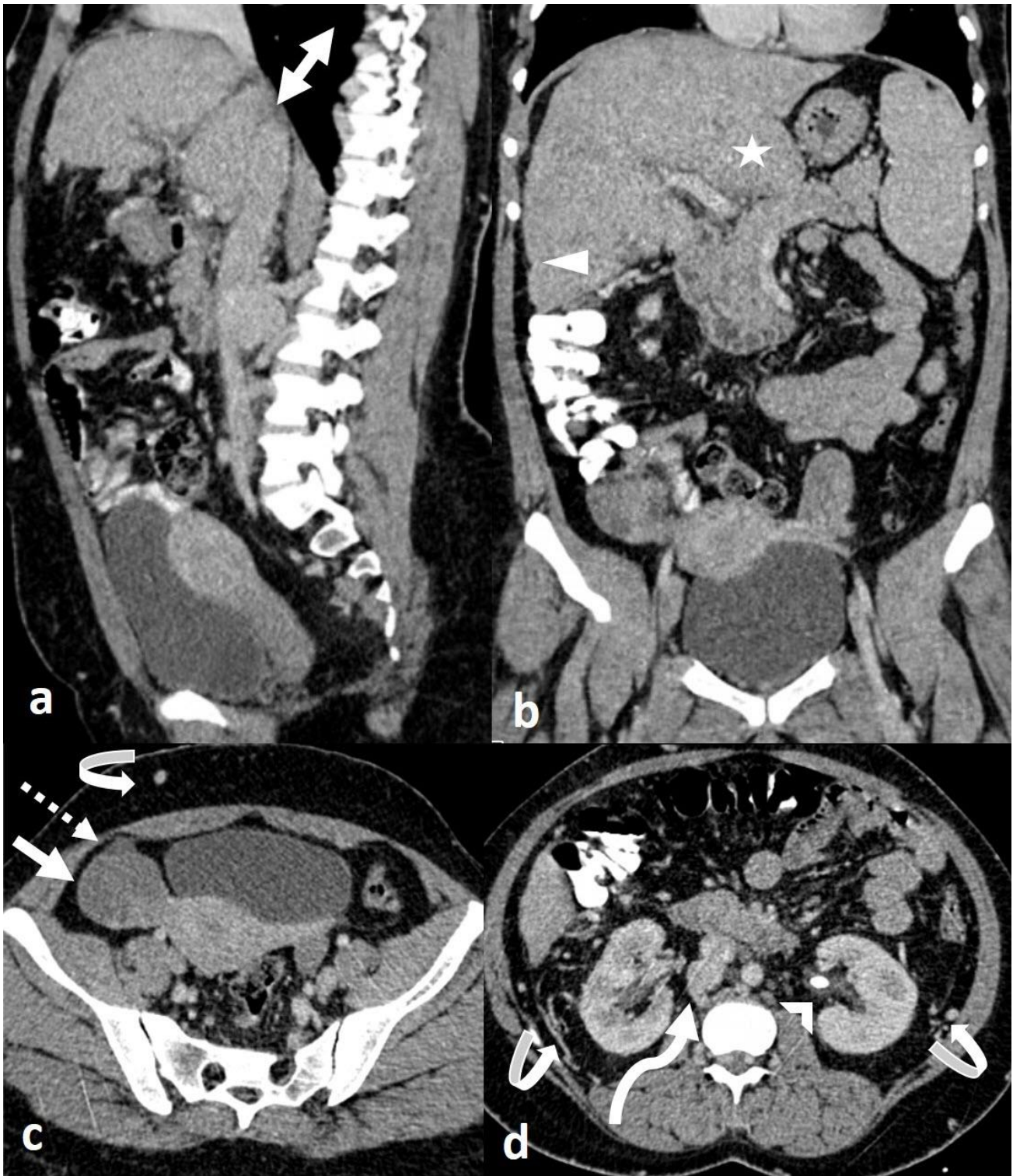


Figure 1: 28-year-old female with IVC web with consequent Budd Chiari syndrome and Massive ovarian edema.

FINDINGS: Sagittal (a) and coronal (b) CECT images of the abdomen show complete occlusion of a segment of intrahepatic IVC (double arrow in a) with irregular hepatic contour (arrowhead in b) and caudate lobe hypertrophy (* in b). Axial CT image of the pelvis (c) shows a well-defined predominantly hypodense lesion in right adnexa (arrow in c). Right ovary is not seen separately. Mild free fluid is seen in pelvis (dashed arrow in c). Axial CT image at the level of kidneys (d) shows dilated ascending lumbar vein on right draining into the IVC (curved arrow in d). Multiple retroperitoneal and abdominal wall collaterals are seen (curved block arrows in c,d). Note is made of left renal pelvic calculus (arrowhead in d).

TECHNIQUE: 128 slice, SIEMENS SOMATOM Definition AS+, contrast-enhanced CT scan [200mAs 120 kV, 1 mm slice thickness] acquired 60 seconds after intravenous contrast injection in the venous phase (60 mL of non-ionic contrast medium; Omnipaque 350mgI/mL, GE Healthcare, USA). Window level: 40 and window width: 300.

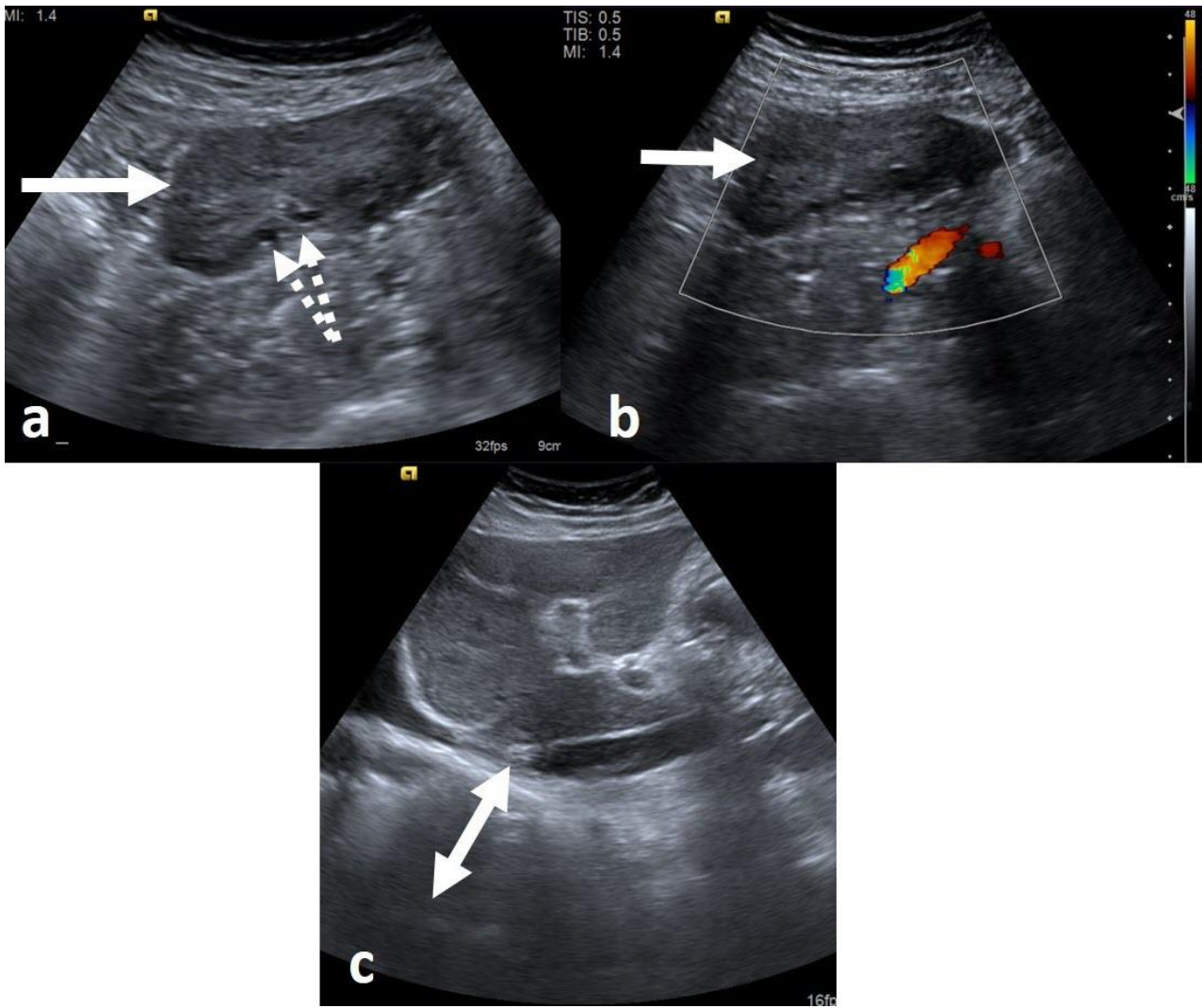


Figure 2: 28-year-old female with IVC web with consequent Budd Chiari syndrome and Massive ovarian edema.

FINDINGS: Axial grey scale (a) and doppler (b) images of the pelvis show enlarged ovary with hypoechoic hypovascular ovarian stroma (arrow in a,b) and peripherally displaced follicles (dashed arrows in a). Longitudinal grey scale ultrasound image of liver (c) shows occlusion of a segment of intrahepatic IVC with echogenic contents (double arrow).

TECHNIQUE: Real-time ultrasound images of the abdomen acquired on SIEMENS ACUSON s2000 with 6 MHz curvilinear probe.

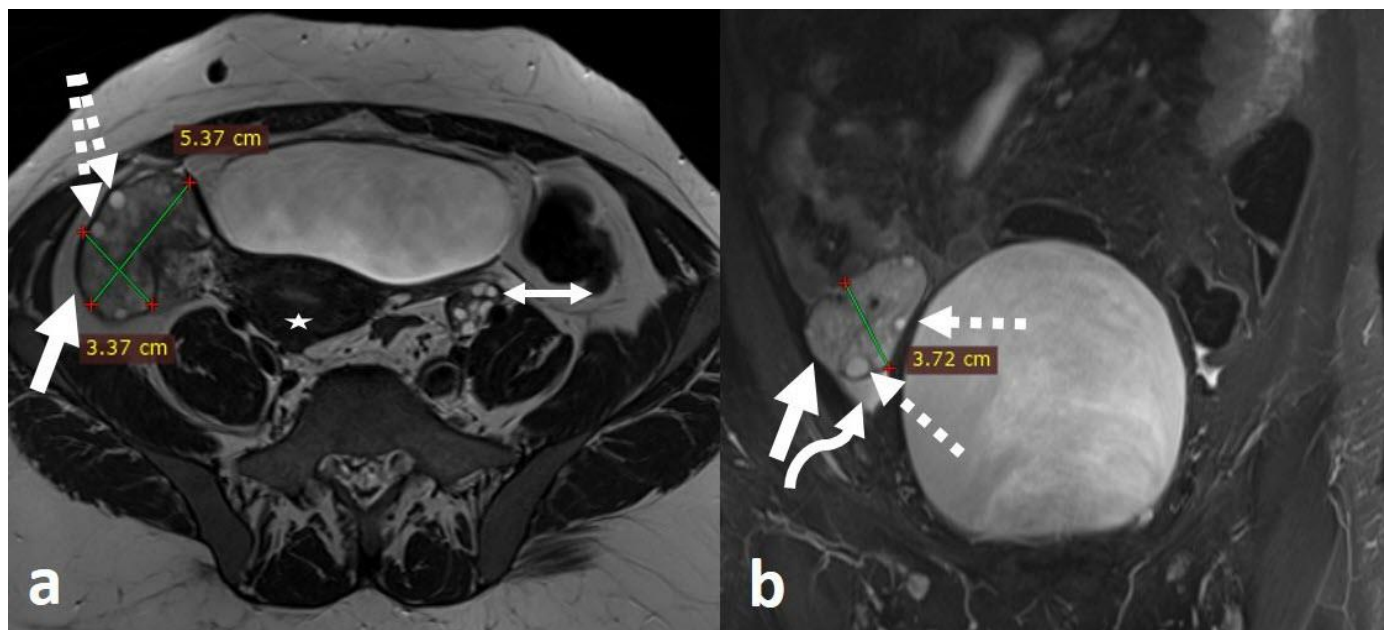


Figure 3: 28-year-old female with IVC web with consequent Budd Chiari syndrome and Massive ovarian edema.

FINDINGS: Axial T2W (a) and STIR Coronal MR images (b) show enlarged right ovary, measuring~ 3.4 (AP) x 5.4 (Trans) x 3.7 (CC) cm with hyperintense stroma (arrow) and peripherally arranged follicles (dashed arrows). Mild free fluid is seen in pelvis (curved arrow in b). Left ovary is normal in size and shows multiple follicles (double arrow in a). Uterus is unremarkable (* in a). No discrete mass lesion is seen.

TECHNIQUE: 3T MR scanner- MAGNETOM SKYRA (Siemens, Erlangen, Germany), images acquired in supine position using a body coil. T2WI axial [TR: 4450ms, TE: 89ms] and STIR coronal [TR: 3520ms, TE: 56ms], non-contrast scans

Etiology	Venous and lymphatic obstruction of the ovary
Incidence	Rare entity; with only case reports and case series existing in literature
Age predilection	Females in 2 nd – 3 rd decade
Risk factors	Recurrent partial torsion of the mesovarium, hypercoagulable state, extrinsic compression on the gonadal vein, left renal vein and IVC
Treatment	Wedge resection with frozen section and fixation of ovary to the uterus and treatment of the cause
Prognosis	Good
Findings on imaging	USG: Solid tumor-like enlargement of the ovary or a complex solid-cystic lesion or an enlarged hypoechoic ovary with increased through transmission and peripherally displaced follicles. MRI: The enlarged ovary shows a homogeneous low signal intensity on T1 and a heterogeneous high signal intensity with peripherally located follicles on T2 weighted image which are better appreciated on post-contrast T1 weighted images.

Table 1: Summary table of massive ovarian edema.

	USG	CT	MRI
Massive ovarian edema	Enlarged hypoechoic ovary with peripherally displaced follicles. Teardrop configuration of ovary may be seen.	Enlarged edematous hypodense ovary with Hounsfield units less than 40, ipsilateral deviation of the uterus.	Enlarged ovary with peripherally located follicles and normal post-contrast enhancement.
Ovarian neoplasms	Discrete solid or solid cystic mass with loss of normal ovarian architecture.	Solid or solid cystic adnexal lesion with/without calcifications, ascites, nodal and distant metastases.	Solid component appears hypointense on T2WI with loss of normal ovarian architecture +/- evidence of metastases.
Ovarian torsion	Enlarged ovary with reduced or absent vascularity and a twisted tubular structure corresponding to vascular pedicle (whirlpool sign).	Enlarged ovary with/without an underlying ovarian lesion (lead point for torsion) with a twisted vascular pedicle. Lack of contrast enhancement and haemorrhage may be seen.	Enlarged ovary with/without an underlying ovarian lesion (lead point for torsion) with a twisted vascular pedicle. Lack of contrast enhancement and haemorrhage may be seen.
Polycystic ovaries	Usually bilateral; enlarged ovaries with increased ovarian volume ($\geq 10\text{mL}$) and ≥ 20 peripherally displaced small follicles per ovary.	Usually bilateral; enlarged ovaries.	Usually bilateral; enlarged ovaries with increased ovarian volume and peripherally displaced small follicles.

Table 2: Differential diagnosis table for massive ovarian edema.

ABBREVIATIONS

CECT = Contrast-Enhanced Computed Tomography
 CT = Computed Tomography
 IVC = Inferior Vena Cava
 MRI = Magnetic Resonance Imaging
 USG = Ultrasonography

KEYWORDS

Massive ovarian edema; IVC web; Budd Chiari syndrome; abdominal pain; Ultrasound; Magnetic resonance imaging; Contrast-enhanced CT

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