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Monochorionic Monoamniotic Twin Pregnancy Complicated by Intrahepatic Cholestasis of Pregnancy and Significant Inter-Twin Growth Discordance: A Rare Case Report

[1]Dr. Ruby Bhatia, Professor & HOD, Department of Obstetrics & Gynecology Maharishi Markandeshwar Institute of Medical Sciences and Research, Abmala

[2]Dr. Sheena Manktala, Resident, Department of Obstetrics & Gynecology Maharishi Markandeshwar Institute of Medical Sciences and Research, Abmala

[3]Dr. Mahak Singhal, Assistant Professor, Department of Obstetrics & Gynecology Maharishi Markandeshwar Institute of Medical Sciences and Research, Abmala

[4]Dr. Vidushi Tewari, Assistant Professor, Department of Obstetrics & Gynecology Maharishi Markandeshwar Institute of Medical Sciences and Research, Abmala

[5]Dr. Himanshi Pareek, Senior Resident, Assistant Professor, Department of Obstetrics & Gynecology Maharishi Markandeshwar Institute of Medical Sciences and Research, Abmala

Corresponding Author: Dr. Sheena Manktala, Resident, Department of Obstetrics & Gynecology Maharishi Markandeshwar Institute of Medical Sciences and Research, Abmala.

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Abstract

Background: Monochorionic monoamniotic (MCMA) twin pregnancies constitute one of the rarest and highest-risk subtypes of twin gestation, occurring in approximately 1% of monozygotic twin pregnancies and in fewer than 0.1% of all pregnancies. These gestations carry an inherently elevated risk of adverse perinatal outcomes, including cord entanglement, congenital anomalies, and intrauterine fetal demise. This risk profile is further compounded when intrahepatic cholestasis of pregnancy (IHCP) coexists with significant inter-twin growth discordance, creating a uniquely complex clinical scenario with limited evidence-based guidance.

Case Presentation: We report the case of a 24-year-old Gravida 3 para 2 (G3P2L2) woman presenting at 34 weeks and 2 days of gestation with an established diagnosis of MCMA twin pregnancy, complicated by IHCP and 33% inter-twin fetal weight discordance. The patient presented with generalised pruritus and was subsequently found to have markedly (232 micromol/L) elevated hepatic transaminases (SGOT 114 U/L; SGPT 87 U/L; ALP 875 U/L).

Ultrasonography confirmed both foetuses in cephalic presentation, with estimated fetal weights of 1,767 g (Twin A) and 1,130 g (Twin B), yielding a discordance of 33%. Umbilical artery Doppler velocimetry remained

within normal parameters. Intensive antenatal surveillance was instituted, comprising serial non-stress tests and biochemical monitoring.

Pharmacological management with ursodeoxycholic acid (UDCA) was initiated for IHCP, and a complete course of antenatal corticosteroids was administered to promote foetal pulmonary maturation. Due to the development of acute foetal distress, an emergency lower-segment caesarean section was performed at 34 weeks and 2 days of gestation.

Outcome: Both neonates were delivered with satisfactory Apgar scores and were subsequently admitted to the neonatal intensive care unit (NICU) for management of prematurity-related complications. The postoperative maternal course was uneventful and babies had NICU stay for observation and later discharged in stable condition.

Conclusions: This case illustrates the compounded perinatal risks and multifaceted clinical challenges encountered in the management of MCMA twin gestation concurrently complicated by IHCP and significant fetal growth discordance. The successful outcome underscores the critical importance of early diagnosis, multidisciplinary surveillance, timely corticosteroid administration, and judicious timing of delivery to optimise both maternal and neonatal outcomes in this rare obstetric constellation.

Keywords: Monochorionic Monoamniotic Twins; Intrahepatic Cholestasis of Pregnancy; Inter-Twin Fetal Growth Discordance; High-Risk Obstetrics; Preterm Delivery; Neonatal Intensive Care.

Introduction

Monochorionic monoamniotic (MCMA) twin pregnancies arise when a single fertilized oocyte undergoes embryonic division between days 8 and 13 post-fertilisation, resulting in both foetuses occupying a shared chorion and a common amniotic cavity without an inter-twin membrane [1]. The reported incidence ranges from 1 in 10,000 to 1 in 35,000 pregnancies, representing approximately 1% of monozygotic twin gestations and fewer than 0.1% of all pregnancies [2]. The absence of an inter-twin membrane predisposes these pregnancies to a constellation of serious complications, most notably umbilical cord entanglement, which occurs in virtually all MCMA pregnancies and constitutes the principal aetiology of perinatal mortality in this group [3]. Additional complications include twin-to-twin transfusion syndrome (TTTS), congenital structural anomalies, and an elevated risk of intrauterine fetal demise, contributing to reported perinatal mortality rates as high as10–15% [2,3].

Intrahepatic cholestasis of pregnancy (IHCP), also termed obstetric cholestasis, is a pregnancy-specific hepatobiliary disorder characterised by pruritus, elevated serum bile acid concentrations, and deranged hepatic transaminases, typically presenting in the late second or third trimester [4]. Its global prevalence varies widely from 0.2% to 2% in most Western populations—though it is significantly higher in twin gestations due to amplified oestrogen-mediated hepatic dysfunction [6]. IHCP is associated with a spectrum of adverse fetal outcomes, including spontaneous preterm birth, fetal cardiac arrhythmias, meconium-stained amniotic fluid, and sudden intrauterine death, the latter risk being disproportionately elevated when total serum bile acids exceed 40 μmol/L [4,5].

Inter-twin fetal growth discordance, conventionally defined as a ≥20% difference in estimated fetal weight (EFW) between co-twins, is observed in approximately 15–20% of twin gestations and is recognised as an independent risk factor for adverse neonatal outcomes, including preterm birth, respiratory distress syndrome, intraventricular haemorrhage, and neonatal death [7].

In the specific context of mono chorionic twin pregnancies, growth discordance most frequently results from unequal placental territory allocation and haemodynamic perturbations arising from inter-twin vascular anastomoses [8].

The coexistence of MCMA twin gestation, IHCP, and significant inter-twin growth discordance within a single clinical case is exceptionally rare, and no established evidence-based management algorithm currently addresses this precise constellation of complications. Both IHCP and growth discordance independently necessitate intensive antenatal surveillance and carry implications for the timing of delivery, yet their concurrent occurrence in the already high-risk background of MCMA twins creates a uniquely challenging clinical scenario [9,10]. We herein present such a case, along with a synthesis of the relevant literature and a discussion of our management strategy.

Case Presentation

A 24-year-old woman (G3P2L2) with a confirmed MCMA twin pregnancy was referred to our department at 34 weeks and 2 days of gestation. She reported progressive generalized pruritus of increasing severity, with no rash, commencing during the third trimester. Her obstetric history was notable for two uncomplicated, spontaneous, full-term vaginal deliveries of significance, her medical history included a prior episode of jaundice during a previous pregnancy, potentially indicative of a predisposition to IHCP. Her surgical history was otherwise unremarkable, and she had no chronic medical co-morbidities.

On clinical examination, the patient was vitally stable. Fundal height was consistent with a 37 weeks, uterus over distended and obstetric palpation confirmed both foetuses in cephalic presentation with both regular fetal heart sounds.

Laboratory investigations revealed a haemoglobin concentration of 10.7 g/dL, consistent with the physiological anaemia of pregnancy. Hepatic function tests demonstrated significant biochemical derangement: serum glutamic-oxaloacetic transaminase (SGOT) 114 U/L (reference range: 10–40 U/L), serum glutamic-pyruvic transaminase (SGPT) 87 U/L (reference range: 7–56 U/L), and alkaline phosphatase (ALP) 875 U/L (reference range: 44–147 U/L non-pregnant). Serum bile acids were (232 micromol/L) elevated, corroborating a diagnosis of IHCP. Coagulation profile, renal function tests, serum albumin, total bilirubin, and complete blood count were within normal parameters. Viral hepatitis serologies were negative. Laboratory investigations revealed a haemoglobin concentration of 10.7 g/dL, consistent with the physiological anaemia of pregnancy. Hepatic function tests demonstrated significant biochemical derangement: serum glutamic-oxaloacetic transaminase (SGOT) 114 U/L (reference range: 10–40 U/L), serum glutamic-pyruvic transaminase (SGPT) 87 U/L (reference range: 7–56 U/L), and alkaline phosphatase (ALP) 875 U/L (reference range: 44–147 U/L non-pregnant). Serum bile acids were (232 micromol/L) elevated, corroborating a diagnosis of IHCP. Coagulation profile, renal function tests, serum albumin, total bilirubin, and complete blood count were within normal parameters. Viral hepatitis serologies were negative.

Ultrasonographic assessment demonstrated both foetuses in cephalic presentation. Estimated fetal weights were 1,767 g for Twin A (corresponding to the 25th–50th centile for gestational age) and 1,130 g for Twin B (below the 3rd centile for gestational age), yielding an inter-twin EFW discordance of 33%—substantially exceeding the 20% threshold defining clinically significant discordance. The umbilical artery pulsatility

index (PI) for both twins, as assessed by Doppler velocimetry, remained within normal reference ranges. Biophysical profiles were reassuring for both foetuses. Amniotic fluid volume was adequate.

The patient was admitted to the high-risk obstetric unit for fetal heart monitoring, steroid cover for fatal lung maturity and subsequent treatment of pregnancy. Pharmacological management with ursodeoxycholic acid (UDCA) at a dose of 15 mg/kg/day in divided doses was commenced for IHCP. A complete course of antenatal dexamethasone (6 mg intramuscularly at 12 hours 4 doses) was administered to accelerate fetal pulmonary maturation. Continuous electronic fetal monitoring via cardiotocography (CTG) was instituted. Serial non-stress tests remained reactive and reassuring throughout the surveillance period.

At 34 weeks and 4 days of gestation, an acute category-1 change in fetal heart rate patterns was detected on continuous CTG monitoring, consistent with fetal distress. Given this development—superimposed on the background of MCMA twin gestation, IHCP with marked biochemical derangement, and severe inter-twin growth discordance—an emergency lower-segment caesarean section was expeditiously performed under spinal anaesthesia.

Intra-operatively, both foetuses were delivered in a cephalic presentation. Twin A was delivered first with a birth weight of 1,740 g (Apgar scores: 7 at 1 minute, 9 at 5 minutes); Twin B was delivered second with a birth weight of 1,120 g (Apgar scores: 6 at 1 minute, 8 at 5 minutes). Umbilical cord entanglement was confirmed at the time of delivery. Both neonates were transferred to the NICU for management of prematurity-related complications, including respiratory support, temperature regulation, and nutritional supplementation. The placenta appeared mono-chorionic mono-amniotic on gross pathological examination, as expected. Twin A had meconium stained liquor and twin b had clear adequate amount of liquor.

The mother's postoperative course was uncomplicated. Hepatic transaminases demonstrated a progressive falling trend following delivery, consistent with the anticipated biochemical resolution of IHCP. She was discharged on postoperative day 5 in satisfactory condition. Both babes were discharged from NICU on day 8 of life.

Discussion

This case exemplifies an exceedingly rare and clinically challenging combination of MCMA twin gestation, intrahepatic cholestasis of pregnancy, and marked inter-twin growth discordance—each individually associated with significant perinatal morbidity and mortality, and collectively representing a compounded obstetric risk. A systematic review of the relevant literature reveals no previously documented case reports describing this precise triad, underscoring its clinical novelty and the necessity of individualised, multidisciplinary management.

- **MCMA Twin Gestation and Cord Entanglement**

Umbilical cord entanglement is a pathognomonic and virtually universal feature of MCMA twin pregnancies, arising from the unimpeded mobility of both foetuses within a shared amniotic cavity [3]. Perinatal mortality attributable to cord entanglement has historically been reported to be as high as 10–15%, though contemporary series employing intensive inpatient monitoring strategies have demonstrated substantially improved outcomes [1]. Heyborne et al. [1] conducted a landmark retrospective cohort study demonstrating that continuous inpatient fetal heart rate monitoring from 24 to 34 weeks of gestation was associated with a perinatal mortality rate of 4.0%, compared with 28.6% in those managed without intensive surveillance. In the present case, continuous

electronic fetal monitoring enabled the timely identification of acute fetal compromise and facilitated expeditious delivery prior to a catastrophic outcome.

- **Intrahepatic Cholestasis of Pregnancy**

Pathophysiology and Fetal Risk. IHCP is a multifactorial disorder in which elevated circulating oestrogen concentrations—particularly oestrogen sulphate metabolites—impair hepatocanalicular bile acid transport, leading to intra-hepatic accumulation of bile acids and their subsequent entry into the fetal circulation. Twin pregnancies are associated with approximately two-fold higher serum oestrogen concentrations compared to singleton gestations, which may partly explain the increased prevalence of IHCP in multiple pregnancies. Elevated bile acid concentrations exert direct cytotoxic effects on fetal cardiomyocytes, precipitating bradyarrhythmias and potentially fatal cardiac events, as documented by Greene and Williamson. Severity Grading. IHCP is classified by peak total bile acid (TBA) levels: mild (19–39 μmol/L) with low stillbirth risk similar to uncomplicated pregnancies; moderate (40–99 μmol/L) linked to higher preterm birth, meconium passage, and cesarean rates; severe (≥100 μmol/L) carrying the greatest perils, including stillbirth peaking around 36 weeks, neonatal asphyxia, and respiratory distress. Williamson et al. established that intrauterine fetal demise risk rises substantially above 40 μmol/L, directly informing expedited delivery recommendations for moderate-to-severe cases. Management Implications. Ursodeoxycholic acid (UDCA) remains the pharmacological cornerstone of IHCP management across severities; a meta-analysis by Bacq et al. demonstrated significant improvements in maternal pruritus and hepatic parameters, plus potential fetal benefit, as observed here. In this MCMA twin case, severity escalation prompted intensified monitoring and timely intervention.33 Inter-Twin Growth Discordance in Monochorionic Gestations. Fetal growth discordance in monochorionic twin pregnancies differs pathophysiologically from that observed in dichorionic twin gestations, primarily due to the role of inter-twin vascular anastomoses (arteriovenous, arterio-arterial, and venovenous) in mediating haemodynamic imbalance [8]. Unequal placental sharing, which is uniquely prevalent in monochorionic placentation, further compounds discordant growth trajectories. Lewis et al.[8] demonstrated that the magnitude and directionality of vascular anastomoses are major determinants of the degree of growth discordance in monochorionic twins, with selective intrauterine growth restriction (sIUGR) representing a clinically significant and distinct complication of this placental architecture. Hiersch et al.[7] further demonstrated that inter-twin growth discordance ≥20% constitutes an independent predictor of adverse neonatal outcomes, including prematurity-related morbidity, irrespective of Doppler findings. In the present case, despite normal umbilical artery Doppler velocimetry, the degree of discordance (33%) warranted heightened surveillance and contributed substantively to the decision for expedited delivery.

- **Timing and Mode of Delivery**

Current evidence-based guidelines from the American College of Obstetricians and Gynecologists (ACOG) [12] and the Society for Maternal-Fetal Medicine (SMFM)[13] recommend planned delivery of uncomplicated MCMA twin gestations at 34 weeks of gestation, given the exponentially increasing risk of intrauterine fetal demise attributable to cord entanglement beyond this gestational age. This threshold is further supported by data demonstrating that the perinatal mortality associated with expectant management after 34 weeks outweighs the neonatal morbidity attributable to iatrogenic prematurity.

In the present case, the additional compounding risks of IHCP and significant growth discordance conferred further urgency to delivery at 34 weeks, and the acute development of fetal distress rendered emergency caesarean section the only appropriate modality. Caesarean section is universally recommended for MCMA twins due to the technical and logistical difficulties in safely achieving vaginal delivery without precipitating acute cord compromise.

- **Role of Antenatal Corticosteroids**

The administration of antenatal corticosteroids to patients at risk of late preterm birth is supported by Level I evidence. The landmark randomised controlled trial by Gyamfi-Bannerman et al.[14] demonstrated that corticosteroids administered between 34 0/7 and 36 6/7 weeks of gestation significantly reduced the composite outcome of respiratory morbidity in late preterm infants, including the need for respiratory support and neonatal ICU admission. In the present case, the timely administration of corticosteroids prior to emergency delivery likely contributed to the relatively satisfactory Apgar scores and neonatal respiratory outcomes observed in both infants.

Intrahepatic cholestasis of pregnancy (IHCP) heightens the risk of other serious obstetric complications, notably preeclampsia and gestational diabetes mellitus (GDM), compounding the challenges in high-risk cases like MCMA twins. Multiple studies confirm these associations, with IHCP patients facing 2-4 times higher odds of GDM and up to 5 times the risk of preeclampsia compared to unaffected pregnancies.

Bile acid excess damages placental vessels via vasoconstriction, oxidative stress, and syncytial knot formation, impairing antioxidant defenses and oxygen delivery. This spurs vasoactive factors like sFlt-1 and endoglin, mirroring preeclampsia's endothelial dysfunction; preeclampsia often emerges 2-4 weeks post-IHCP diagnosis. Shared dyslipidemia and inflammation amplify vascular risks.

Preeclampsia risk escalates particularly with severe IHCP (bile acids >40 μmol/L) or early onset, often manifesting 2-4 weeks post-IHCP diagnosis and preceding hypertension via proteinuria. This link persists across populations, including twins, urging vigilant bloodpressure and urine protein monitoring alongside IHCP management.

GDM occurs more frequently regardless of IHCP severity, even in mild cases, likely tied to shared metabolic disruptions from elevated estrogens in multiples. Routine screening at 24-28 weeks remains essential, with IHCP as an independent risk factor (adjusted OR~1.4-3), further justifying multidisciplinary surveillance in this case.

High sulfated progesterone metabolites in IHCP antagonize FXR (farnesoid X receptor), a bile acid sensor that curbs gluconeogenesis and boosts insulin sensitivity; this leads to impaired glucose tolerance, higher postprandial glucose, and reduced GLP-1 (an insulin-promoting incretin via TGR5 receptor). Result: IHCP women show dysglycemia (e.g., 30% GDM rate vs. 0% controls), elevated triglycerides/LDL, and low HDL during continuous monitoring. UDCA may partially restore GLP-1, easing effects. These comorbidities amplify perinatal risks like preterm birth and stillbirth, reinforcing the need for expedited delivery and holistic care as applied here. Integrating IHCP with preeclampsia/GDM vigilance enhances outcomes in such rare triads.

Conclusion

We present an exceptionally rare case of MCMA twin pregnancy complicated concurrently by intrahepatic cholestasis of pregnancy and significant inter-twin fetal

growth discordance, managed successfully through intensive antenatal surveillance, pharmacological intervention, and timely surgical delivery. This case reinforces several critical principles in high-risk obstetric management: the indispensable role of continuous electronic fetal monitoring in MCMA gestations; the importance of early biochemical diagnosis and UDCA therapy in IHCP; the heightened clinical significance of inter-twin growth discordance in the context of monochorionic placentation; and the necessity of integrating multiple compounding risk factors into an individualised delivery timing strategy. The successful neonatal outcome in this case was contingent upon a coordinated multidisciplinary approach encompassing obstetrics, neonatology, and Anaesthesiology. Further prospective data collection and case series reporting are warranted to inform the development of standardised management guidelines for this rare and complex obstetric scenario.

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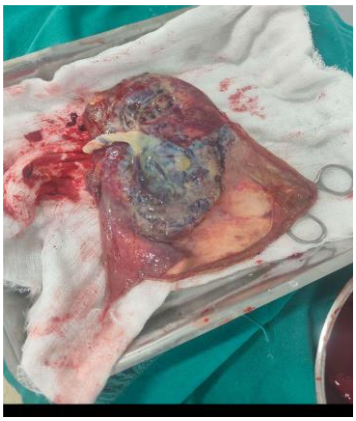
Legend Figures



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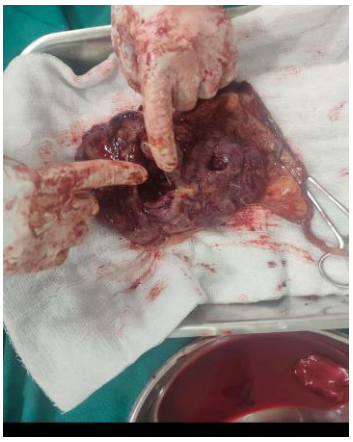

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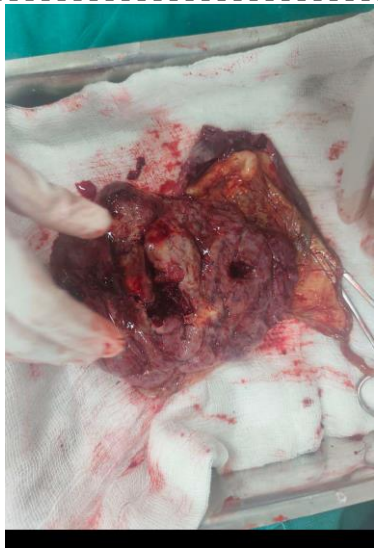

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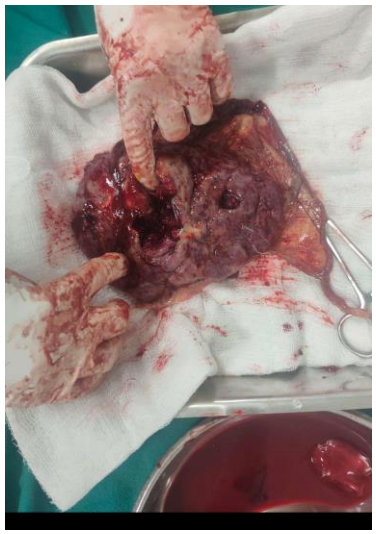

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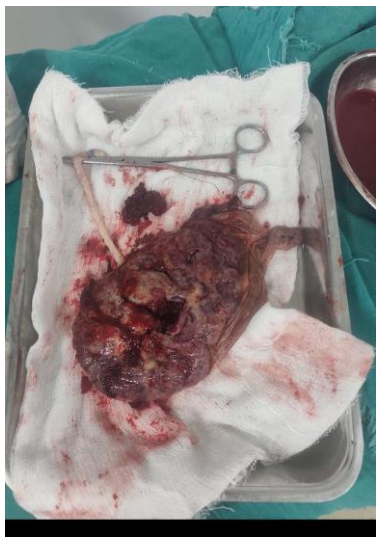

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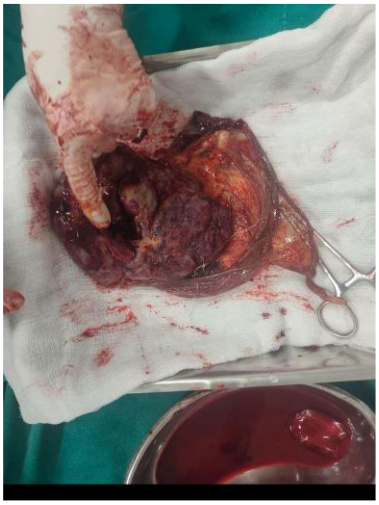

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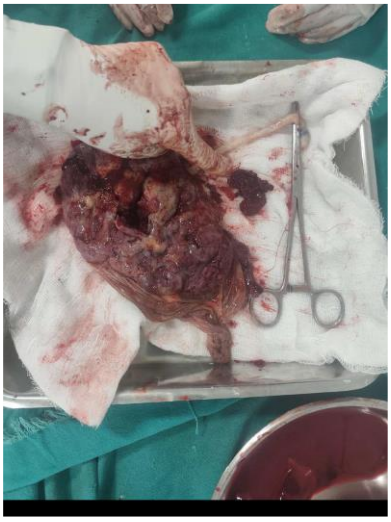

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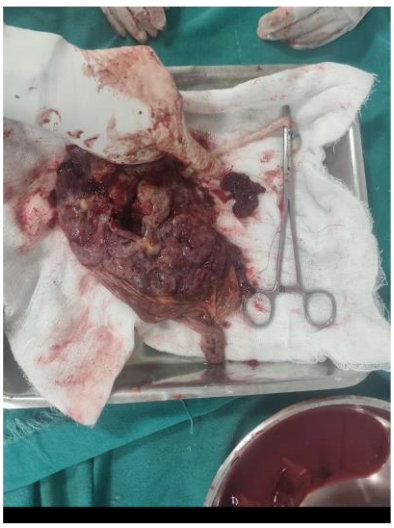

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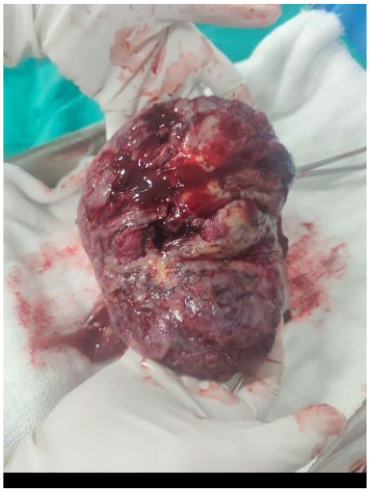

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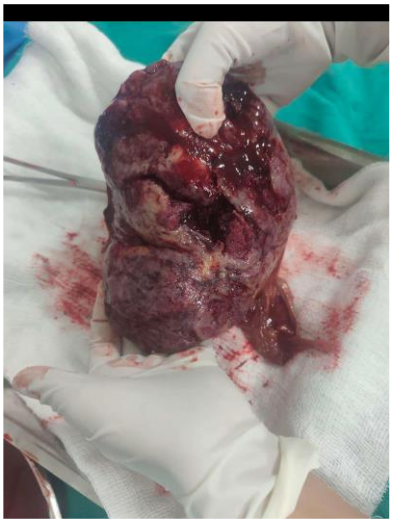

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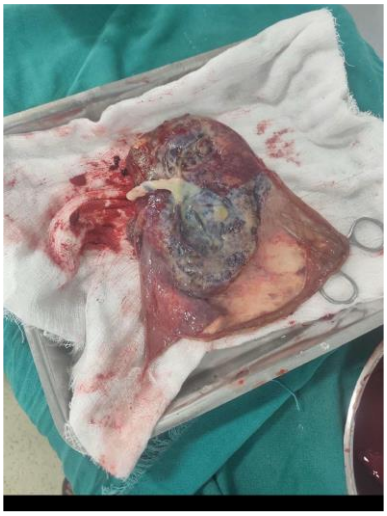

Figure 11: