POLYCYSTIC LIVER DISEASE IN PREGNANCY:
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Abstract

Introduction: Polycystic liver disease (PLD) in pregnancy, an uncommon disorder in daily practice is associated with severe morbidity and often results in mortality. The disease is common among the elderly and a rare entity among women of reproductive age group. We present a report of a young woman that presented with polycystic liver disease in pregnancy, managed conservatively to term and celebrated with a live foetus.

Key words: polycystic liver, Pregnancy, Preterm, life foetus.

INTRODUCTION

Mrs A. U was a 36 year old Gravida 4 Para 1+2 one alive, who reported at antenatal clinic at 19 weeks gestation. She complained of abdominal pain of 4 years duration which had worsened since conception. She was diagnosed with polycystic liver disease 4 years prior to conception and was being managed by a gastroenterologist. There was no history of jaundice. She had been on intermittent tramadol for pain relief for the past one year. Mrs A. U's mother was diagnosed with polycystic liver disease about 16 years earlier. Her first and second pregnancy ended in spontaneous first trimester miscarriages. Her third pregnancy was uneventful, it was carried to term and she had a vaginal delivery of a female baby 7 years prior to presentation.

She was well nourished. She was neither febrile nor pale. She was anicteric and had no pedal oedema. Her weight and height were 113.6kg and 1.8m respectively. Her pulse and blood pressure were normal. The abdomen was enlarged with moderate tenderness on the right hypochondrium. The liver was enlarged, tender, 24cm below the right coastal margin with a span of 33cm. The uterus was displaced to the left by the enlarged liver. The symphysiofundal height was 20cm which was compatible with her gestational age of 19 weeks.

An abdominal ultrasound scan done at booking showed a single live intra-uterine foetus at 19 weeks with a fundal placenta and adequate amniotic fluid. The liver was enlarged and contained multiple parenchymal anechoic cystic lesions of varying sizes consistent with PLD. The intrahepatic and vascular channels were distorted by the cysts. Other abdominal organs were normal (Figure 1).

The liver function tests showed mildly elevated total and direct bilirubin while the other parameters were normal. The full blood count, renal function tests, and clotting profile were all within normal limit. The oral glucose tolerance test was normal.

A diagnosis of polycystic liver disease in pregnancy was made. She was placed on routine antenatal medications and received three doses of sulphadoxine-pyrimethamine. Pentazocine injection was administered intermittently for control of pain. She was counselled for bed rest at home.

She was seen weekly at the antenatal clinic until 30 weeks when she had marked abdominal distension with pain. There were no uterine contractions, vaginal bleeding or drainage of liquor. She was afebrile, not pale, anicteric and there was no pedal oedema. The liver at this point was 36cm below the

coastal margin, the spleen was not enlarged and kidneys were not ballotable. There was no ascites. She was admitted and co-managed with the Gastro-enterologist and placed on intramuscular pentazocine 60 mg 8 hourly for 48 hours and a course of dexamethasone to accelerate lung maturity.

She remained stable on admission until at 35 weeks gestation when abdominal distension and discomfort became unbearable. The liver was then found to have extended to the right iliac fossa (39cm below the coastal margin). The uterus was pushed to the left hypochondrium and the symphysiofundal height was 38cm. The foetal lie was longitudinal and the presentation was breech. She had a caesarean section at 35 weeks gestation. A live female baby with Apgar scores of 8 and 9 in the first and fifth minutes respectively was delivered with a birth weight of 3.2 kg. The abdominal cavity was mainly filled with the enlarged multi-cystic liver, with each cyst containing clear fluid. There was no ascites (Figure 2).

Post operatively she had 1 litre of intravenous dextrose saline 8 hourly for 24 hours with intravenous augmentin 1.2gm 12 hourly for 48 hours and 625mg bd orally for 5 days thereafter. She also had intramuscular pentazocine 60mg 8 hourly for 48 hours and thereafter oral diclofenac 50mg bd for 5 days. She felt a lot of relief of the abdominal distension and her vital signs remained stable. She was discharged on the 4th day post-operative. She was seen at the post-natal clinic at 2 and 6 weeks and was found to be stable with her baby. The liver size however, remained unchanged but she had relief from abdominal discomfort and pain. She was counselled on contraception and had an intrauterine contraceptive device (IUCD) inserted. She was referred back to the gastroenterologist for further management.

Discussion

Polycystic liver disease (PLD) is an autosomal dominant condition which is commonly seen in females, usually in their fifth decades of life and its coexistence with pregnancy is rare. No case appeared to have been reported in Africa with only three cases ever reported in literature to the best of the researchers' knowledge, from USA, Australia and Spain respectively.^{2,3,4} This underscores the importance of this case report in adding to the body of literature on the subject matter.

The main risk factor for polycystic liver disease is having a family member with the condition due to its Autosomal dominant pattern of inheritance⁵ consistent with Mrs A. U whose mother had a similar disease.

She was referred to us by the Gastroenterologist as a case of polycystic liver disease in pregnancy with history of

abdominal distension, positive family history of polycystic liver disease and an enlarged liver on physical examination. This was confirmed by an abdominal ultrasound scan which showed a markedly enlarged liver with multiple parenchymal anechoic cystic masses coexisting with intrauterine pregnancy. The kidneys were normal, hence the diagnosis of isolated polycystic liver disease. A Magnetic Resonance Imaging (MRI) was considered but this was unaffordable for our patient.

The clinical course is said to be benign as the hepatic structure is conserved. Mrs A.U however had mildly elevated liver enzymes but she was not jaundiced. She also had abdominal pain and discomfort perhaps, due to gross abdominal distension from the hepatic enlargement and pregnancy consistent with findings of previous literatures. 2,3,4 The foetal presentation was breech probably due to uterine displacement by the enlarged liver. Vaginal delivery would have been considered if the presentation was cephalic. The three previous reports all had vaginal deliveries. 2,3,4 The deliveries occurred at term in two of the cases 2,4 while preterm birth at 35 weeks occurred in one as found in our patient 3.

The complications of polycystic liver disease include cyst rupture, bleeding into the cyst, infection, bile duct obstruction and jaundice. 4,5 None of these occurred in our patient. The general physiologic changes of pregnancy may aggravate PLD in addition to direct competition for space with the gravid uterus. This was seen in our patient whose symptoms worsened with the pregnancy and the uterus was pushed to the left by the enlarged liver. Female steroid hormones are known to cause enlargement of the cysts 4 and hence, hormonal contraceptives were avoided in this patient to prevent further enlargement.

Treatment generally involves cyst aspiration combined with ultrasound guided application of sclerosants, cyst fenestration and liver resection. 3,4,5 These were not contemplated in our patient as they are mainly procedures of gastroenterologist preferably outside pregnancy. Perhaps, cyst aspiration during pregnancy may further worsen pregnancy condition as the degree of intra peritoneal bleeding may be unpredicted necessitating premature laparotomy. The only known cure for PLD is liver transplant when all other measures have failed. 4,5 Following an uneventful puerperium, she was referred back to the gastroenterologist to continue management.

Polycystic liver disease in pregnancy is indeed rare and a high index of suspicion as well as multi-disciplinary approach may improve pregnancy outcome.

Figures 1 and 2 are ultrasound prints showing the multiple hepatic cysts and portion of the foetal head adjacent to cystic liver.

Figure 1

Figure 2

Figure 3 shows arrows pointing at the lower edge of the polycystic liver during caesarean section.

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