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ONLINE CASE REPORT

Twin Pregnancy with a Complete Hydatidiform Mole and a Coexisting Live Fetus

Rare entity

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الحمل التوأم بالحمل العنقودي الكامل مع جنين حي

حالة نادرة

شهيلة الشيخ، نهال الريامية، ناميثا راشيل ماثيو، راشد السكيتي، عاصم قريشي، مريم ماثيو

ABSTRACT: A hydatidiform mole with a coexisting live fetus is a rare occurrence and the optimal management for this condition is not yet known. We report the case of a 32-year-old woman (*gravida* 3, *para* 2) who presented to the Sultan Qaboos University Hospital, Muscat, Oman, in March 2012 at 13 gestational weeks with abdominal pain and vaginal bleeding. An ultrasound examination revealed a hydatidiform mole pregnancy coexisting with a live fetus. After extensive counselling, the patient and her husband opted for a conservative management approach. Unfortunately, a hysterotomy had to be performed at 17 gestational weeks due to severe haemorrhage. The post-operative period was uneventful and histopathology results confirmed one complete mole with a coexisting fetus and normal placenta. The patient's serum β-human chorionic gonadotropin level remained normal for 18 months following her surgery.

Keywords: Hydatidiform Mole; Twin Pregnancy; Fetus; Hysterotomy; Placenta; Human Chorionic Gonadotropin; Case Report; Oman.

الملخص: الحمل العنقودي الكامل مع جنين حي هو حالة نادرة الحدوث والعلاج الأمثل لهذه الحالة غير معروف حتى الآن. نحن نبلغ عن حالة لإمرأه في 32 من عمرها (في حملها الثالث وولادتين سابقتين) والتي جاءت إلى مستشفى جامعة السلطان قابوس في مسقط، عمان، في مارس 2012 في الإسبوع 13 من الحمل بالآم في البطن ونزيف مهبلي.الأشعة الفوق الصوتية أوضحت حمل عنقودي مع جنين حي. وبعد المشورة واسعة النطاق اختارت المريضة وزوجها المحافظة والإستمرار في الحمل. للأسف كان لابد من إجراء عملية إنزال للجنين في الأسبوع 17 من الحمل عن طريق فتحة في الرحم بسبب نزيف حاد. كانت فترة ما بعد الجراحة خالية من الأحداث وأكد التشريح المخبري حمل توأم بحمل عنقودي واحد وجنين آخر مع مشيمة طبيعية. ظل مستوى هرمون الحمل طبيعي لفترة 18 شهر بعد العملية.

مفتاح الكلمات: حمل عنقودي؛ حمل توأم؛ جنين؛ فتح الرحم؛ مشيمة؛ هرمون الحمل؛ تقرير حالة؛ عمان.

A hydatidiform mole coexisting with a normal live fetus in a twin pregnancy is an extremely rare phenomenon, with a worldwide incidence ranging from one in 22,000 to one in 100,000 pregnancies.[1,2] This condition presents a significant management challenge for physicians due to its associated complications, including heavy vaginal bleeding, severe pre-eclampsia, intrauterine fetal death or growth restriction, miscarriage, preterm birth and hyperthyroidism, as well as ovarian cyst rupture or torsion in cases with *theca* lutein cysts.[1] Twin molar pregnancies may also have an increased risk of persistent trophoblastic disease compared to a single molar pregnancy.[2]

Case Report

A 32-year-old *gravida* 3, *para* 2 woman was admitted to the Emergency Department of the Sultan Qaboos University Hospital (SQUH), Muscat, Oman, in March 2012 at 13 gestational weeks with lower abdominal pain and vaginal spotting. Upon examination, she was haemodynamically stable and not in distress. Abdominal palpation indicated mild tenderness and a uterine size corresponding to 16 gestational weeks. An ultrasound examination revealed a twin pregnancy, including one normal fetus with the placenta covering the cervical opening and one molar pregnancy [Figure 1A]. Both ovaries were enlarged with multiple *theca* lutein cysts; the right ovary measured 12.2 x 8.1 x 6.9 cm while the left measured 13.3 x 7.3 x 10.6 cm [Figure 1B]. The patient's serum β-human chorionic gonadotropin (hCG) level was 1,386,570 IU/L. Chest X-ray findings were normal as were the results of the thyroid function tests and coagulation profile.

The patient and her husband were counselled in detail regarding the risks associated with this type of pregnancy; opinions from international experts

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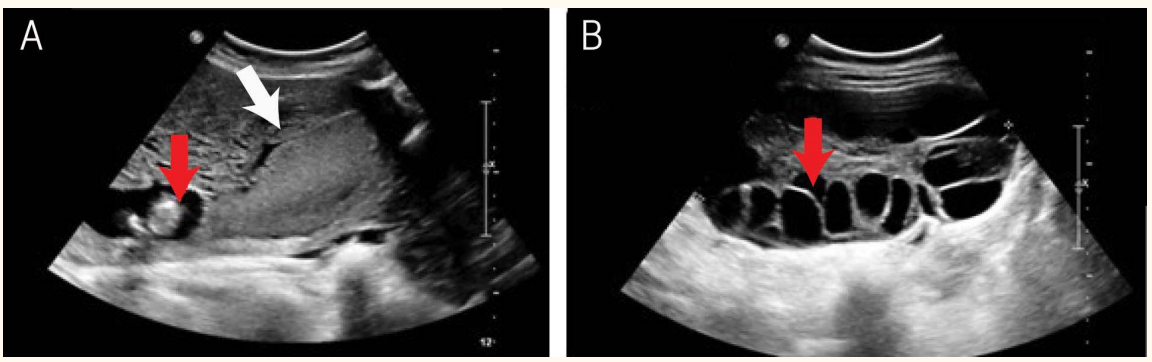

Figure 1A & B: Ultrasound scans of a patient with a complete hydatidiform mole and coexisting live fetus showing **(A)** the junction between the normal placenta and the molar tissue (white arrow) and the normal fetus in the gestational sac (red arrow) and **(B)** the enlarged left ovary with multiple *theca* lutein cysts (red arrow).

were elicited and the couple was informed that a review of the literature indicated a 25% chance of a live birth and a 40% chance of early fetal loss.[3] A multidisciplinary team including a consultant haematologist, an anaesthetist, an interventional radiologist and several obstetricians were involved in the decision-making process. After consideration, the couple opted to continue the pregnancy with close follow-up and the patient was discharged after three days.

Despite close follow-up, the patient was readmitted at 17 gestational weeks with vaginal bleeding and tachycardia. A significant drop in haemoglobin from 10 g/dL to 7 g/dL was also noted, requiring a transfusion of two units of packed red blood cells. At this time, the uterine size corresponded to 28 gestational weeks. The couple were advised that the maternal risks had now increased significantly and they agreed to have the pregnancy terminated. Bilateral uterine artery balloon catheters were placed prophylactically and inflated to control the intraoperative blood loss; this provision would also enable the postoperative embolisation of the uterine arteries if required.

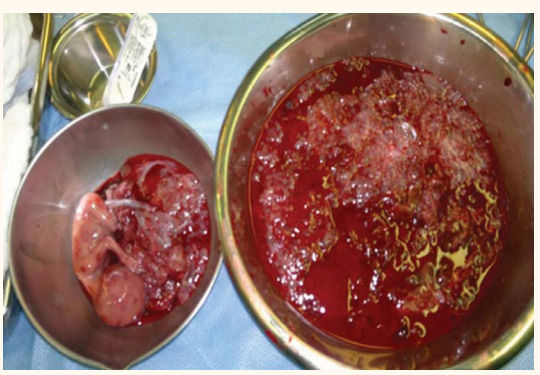

Figure 2: Photograph of the normal fetus and placenta and the molar tissue following a hysterotomy in a pregnant patient with a complete hydatidiform mole and a coexisting live fetus.

A hysterotomy was performed under general anaesthesia via a lower segment transverse incision. Molar tissue weighing 1,040 g was evacuated, along with a fetus weighing 105 g and a normal placenta of 65 g [Figure 2]. The ovaries were enlarged with bilateral *theca* lutein cysts. The intraoperative blood loss was 800 mL. The uterine artery balloon catheters were removed six hours after the operation as there was no active bleeding and embolisation was not deemed necessary. The postoperative recovery period was uneventful.

Histopathology results confirmed a complete hydatidiform mole with a coexisting fetus and normal placenta [Figure 3]. Three months after the surgery, the patient's serum β-hCG level had returned to normal without any cytotoxic therapy. The enlarged ovaries with multiple *theca* lutein cysts took six months to return to a normal size. There was no evidence of any persistent trophoblastic disease one year later and the serum β-hCG level remained normal for 18 months following the surgery. At the time of writing, the patient was planning her next pregnancy.

Discussion

Complete hydatidiform moles form part of the tumour spectrum of gestational trophoblastic disease. Molar pregnancies are subdivided into either complete or partial types based on genetic and histological features. A complete hydatidiform mole consists of oedematous villous tissue and trophoblastic hyperplasia with no fetal tissue.[1,3] While a complete mole is diploid, its chromosomes are entirely of paternal origin, following the duplication of a single sperm fertilising an empty ovum.[1] However, the majority of partial moles are triploid in origin and contain two sets of paternal and one set of maternal haploid genes. This occurs following the fertilisation of a normal ovum by two sperm cells. In a partial mole, a fetus or fetal red blood

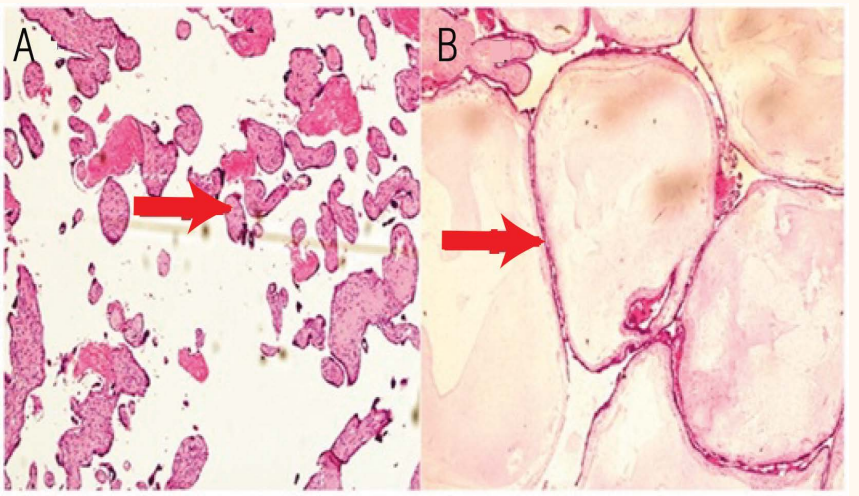

Figure 3A & B: Haematoxylin and eosin stains of the molar tissue showing (**A**) normal trophoblasts (arrow) at x10 magnification and (**B**) avascular oedematous villi (arrow) in a pregnant patient with a complete hydatidiform mole and a coexisting live fetus.

cells are evident.[2] The current patient is the first case of a complete hydatidiform mole coexisting with a live fetus at SQUH. A conservative approach was initially deemed possible due to the availability of resources and high standard of care. However, close monitoring of the patient and timely intervention were crucial.

The incidence of gestational trophoblastic disease is higher among women of Asian, West African and South American origin, compared to those from Europe and North America.[4] It is normally seen in the very young and elderly. A previous molar pregnancy increases the chance of a repeat molar pregnancy by 1–3%.[4] However, with two previous molar pregnancies, the risk goes up to almost 15%.[4] Aside from ethnicity, none of the risk factors mentioned above were relevant to the patient described in the current report.

Twin pregnancy with a complete hydatidiform mole and a coexisting live fetus is also known as sad fetus syndrome, because the coexisting fetus is usually chromosomally normal and potentially viable.[5,6] In the present case, amniocentesis could not be performed for the purposes of fetal karyotyping as the molar tissue was covering the entire anterior wall of the uterus, pushing the live fetus into a posterior position. The pregnancy had to be terminated due to haemorrhage, which is a known and potentially fatal complication associated with this condition.

Sebire *et al*. assessed 77 twin pregnancies with complete hydatidiform moles and healthy co-twins to ascertain the risk of continuing the pregnancy as opposed to termination.[7] They concluded that there was a 40% chance of a live birth with a very low chance of serious obstetric complications, including the development of persistent gestational trophoblastic disease, if the pregnancy was continued.[7] In the past decade, clinicians have supported the option of conservation, providing that the patient remains under strict hospital-based observation and follow-up.[8,9] With the current medical facilities available, most expected complications can be diagnosed early and managed appropriately. Although the risk of persistent trophoblastic disease cannot be excluded in cases of conservative management, this risk does not seem to increase with gestational age.[10,11] Conservative management in a tertiary care environment is indicated if a multi-disciplinary team is present and able to quickly respond in the event of an emergency. Close surveillance of the patient is also recommended to detect early signs of maternal and fetal complications. Termination of the pregnancy should be considered only if the patient develops severe pre-eclampsia, thyrotoxicosis or significant vaginal bleeding, or if there is evidence of trophoblastic embolisation.[12]

Measures to minimise blood loss during surgery for complete hydatidiform twin molar pregnancies with live co-twins are not described in the literature, although torrential haemorrhage is an expected complication. As such, the authors of this report strongly recommend preoperative placement of bilateral uterine artery balloon catheters for patients at centres with interventional radiology facilities. This will assist in decreasing the intra- and postoperative blood loss and minimise the need for blood and blood product transfusions.

Conclusion

In twin pregnancies with one complete hydatidiform mole, the likelihood of delivering a healthy viable fetus is low. This type of pregnancy creates a

significant management dilemma for the treating physician, especially if the pregnancy was planned. Comprehensive counselling, close follow-up and timely intervention are imperative during the conservative management of this rare clinical entity.

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