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Low-set umbilicus in a pregnant woman with bladder pseudoexstrophy: A case report

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ABSTRACT

Bladder pseudoexstrophy is a rare form of bladder exstrophy. Bladder exstrophy occurs in 1 in every 30,000 to 50,000 live births. Because bladder pseudoexstrophy is asymptomatic, it may be undiagnosed even in adulthood.

A 31-year-old woman with uterus bicornis unicollis and a low-set umbilicus underwent emergency cesarean delivery during the 37th week of pregnancy for chorioamnionitis. Perioperatively, asymptomatic anatomical abnormalities were identified, which included separated rectus abdominis muscles and diastasis of the symphysis pubis. The urinary tract was normal. The patient was diagnosed with bladder pseudoexstrophy.

A low-set umbilicus may be a potential marker for the diagnosis of bladder pseudoextrophy.

1. Introduction

Bladder exstrophy is a rare congenital malformation that occurs in 1 in every 30,000 to 50,000 live births [1]. Pseudoexstrophy, first described by Hejitmancik et al. in 1954 [2], is a rare type of bladder exstrophy. Bladder exstrophy is a malformation that results from the failure of the mesoderm to fuse in the midline during fetal life, with abnormal positioning of the umbilicus and various degrees of separation of the rectus abdominis and pubic symphysis. Unlike bladder exstrophy, which is associated with external urogenital malformations, patients with pseudoexstrophy have normal external genitalia. Separation of the rectus abdominis muscle and pubic symphysis is often asymptomatic and there are no urinary tract problems [3,4]. However, pseudoexstrophy is associated with an increased risk of miscarriage and premature birth due to co-existent uterine malformations requiring diagnosis and management during pregnancy and delivery [5].

A case of pseudoexstrophy diagnosed postpartum in a woman with a low-set umbilicus is reported.

2. Case Presentation

A 31-year-old woman with a history of two spontaneous miscarriages was diagnosed with uterus bicornis unicollis and hyperprolactinemia. The location of her umbilicus was not noted. Spontaneous pregnancy was achieved with cabergoline. She was diagnosed with gestational diabetes mellitus at 31 weeks of gestation and received dietary glycemic control. No abnormalities in fetal growth were observed.

A low-set umbilicus, located at two fingers above her pubic bone, had been noted since early pregnancy (Fig. 1). Transabdominal ultrasound scanning revealed the bladder directly below the umbilicus.

The patient experienced spontaneous rupture of membranes at 37 weeks and 5 days of gestation, resulting in labor but not delivery. Approximately 16 h later, her temperature was 38 °C, she was tachycardic and her white cell count was elevated. She was diagnosed with clinical chorioamnionitis. She therefore underwent emergency cesarean delivery.

To avoid bladder injury, cesarean section was performed via a lower transverse abdominal incision at the height of one finger above the umbilicus. The subcutaneous tissue in her midsection was thin, fascia was observed just below the skin incision, and the rectus abdominis muscle was spontaneously separated just below the transverse incision in the fascia. The fascia and mural peritoneum were fused in the defect of the rectus abdominis muscle, and only fasciotomy was required to reach the abdominal cavity. The fetus was delivered through a transverse incision in the lower uterine segment. The fetus was a female weighing 2286 g with an Apgar score of 8 at 5 min after birth and an umbilical artery blood gas pH of 7.310. No malformation or infection was detected in the neonate. Postoperative abdominal radiography showed diastasis of the pubic symphysis (Fig. 2).

Pseudoexatrophy was diagnosed postoperatively and the

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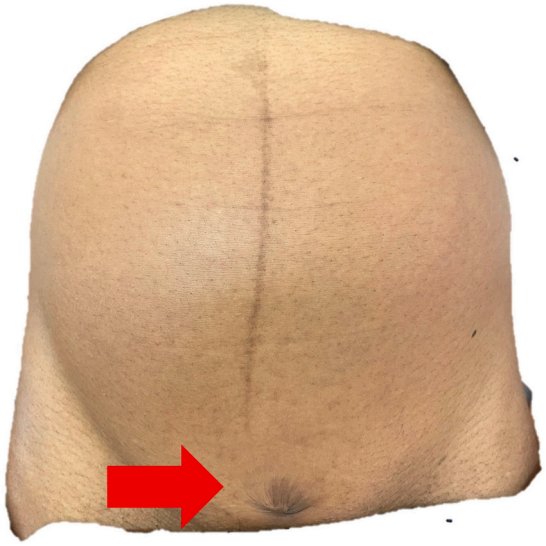

Fig. 1. Patient's abdomen at 35 weeks of gestation. The umbilicus is slightly cranial to the pubic symphysis.

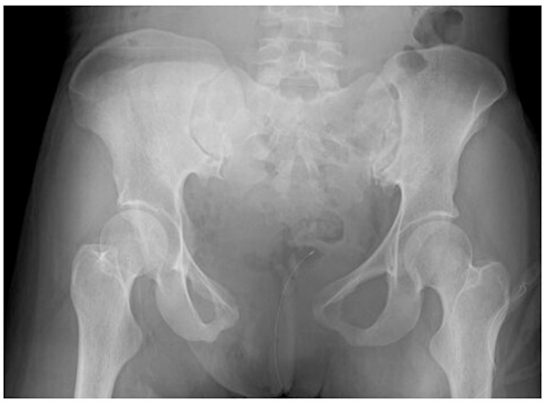

Fig. 2. Pelvic radiograph after cesarean section showing diastasis of the pubic symphysis.

implications discussed (Fig. 3).

At the time of writing the patient was pregnant with her second child.

3. Discussion

Bladder exstrophy is a rare congenital malformation, occurring in 1 in 30,000 to 50,000 live births. Pseudoexstrophy is very rare, accounting for <10% of bladder exstrophy cases [4,6]. Bladder exstrophy is caused by the failure of mesoderm components to fuse in the midline during fetal life and is characterized by defects in the anterior wall of the lower abdomen, dysplasia, loss of bladder musculature, dilatation of the urethra due to dysplasia of the bladder neck, anterior deviation of the anus, downward deviation of the umbilicus, separation of the pubic symphysis, and loss of the anterior pelvic floor muscles. In addition to a split genitalia and short perineum, the vagina and uterus may also be associated with morphological abnormalities and pelvic organ prolapse due to the loss of the anterior pelvic floor muscles [7].

Pseudoexstrophy is a variant of bladder exstrophy and is characterized by normal external genitalia, no abnormalities of the urinary tract system, a low-set umbilicus, and asymptomatic dissection of the rectus abdominis muscle and pubic symphysis. Because there is little or no dysuria, it often remains undiagnosed in childhood and may be incidentally noted in adulthood, as in the present case [3].

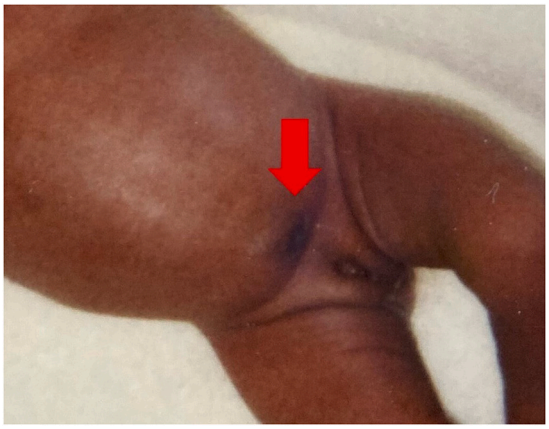

Fig. 3. Patient's abdomen after delivery.

A normal umbilicus is located in the center of the anterior abdominal wall at the fibrocartilage level between the third and fourth lumbar vertebrae [8], and a low umbilicus, as in the present case, may trigger the diagnosis of pseudoexstrophy.

Because pseudoexstrophy is very rare, there have been no comprehensive reports on the prognosis of pregnancy and delivery. It is believed that there are cases of pregnancy and delivery without the condition being diagnosed, as in this case.

In contrast, previous studies reported that bladder exstrophy results in a high-risk pregnancy and delivery for both the mother and child, with a high miscarriage rate, uterine prolapse after vaginal delivery, temporary dysuria, postpartum hemorrhage, and urinary tract infection [9,10]. Although vaginal delivery is possible in patients with bladder exstrophy, multidisciplinary management of both the mother and child is desirable because of the risk of injury to the mother during delivery and the associated risk to the child [11]. Multidisciplinary care is also desirable during cesarean delivery to minimize the occurrence of complications during laparotomy resulting from anatomical abnormalities and childhood plastic surgery [12].

Pseudoexstrophy may be undiagnosed even in adulthood, and the risks to pregnancy and delivery may be lower than in classical bladder exstrophy. However, because this variant of bladder exstrophy is heterogeneous and might be associated with both functional and morphological abnormalities, the risk of pelvic organ prolapse should be considered. With regard to pregnancy and delivery in patients with pseudoexstrophy, Cai et al. are the only authors, to the best of our knowledge, to report a case of repeated placental abruption due to bicornuate uterus caused by pseudoexstrophy [5]. These authors also described the risk of organ damage during laparotomy due to anatomical abnormalities and wound scar herniation after abdominal closure.

We cannot deny the possibility that in our case, the two miscarriages that occurred prior to this pregnancy were also influenced by uterine malformations, and we should have been fully aware of the risks of miscarriage and premature birth during and after laparotomy. Although cesarean section was appropriate in this case, pseudoexstrophy should have been diagnosed earlier, and the perinatal and perioperative risks should have been understood.

In conclusion, a low-set umbilicus should alert clinicians to bladder pseudoextrophy.

Contributors

Yusuke Taira participated in the conception of the case report and drafted the manuscript.

Nana Yara contributed to the review, obstetric management and editing of the manuscript.

Yoshino Kinjo contributed to the review, obstetric management and editing of the manuscript.

Tadatsugu Kinjo contributed to the review, obstetric management and editing of the manuscript.

Keiko Mekaru contributed to the review and editing of the manuscript.

Yoichi Aoki contributed to the review and editing of the manuscript.

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Patient consent

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Provenance and peer review

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Conflict of interest statement

The authors declare that they have no conflict of interest regarding the publication of this case report.

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