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Antenatal Ultrasound Profile of Bilateral Multicystic Renal Dysplasia: Report of Two Cases in Kati

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Abstract

Bilateral Multicystic Renal Dysplasia (MCRD) is a rare condition characterized by large cystic kidneys and completely remodeled, non-functional parenchyma. Ultrasound is the gold standard for prenatal diagnosis. The fetal prognosis is poor. Prenatal management relies primarily on ultrasound monitoring of pregnancy progression, the pathology, and the amount of amniotic fluid. We report two cases observed at the Clinique Médicale Amitié in Kati to study the contribution of ultrasound in the management of this condition.

Keywords

Bilateral Renal Dysplasia, Prenatal Diagnosis, Ultrasound, Poor Prognosis

1. Introduction

Multi-Cystic Renal Dysplasia (MCRD) is a usually unilateral renal developmental anomaly characterized by a large cystic kidney and completely remodeled, non-functional parenchyma. It represents the most frequent clinical manifestation of the congenital anomalies of the kidneys and urinary tract known as Congenital Abnormalities of Kidney and Urinary Tract (CAKUT) [1] [2]. It is a rare condition, with an incidence that varies considerably between 1 in 500 and 1 in 6700 births [1] [3]. Obstetric ultrasound is the gold standard for prenatal diagnosis, which will necessitate the search for other associated malformations. Prenatal management relies primarily on ultrasound monitoring of pregnancy progression, the pathology, and the amount of amniotic fluid [4]. The objective was to describe the ultra-

sound features and to identify associated anomalies.

Case 1: Ms. O. D., 30 years old, gravida 3, parity 1, miscarriage 1 (third pregnancy, primiparous with one live infant and a history of miscarriage). She had not had any ultrasounds during this pregnancy. She was referred to us for a routine obstetric ultrasound on June 7, 2023, at the Amitié Medical Clinic. The examination performed by a radiologist revealed a viable, singleton intrauterine pregnancy of 36 weeks' gestation. This ultrasound examination showed large, hyperechoic, poorly differentiated kidneys with millimeter-sized, multi-locular cysts (Figure 1(a)). An occipital defect causing a meningoencephalocele was also present (Figure 1(b)). We also observed anhydramnios. Based on the ultrasound findings, outpatient monitoring was initiated. One week later, Ms. O. D. returned with a lack of perception of fetal movements. A second ultrasound performed that day confirmed a missed miscarriage. Labor was induced after obtaining informed consent from the parents.

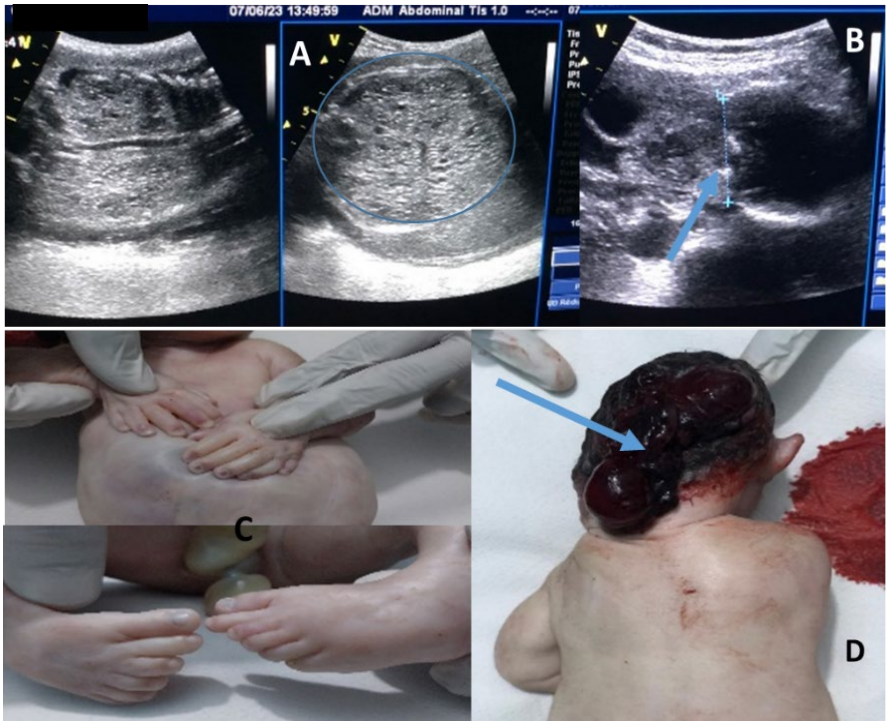

Figure 1. Obstetric ultrasound and photo of the newborn showing: (A) DRMK (circle); (B) a defect, the meningoencephalocele (arrow); (C) supernumerary fingers and toes; (D) an occipital mass corresponding to the meningoencephalocele (arrow).

After 8 hours, she delivered a stillborn female infant weighing 2950 grams and measuring 52 cm, with a large abdomen corresponding to enlarged kidneys due to dysplasia, and an occipital mass consistent with a meningoencephalocele. Well-developed supernumerary fingers and toes were also noted (Figure 1(c) and Figure 1(d)), which had not been visualized on ultrasound. The postpartum period was straightforward.

Case 2: Ms. M. K., 40 years old, gravida 7, parity 5, 1 miscarriage (seventh grav-

ida, fifth parity with 6 live births, including one twin birth and a previous miscarriage). She had only one ultrasound scan in the first trimester at 12 weeks of gestation. She was referred to us on July 21, 2024, for a third-trimester obstetric ultrasound. The examination performed by a radiologist revealed a non-viable, singleton intrauterine pregnancy without overlapping of the skull bones, estimated at 38 weeks of gestation. Large, hyperechoic, poorly differentiated kidneys with millimeter-sized, multi-locular cysts were noted (**Figure 2(a)**). These were associated with generalized skin detachment and anhydramnios. Based on these ultrasound findings, we concluded that she had bilateral renal dysplasia complicated by edema and intrauterine fetal death. A meningoencephalocele was also observed (**Figure 2(b)**). Labor was induced after the couple agreed. She gave birth to a fresh stillborn female, weighing 3500 grams and measuring 53 cm, confirming the same observations made on ultrasound (**Figure 2(c)**). The postpartum period was uneventful.

Iconography: these images illustrate our observations.

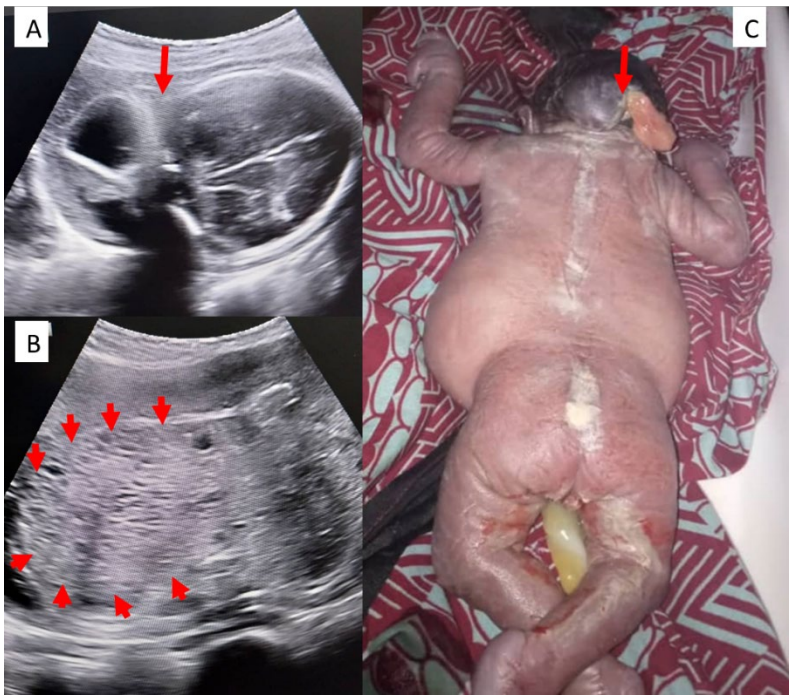

Figure 2. Antenatal ultrasound and posterior view photo of the newborn showing: (A) DRMK (arrowheads); (B) meningoencephalocele (arrow); (C) occipital mass corresponding to the meningoencephalocele.

2. Discussion

2.1. Epidemiology

Renal dysplasia is a rare entity. Its incidence is 2.6 cases per year [4]. For others, it is 1/4300 births [2]. We observed 2 cases in 7 years out of more than 10,000 obstetric ultrasounds performed during the 2nd and 3rd trimesters. It is generally unilateral, affecting the left side in 75% of cases [4]. Bilateral forms are rarer, rep-

resenting 1/4 of cases, and are usually fatal [5]. However, in the study by Hekmat Chaara *et al.* [4], it was bilateral in 61% of cases, which corroborates our case. Renal dysplasia most often affects boys in 60% of cases, with a sex ratio of 1.48 [6], according to most authors [4] [6]. Hekmat Chaara *et al.* [4] reported a male predominance of 71% and a sex ratio of 2.6/1. Our cases were female, as in the case of Faye Dieme M.E [1]. This could be explained by the small number of cases in our series.

2.2. Diagnosis

It is made by ultrasound in almost all cases (94%) prenatally [4] [7] during the morphological examination between 20 and 22 weeks of gestation. The sensitivity of ultrasound varies between 80 and 100% with a false-positive rate of approximately 2% [8]. Classically, ultrasound reveals anechoic intrarenal cysts, often large, of unequal size, varying in number, with a chaotic distribution within the kidney, and non-communicating. The residual renal parenchyma is echogenic, completely remodeled and fibrous, thinned, and sometimes unidentifiable, which explains the non-functional nature of the affected kidney. Color Doppler ultrasound shows the absence of parenchymal vascularization. The discovery of Renal Cell Carcinoma (RCC) should prompt a careful search for an associated anomaly. Some studies have reported extrarenal malformations associated with multicystic renal dysplasia such as esophageal or duodenal atresia, meningocele or heart disease [4]. In our setting, prenatal diagnosis was delayed, occurring between 36 and 38 weeks of gestation, which limited the search for certain associated malformations. However, we did observe meningoencephalocele in both cases, which is consistent with the literature. This diagnostic delay was due to the fact that the patients were seen late, and a lack of resources was the primary reason. We also noted well-developed supernumerary fingers and toes in one case after birth, which had not been seen prenatally. However, if second-trimester ultrasounds had been performed, all these malformations could have been visualized, hence the importance of morphological ultrasound. Chaara *et al.* [4] noted brain malformations in 17% of cases, followed by cardiac involvement (11%) and facial and neck malformations (also 11%). The amount of amniotic fluid should also be assessed [4]. Oligohydramnios is common in bilateral forms. In our case, anhydramnios was noted in both cases, which may indicate bilateral renal insufficiency, worsening the prognosis. This condition differs from polycystic kidney disease in several important ways. First, it is generally unilateral—affecting only one kidney in 95% of cases [9]. The cysts are non-communicating, and the affected kidney does not function at all, unlike polycystic kidney disease where the kidney may be functional, but this renal function gradually declines [10].

Monitoring: Once the diagnosis is made, prenatal ultrasound monitoring is necessary to:

—Assess the progression of polycystic kidney disease, which may be stable, regress, or progress during fetal life;

—Evaluate the function and morphology of the contralateral kidney, if it is unilateral, to establish a prognosis;

—Monitor for the appearance of new renal or extra-renal abnormalities;

—Quantify amniotic fluid;

—Although the monitoring schedule is not well codified in the literature. In the series by Hekmat Chaara *et al.* [4], monitoring was based on repeated obstetric ultrasounds performed monthly to bimonthly to look for signs of worsening renal function. According to studies, most DRMKs tend to involute and decrease in size prenatally, either completely or partially [11]. In our case, the prenatal diagnosis was late, and this monitoring was punctuated by a fetal death in utero at one week.

2.3. Prognosis

The literature reports a poor prognosis for the bilateral form, describing it as lethal [1] [4], which is consistent with our observation. Hekmat Chaara *et al.* [4] reported two cases of bilateral DRMK that survived postpartum in a series of 18 cases.

3. Conclusion

Multicystic renal dysplasia, although rare, is the most common congenital kidney disorder. Diagnosis is made by ultrasound. It is frequently associated with other malformations. The fetal prognosis is poor in bilateral cases.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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