A case report of congenital labial fusion

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Objective

To describe a rare severe case of congenital labial fusion.

Methods

Case report.

Results

We describe a rare case of labial fusion diagnosed in 20th week of pregnancy. A 32 year-old healthy Gravida 2 was referred to our department for her routine 20-22nd week scan. The ultrasonographic examination showed single viable pregnancy with appropriate biometric assessment. Detailed anatomical scan revealed presence of thin-wall cystic anechogenic structure related to external genital with no other pathological findings. Amniocentesis confirmed normal female karyotype. We excluded infectious origin and congenital adrenal hyperplasia. There was no history of teratogens exposure. Later in 23rd week of pregnancy ascites was detected and the patient decided to terminate pregnancy. On autopsy, the fetus (440g/24cm) was diagnosed with ascites and severe deformation of vulva with elongated fused labia and dilated urethra. Internal genital structures were normal.

Conclusion

Labial adhesion or labial fusion are common gynecological disorders in the pediatric population defined as complete or partial adherence of labia minora or majora. The age at which these disorders are commonly seen ranges from 13-23 months with an incidence of 1, 8%. This usually benign disorder is noticed due to frequent urinary tract infections. The congenital forms are associated with exposure to drugs such as chlorpyrifos or danazol or are associated with adrenal steroid 21-hydroxylase deficiency. It has also been described as part of Fraser syndrome. The most common complications are urinary tract infections, bladder distention and hydronephrosis due to obstruction. Treatment depends on the severity of labia adhesions; in mild form topical estrogen therapy is usually sufficient but fibrous adhesions may need adhesiolysis or genitoplasty. In the world literature we found only one case of severe congenital labial fusion associated with ascites. It was a case with poor perinatal outcome at 28th week of pregnancy and the presence of ascites was explained by flow of urine through vagina, uterus and tubes into the abdominal cavity, which was also confirmed biochemically.