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# Congenital vulvar teratoma: a case report

I Nyoman Hariyasa Sanjaya[1*], Ryan Saktika Mulyana[1], Evert Solomon Pangkahila[1], Hartanto[2]


## ABSTRACT

**Introduction:** Teratoma is an embryonal neoplasm consisted of 3 germinal layers. Teratoma usually arises along the line of embryonic cleft and sinus closure formed by the fusion of skin during the embryonic development. In this case report, we present a case of fetal Teratoma on the vulval region, a very rare variant of the disease.

**Case:** A 28-year-old woman came for a routine check-up for her first pregnancy at her 26th weeks of gestational age. On ultrasound examination, a mass was found around the fetal vulva. There was no mass or malformations on other body parts. Placenta and the amount of amniotic fluid were normal. History of congenital anomaly in the family was denied. The patient then gave birth to a female baby at 38 weeks of gestational age without any complication. The baby was healthy, 2650 gram in weight, 50 cm in length, with good APGAR Score (8-10). On the baby left labia, there was a mass measured 5 cm x 4 cm x 3 cm, with slight discoloration. We recommend an early tumor excision to prevent unnecessary complication such as malignant transformation. Although at the time of writing, the patient still waiting for the schedule to remove the tumor due to a non-medical factor.

**Conclusion:** Congenital Teratoma in the vulvar region was a very rare event. First-line therapy is early tumor excision and usually carries an excellent prognosis.


**Keywords:** Congenital Teratoma, vulva, case report, Bali

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[1]Fetomaternal Division, Department of Obstetrics and Gynecology, Faculty of Medicine, Universitas Udayana/Sanglah General Hospital, Bali, Indonesia
[2]Obstetrics and Gynecology Department, Faculty of Medicine, Universitas Udayana/Sanglah General Hospital, Bali, Indonesia

*Corresponding to:
I Nyoman Hariyasa Sanjaya;
Fetomaternal Division, Departement of Obstetrics and Gynecology, Faculty of Medicine, Universitas Udayana/Sanglah General Hospital, Bali, Indonesia;
hariyasasanjaya@gmail.com

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## INTRODUCTION

Teratoma was an embryonal neoplasm consisted of three germinal layers. Teratoma often formed along the embryonic cleft and sinus formed by the skin fusion during the embryonic phase.[1] Teratoma on vulva and vagina was a very rare case. If we did not count the incidence on the testis, then teratoma incidence on girl was 75%, and approximately 80% of them are benign. Teratoma that occurs on an early age was usually extragonadal. In contrast, Teratoma that occurs on older children was usually in the gonad.[2] Immature Teratoma was one of the ovary germ cell malignancy that happens most often in which 20-30% was benign. The immaturity of Teratoma was defined as a combination of the mature and immature component in which this component usually did not have a structured growth pattern.[3]

Teratoma can be classified using Gonzalez-Crussi classification system. According to this system, Teratoma can be classified into 0 or mature (benign), 1 or immature (probable benign), 2 or immature (probable malignant) and 3 or teratosarcoma. Stage 0 was a neoplasm that was consisted of mature tissue, while stage 3 which the neoplasm contains a lot of immature tissue.[4] Teratoma that was malignant can include embryonal carcinoma, *yolk sac* tumor, germinoma, dysgerminoma, or seminoma.[5]

The Teratoma on neonates was most often located on the sacrococcygeal and presacral region (47%), reproductive organ/the gonads (36%), mediastinum (6%), dan the rest was spread at the mesenterium, retroperitoneum, pericardium, spinal canal, cervical, nasopharynx, orbital and neurocranium (11%).[2] On rare cases, Teratoma on the liver, stomach, adrenal and umbilical cord was also reported. Nevertheless, the case report about Teratoma in the vulva region was very rare. To our knowledge, there were only a few other case reports that discuss Teratoma on the vulva. This report discusses a case about congenital Teratoma that was located on the vulva.

## CASE

A 28-year-old woman came for a routine check-up for her first pregnancy at her 26th weeks of gestational age. On ultrasound examination, a mass was found around the fetal vulva, as shown in Figure 1. There was no mass or malformations on other body parts. Placenta and the amount of

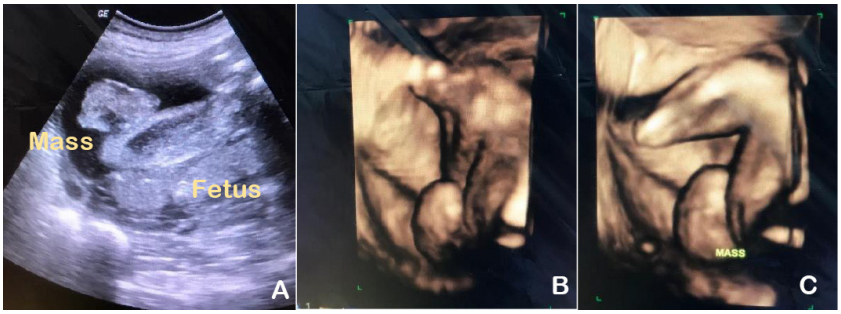


**Figure 1.** Intrauterine fetal ultrasound examinations. (A) The two-dimensional ultrasound examinations that showed a mass in the fetal vulva. (B) and (C) were images taken from four-dimensional USG examinations around the fetal vulva.

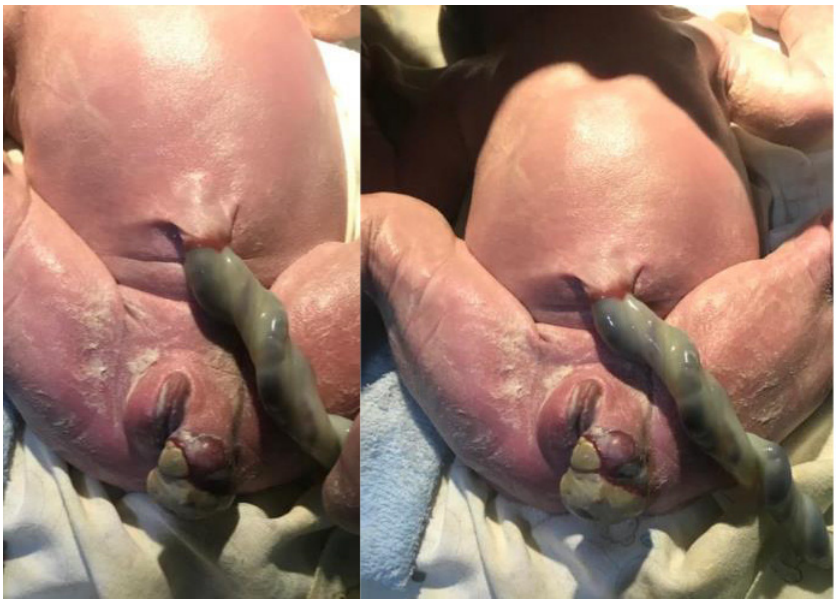

**Figure 2.** The photograph of the vulvar mass was taken just after the baby was born.

amniotic fluid were normal. History of congenital anomaly in the family was denied.

The patient then gave birth to a female baby at 38 weeks of gestational age without any complication. The baby was healthy, 2650 gram in weight, 50 cm in length, with good APGAR Score (8-10). On the baby left labia, there was a mass measured 5 cm x 4 cm x 3 cm, with slight discoloration. The patient was not yet scheduled for tumor excision due to a non-medical factor.

## DISCUSSION

Teratoma was initially defined by Willis at 1953 as a tumor consisted of exotic tissue of the body parts where the tumor grows.[6] The word Teratoma originated from the Greek, which is *téras oma* that translate into the monster tumor. Teratoma was usually congenital and benign in which the components came from three germs layer, endoderm, ectoderm, and mesoderm. Teratoma was most often located in the sacrococcygeal region with an incidence rate of 1 every 5.000 – 40.000 live births. Teratoma often diagnosed with prenatal USG or postnatal physical examinations.[5,7] Almost half of the Teratoma in neonates located on the sacrococcygeal and presacral region (47%) followed by reproductive organ/the gonads (36%), mediastinum (6%), and the rest was spread in various part of the body (11%).[2] Teratoma on the vulvar region just like, in this case, was an infrequent event.

Even though the exact cause of Teratoma was still unclear, there were few theories about the pathogenesis of Teratoma. One of them was the germ cell theory. Primitive germ cells have the ability to differentiate to form various tissue that we could found in Teratoma. Migration pattern of those cells can explain its location on the gonads, midline and paramedian.[8] While Osuoji hypothesized that ectopic epiblast cell that came from the vulva could grow into a teratoma. This was supported from his case report that a defect on the thorax and rectovaginal fistule can also accompany a vulva teratoma.[3] Other theories hypothesized the possibility of germ cell migration from the primitive yolk sac, remnants of primitive streak and Henson's Node. All these cells were the progenitor of mesenchymal cells that were very potent, and those cells can grow to be a teratoma.[5]

Before conduct a treatment on Teratoma, some suggested that additional examinations should be done, such as CT-scan, MRI or a more detailed USG. The reason was to detect other mass or another congenital anomaly of the fetus. On MRI examinations with T1-imaging, there would be a high-intensity image due to the fat layer in Teratoma. Differentiating mature and immature Teratoma was challenging since there were no specific criteria. Immature Teratoma was usually larger in size, cystic characteristics with a solid component, and often secrete AFP (a-Fetoprotein). In mature Teratoma, a solid component that was called the Rokitansky protuberance could be seen.[9] CT-scan had an excellent sensitivity to detect mature Teratoma in which the lesion contains both cystic and solid components. Even though CT-Scan was more sensitive, it posed a radiation-related risk.

Management of vulva teratoma was excision of the mass and staging of neoplasm malignancy. However, when would be the best time for teratoma excision on neonate patient? Cakmak et al. on his case report did the excision of vulva teratoma on

the fourth day after delivery while Osuoji et al. did the excision after 6-month of age. However, it was still recommended that the excision of Teratoma was done as soon as possible to prevent malignant transformation, and also the patient must be followed up regularly to assess the possibility of recurrence. Teratoma malignant transformation rate will increase after two months. Thus it was recommended to do excision before.[5] In this case, the primary treatment modality is tumor excision to prevent tumor rupture and malignant transformation. However, due to non-medical factor patient still wait for the operation schedule, and the patient was closely observed to assess tumor progressivity.

The prognosis of Teratoma depends on its size and location. Tumor recurrence was also a factor that affects the prognosis on neonates. A research was done by Wang et al., in 2017, the survival rate on neonates with recurrent Teratoma was 66.7% compared with those that did not have a recurrence (94.4%).[10] Frequently, another congenital anomaly could accompany a teratoma. Those anomalies include defects in the gastrointestinal system (imperforate anus), cardiovascular (left heart hypoplastic syndrome), and neurologic (absence of corpus callosum and arachnoid cyst).[6] In this case, the baby was born with an isolated vulvar teratoma.

In this case, it was recommended to have multidiscipline modality therapy with Pediatric surgery and the pediatric department to determine further treatment of vulvar Teratoma. Evaluation of other congenital anomaly was also recommended, as mentioned in a case report by Osuoji et al., that Teratoma was associated with VACTERL syndrome.[3] One of the treatment modality that can be considered was delivery by caesarean section and followed by intrapartum resection ex utero in which fetus still have support from the placenta to increase fetal survivability.[11] In this case, the baby was born through normal vaginal birth after consideration that the location of Teratoma would not disturb the delivery process.

## CONCLUSION

Congenital Teratoma on the vulvar region was a very rare event. Diagnostic modality other than USG can be utilized to find other masses or accompanying congenital anomalies. First-line therapy was tumor excision as soon as possible to prevent unnecessary complications. The prognosis of congenital Teratoma on the fetus usually excellent, but thorough examinations must be conducted to differentiate the benign and malignant variant.

## CONSENT

Patients have agreed and given their consent to be reported for academic purpose.

## CONFLICT OF INTEREST

All authors declare that there is no conflict of interest.

## AUTHOR CONTRIBUTION

All authors have contributed equally during all phase of the study, including, conducting the study, drafting and revising the manuscript, giving final approval and have agreeing to be accountable.

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