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Asymmetric twinning: A preterm baby with sacrococcygeal teratoma and parasitic twin born to a 16-year-old primigravida

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Abstract:

A 16-year-old primigravida, at 33 weeks and 5 days age of gestation came in due to preterm labor. Sonographic examination revealed an incidental finding of a mass attached to the sacrococcygeal area. The mass has a cystic and solid component diagnosed as sacrococcygeal teratoma. Attached to the mass were two lower extremity structures identified as femurs with feet and was considered as an underdeveloped parasitic twin. A classical cesarean section was performed because of the advanced preterm labor, and a live female infant weighing 2500 g was delivered. The parasitic lower limbs, however, inadvertently detached during delivery. There was a high index of suspicion because of a larger fundic height of 37 cm compared to the age of gestation and the difficult palpation on Leopold's maneuver. Accuracy of ultrasound findings helped the obstetricians to a timely and prepared for delivery.

Keywords:

Parasitic twin, pediatric tumor, sacrococcygeal teratoma

Introduction

Conjoined twins are rare and it has an estimated frequency of 1 in 50,000–100,000 live births. Parasitic or heteropagus twinning is an extremely rare condition, with an estimated incidence of <0.1 in 100,000 births.[1] Asymmetric and parasitic conjoined twins are rarer anomalies of monochorionic monoamniotic twins, consisting of an incomplete twin (parasite) attached to the fully developed body of the co-twin (autosite).[2] In the Philippines, there is only one published journal about parasitic twinning to date.

Sacrococcygeal teratoma is the most common tumor diagnosed in neonates and approximately 1 in 35,000 live births. There is a striking female preponderance, with a female-to-male ratio of approximately 4:1.[3] There are four published journals related to sacrococcygeal teratoma in the Philippines, which are discussed below.

At present, there is no published case of sacrococcygeal teratoma and parasitic twin similar to this case in the Philippines. Internationally, there are two published journals that both dealt with a diagnostic dilemma of whether it was solely a teratoma or a teratoma with a parasitic twin and the similar issue was encountered in this case.

Case Report

This is a case of a 16-year-old primigravida at 33 weeks and 5 days age of gestation,

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who came to the emergency room due to preterm labor. She had five prenatal checkups at Bahay Toro Health Center. She has unremarkable past medical history, personal history, menstrual history, and sexual history. There was no family history of congenital anomalies, and she had taken no medications during her pregnancy, except for prenatal vitamins.

Physical examination showed a fundic height of 37 cm incompatible to age of gestation, with nondistinct fetal back and fetal small parts upon palpation on Leopold's maneuver. Internal examination revealed 2 cm dilated, effaced, cephalic, intact bag of waters at station-2. Assessment on admission was an intrauterine pregnancy at 33 weeks and 5 days gestation by last menstrual period, cephalic in preterm labor with a consideration of a fetal congenital anomaly.

Baseline fetal heart rate on cardiotocogram was noted at 140–145 bpm, with minimal variability, no accelerations, and no decelerations with strong uterine contractions every 5–6 min; hence, tocolytics were given. However, labor progressed to 4 cm dilated, effaced, cephalic, intact bag of waters at station-2 after 12 h.

Sonographic examination was done and revealed a live, singleton pregnancy, in cephalic presentation with an estimated fetal weight of 1976 g. The fetus had good cardiac activity with noted polyhydramnios and amniotic fluid index of 29.38 cm. There was incidental finding of a mass with cystic and solid components arising at the sacrococcygeal area measuring 15.19 cm × 8.96 cm × 12.32 cm (volume 877.96 ml) that could be a sacrococcygeal teratoma [Figure 1a]. Seen attached to the mass were two structures identified as femurs compatible with 24 weeks and 1 day measurement with feet and tiny toes and was considered as an underdeveloped parasitic twin [Figure 1b]. With the aid of ultrasound, an assessment was made, Sonographic assessment was a fetus measuring 33 weeks by biometry with fetal congenital anomaly, to consider sacrococcygeal teratoma versus parasitic twin. A plan to terminate the pregnancy was made because of the advanced preterm labor.

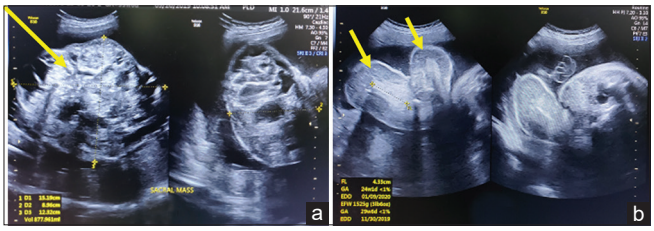

Figure 1: (a) This is a pelvic scan showing a sacral mass with cystic and solid component measuring 15.19 cm × 8.96 cm × 12.32 cm (volume 877.96 ml), suggestive of sacrococcygeal teratoma. (b) Seen attached to the mass are two structures identified as two lower extremities with identified femur compatible with 24 weeks and 1 day, nonidentified tibia-fibula, and feet with tiny toes, considered as an underdeveloped parasitic twin

Classical cesarean section was performed which provided large incision in the uterus to avoid rupture and hemorrhage of the mass at the time of delivery. However, difficulty was encountered during delivery of the large sacrococcygeal mass and identified limb-like structure attached to it. The limb-like structure that was attached to the sacrococcygeal mass inadvertently detached from the sacrococcygeal mass which seems to be connected by a soft tissue pedicle [Figure 2]. Minimal bleeding was noted on the site of detachment which was controlled immediately with compression. A live female infant weighing 2500 g, 44 cm long was delivered with an Apgar score of 7.9 and a Ballard's score of 33 weeks. There was a 17 cm × 10 cm × 9 cm mass connected to the sacrococcygeal area with overlying skin and noted with mixed cystic-solid consistency [Figure 3a]. The placenta was singleton weighing 350 g with cord length of 40 cm. Placenta was sent for histopathology. The placental histopathological result revealed a mature, singleton placenta, with noted infarction and intervillous hemorrhages.

The baby was preterm, pink, with noted subcostal retractions and tight air entry upon auscultation. She has normal heart rate with regular rhythm. The umbilical cord was noted with two arteries and one vein, and genitalia was grossly female. There was a 17 cm × 10 cm × 9 cm sacrococcygeal mass with overlying skin with mixed cystic-solid consistency with noted 2 cm × 2 cm depression with soft tissue-like pedicle [Figure 3b]. The detached limb-like structure measuring 8 cm × 6 cm is seemingly connected posteriorly to the sacrococcygeal mass depression by a soft tissue-like pedicle with noted penile-like structure [Figure 2]. The baby has patent anus and with grossly normal extremities. Pediatric assessment was a preterm female with Apgar 7,9 and birthweight of 2500 grams that was appropriate for 33 weeks' gestation with a sacrococcygeal teratoma with parasitic twin.

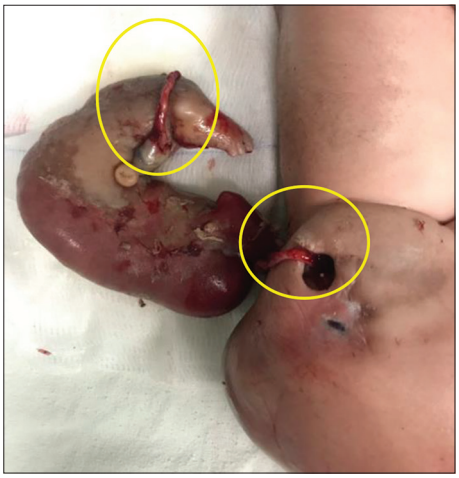

Figure 2: Detached limb-like structure measuring 8cm x 6cm connected to the sacrococcygeal mass by a soft tissue-like pedicle

The babygram revealed a large soft tissue mass density at the projection of the sacrococcygeal region, compatible with teratoma. The detached extracorporeal soft tissue mass was placed on the right side of the baby for documentation and was noted with apparent pelvic and lower and extremity osseous structures [Figure 4]. Computed tomography was done and revealed a well-defined enhancing complex mass in the sacrococcygeal region extending toward the pelvis, exhibiting cystic attenuation, calcification, septations, and soft tissue components measuring about 13.6 cm × 8.4 cm × 12.8 cm, suggestive of teratoma. Hence, the patient was referred to surgery service for co-management. As for the limb-like structure, it was committed to be a parasitic lower limb by the pediatrics and surgery service.

The baby underwent excision of the sacrococcygeal teratoma at 19th day of life. The patient was managed by the pediatric surgery for the excision of the mass and the reconstructive and plastic surgery for the skin advancement flap. An elliptical incision was done to obtain a cross-section of a large area of tissue and to histologically see the transition from normal to abnormal skin and get the excess skin. The mass excised was measured 20 cm × 25 cm × 10 cm [Figure 5]. Three specimens were sent for histopathology. The first specimen was labeled "teratoma," consisting of an irregular, multinodular, tan brown to tan gray, doughy to soft tissue measuring 15.0 cm × 13.0 cm × 6.5 cm and weighing 750 g [Figure 6a] which revealed an immature teratoma, Grade 3 with negative tumor involvement. Other microscopic findings included pancreatic tissue, mature and immature cartilage, osteogenic foci with calcifications, transitional epithelium, uveal tissue, and skin adnexae [Figure 6]. The second specimen labeled as "coccyx" was positive for tumor involvement and signed out as immature cartilaginous tissue. The specimen labeled "parasitic twin's bowels" revealed vascular congestion and edema and negative for tumor involvement. The baby was weighed after the surgery and noted to be 1500 g. Assessment was sacrococcygeal immature teratoma, grade 3.

Pediatric oncology service did serial monitoring of serum alpha-fetoprotein (AFP) levels every 3 months for the sacrococcygeal immature teratoma, grade 3. Initially, the level of AFP is >20,000 ng/ml taken on her 5th day of life. On her 44th day of life (postoperative day 25), it was noted at >2000.00 ng/ml, and on her 52nd day of life (postoperative day 33), it was elevated to 6931.04 ng/ml. Few days before discharge, on her 91st day of life (postoperative day 77), it was noted with significant decline of serum AFP >400 ng/ml. The baby was discharged on her 100th day of life, and the baby was lost to follow-up with the pediatric oncology.

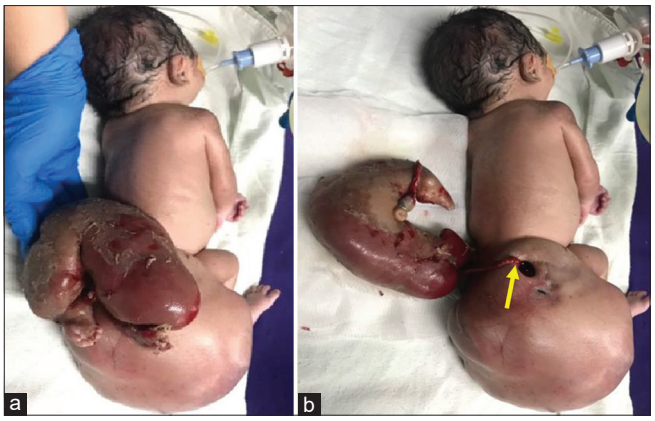

Figure 3: (a) There is a 17 cm × 10 cm × 9 cm mass connected to the sacrococcygeal area with overlying skin and noted with mixed cystic-solid consistency. (b) There is a 2 cm × 2 cm depression on the sacrococcygeal mass with noted soft tissue-like pedicle

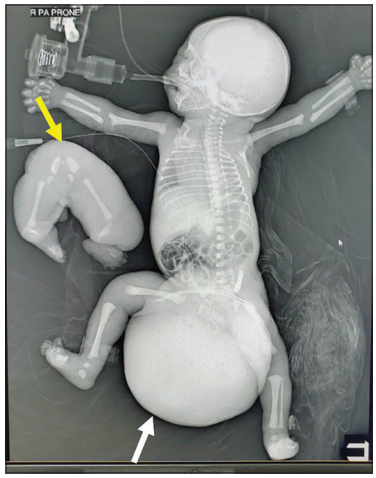

Figure 4: Babygram showing a large soft tissue mass density at the projection of the sacrococcygeal region (white arrow). The extracorporeal soft tissue mass at the right side of the baby with apparent pelvic and lower extremity osseous structures (yellow arrow)

Figure 5: Postoperative picture of the patient with the excised mass measuring 20 cm × 25 cm × 10 cm weighing 750 g

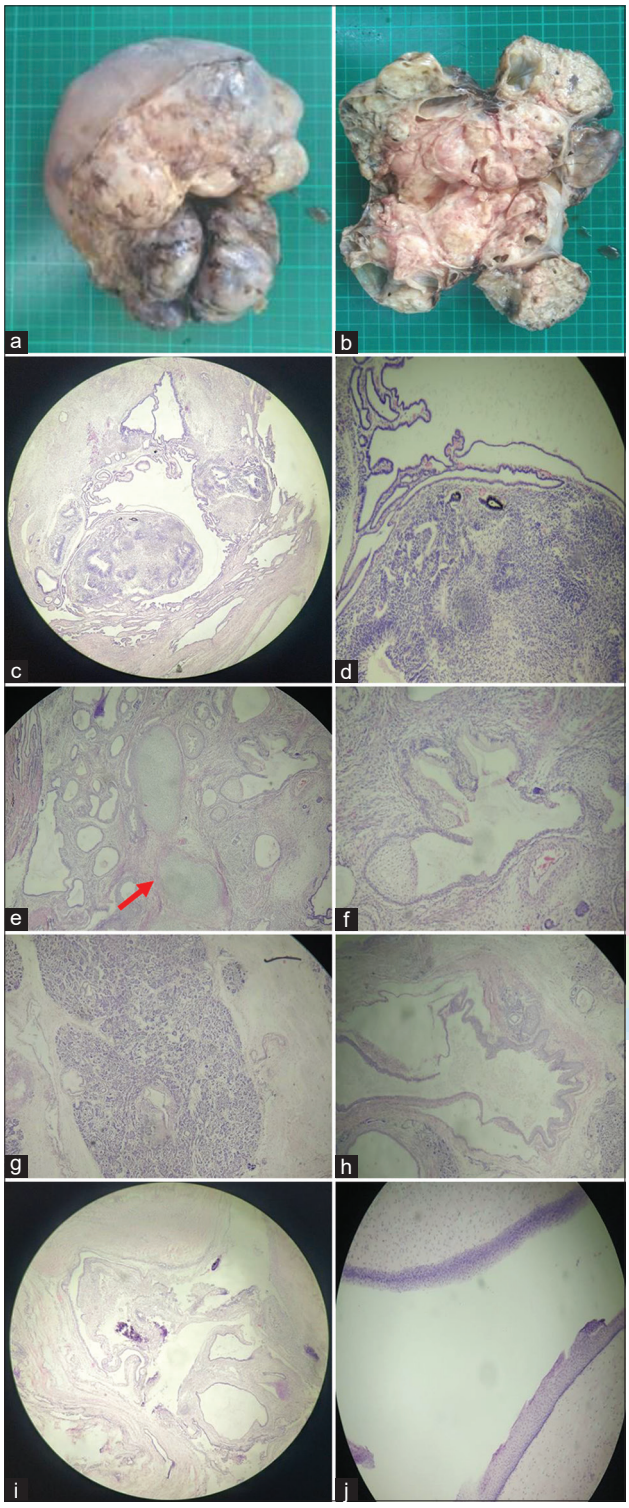

Figure 6: Histopathology pictures. (a) Specimen was labeled as teratoma. It was irregular, multinodular, tan brown to tan gray, doughy to soft tissue measuring 15.0 cm × 13.0 cm × 6.5 cm and weighing 750 g. (b) On cut section, it showed a highly multiloculated interior, some forming spongy surfaces, with various sized, thin-walled cysts containing serous fluid. Few necrotic and hemorrhagic foci were observed. Microscopically, it showed the presence of (LPO view) immature neuroepithelial elements merging with cellular stroma (c); (HPO view) primitive retinal tissues (d); (LPO view) immature cartilage (e); (LPO view) intestinal type glands (f); (HPO view) primitive pancreatic tissue (g); (LPO view) transitional epithelium (h); (LPO view) calcification with osteogenic foci (i); and (LPO view) the presence of skin epithelium (j)

Case Discussion

Distinguishing between heteropagus parasitic twin and sacrococcygeal teratoma is important because of the malignant potential of teratoma. However, to encounter both cases at the same time in one patient is extremely rare.

There are two major theories of the embryologic origin of conjoined twins, and it was labeled "fission" and "fusion." Proponents of the first suggest that incomplete fission of the blastocyst inner cell mass during the primitive streak stage, days 13–15 postfertilization, results in two centers of axial growth that retains a connection at some point, while "fusion," in contrast, refers to two originally distinct inner cell masses that coalesce secondarily at a later stage.[4] However, the most commonly accepted theory explaining the occurrence of conjoined twins is incomplete splitting of the embryonic axis.[5]

Parasitic twins occur when one embryo of a pair of monozygotic twins starts to develop, but the pair does not fully separate and one embryo's development prevails over the other. Rather than conjoined, it is considered parasitic because it is incompletely formed or wholly dependent on the body functions of the complete fetus.[1] A standardized terminology was postulated by Spencer using terms such as cephalopagus, ischiopagus, parapagus, craniopagus, rachipagus, and pygopagus. Parasitic twins result from the selective destruction of part of one embryo, the subsequent location indicating the original site of attachment.[6]

Based on the classification cited by Spencer, this is a case of pygopagus twin wherein the attachment is at the sacral region. The detached parasitic leg was noted with macerated areas and different skin color from the autosite, and this is supported by the study by Sharma *et al.*, which vascular compromise causes tissue of the parasitic twin to become dependent on collaterals derived from the autosite and selective ischemic atrophy of the deprived portion of the parasite's body follows.[4]

The diagnosis of parasitic twin is made when one or more of the following was fulfilled: (a) they should be enclosed within a distinct sac and be partially or completely covered by normal skin, (b) with grossly recognizable body parts; (c) it is attached to the host by only a few relatively large blood vessels, and (d) be located immediately adjacent to one of the sites of attachment of conjoined twins.[7]

In the Philippines, there is one published report about parasitic twinning entitled "Craniopagus parasiticus-Rare conjoined twinning: A case report" by

Brillantes. It was a case of 14-day-old twins who were attached to each other at the parietal region.

The main differential diagnosis of a pygopagus parasitic twin is a sacrococcygeal teratoma. Sacrococcygeal teratoma is the most common tumor diagnosed in neonates and approximately 1 in 35,000 live births. There is a striking female preponderance with a female-to-male ratio of approximately 4:1.[3]

Only four reports were published in the Philippines. One is entitled "Sacrococcygeal teratoma in Filipino children" by Menes r. Second is a case report by Bitera entitled "Dystocia and sacrococcygeal teratoma" in which they dwell in the problem it posed during delivery and surgical intervention that needed the attention of several specialties, including pediatric and orthopedic surgery. Third is a case report entitled "Sacrococcygeal teratoma including a case report of a well differentiated tumor in a 3 month old infant" by Buenviaje *et al*. At last, "Sacrococcygeal teratoma: Diagnosis and management" by Balbido *et al*. These cases only reported a diagnosis of sacrococcygeal teratoma but not parasitic twins. The presence of both the parasitic twin and teratoma made this case unique from the other cases that were published locally.

A primary complication associated with sacrococcygeal teratoma is dystocia during delivery caused by the sheer size of the tumor. The average size is 8 cm with a range of 1–30 cm. Large-sized sacrococcygeal teratomas can cause dystocia during delivery in approximately 6% to 13% of the time requiring urgent resection during cesarean delivery.[8] This supported the classical cesarean section done by the team which also provided adequate space for extraction of the baby with a large mass.

Before 2 months of age, most of these lesions are benign and surgical extirpation with coccygectomy can be accomplished with relatively low morbidity and mortality. Excision of the coccyx is mandatory in all cases. Failure to remove the coccyx results in 30%–40% recurrence rate, with noted higher probability of malignancy.[9] In our case, surgery was done on her 19th day of life. For the surgical approach, a chevron or inverted V incision was the standard approach; however, in our patient, an elliptical incision was made to obtain a cross-section of a large area of tissue, to histologically see the transition from normal to abnormal skin and get the excess skin. In this case, the coccyx was positive for tumor involvement and signed out as immature cartilaginous tissue.

Sacrococcygeal teratoma is graded histologically as follows: grade 0, tumor contains only mature tissue; grade 1, tumor contains rare foci of immature tissues; grade 2, tumor contains moderate quantities of immature tissues; and grade 3, tumor contains large quantities of immature tissue with or without malignant yolk sac elements.[8] In this case, it was grade 3 because of the noted microscopic findings of pancreatic tissue, mature and immature cartilage, osteogenic foci with calcifications, transitional epithelium, uveal tissue, and skin adnexae. Postoperatively, adjuvant chemotherapy is used in malignant cases in the form of combination of vincristine, actinomycin D, and cyclophosphamide, with or without Adriamycin.[9] Hence, in our case of immature teratoma, adjuvant chemotherapy has no benefit since early surgical intervention is associated with better prognosis. The plan of the pediatric oncology service was serial monitoring of serum AFP every 3 months until values normalize. A repeat serum AFP >400 ng/ml was done her 91st day of life (postoperative day 77) with noted significant decline before discharge.

The use of AFP as a tumor marker is well established and persistent elevated level may indicate a residual tumor, recurrence, or malignant degeneration.

It was fortunate to document these extremely rare conditions in one patient. The pediatric and surgery service committed that the limb-like structure is a parasitic twin, and the teratoma was supported with the histopathology results.

Summary

Distinguishing between heteropagus parasitic twin and sacrococcygeal teratoma is important because of the malignant potential of a teratoma. This is a case of a preterm baby girl with an externally attached parasitic twin and a sacrococcygeal teratoma. This was agreed upon by the pediatrics and surgery services, as evidenced by the findings on babygram of apparent pelvic and lower extremity osseous structures. The recognizable lower limb structure attached to the sacrococcygeal mass by a soft tissue-like pedicle that bled upon its detachment strengthened the diagnosis of a parasitic twin.

The sacrococcygeal teratoma was confirmed by the microscopic findings of pancreatic tissue, mature and immature cartilage, osteogenic foci with calcifications, transitional epithelium, uveal tissue, and skin adnexae. Hence, diagnosed as an immature teratoma, grade 3 was made.

Sacrococcygeal teratoma is a common neoplasm in neonates, found approximately one in 35,000 live births. However, heteropagus or parasitic twinning is an extremely rare condition, with an incidence of <0.1 in

100,000 births. Given that there were no reported cases published locally, being able to diagnose both cases in a patient was clinically remarkable.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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