Congenital Nasopharyngeal Teratoma (Epignathus)

NANTANA KAEWPILA, M.D.*, MANA ROCHANAWUTANON, M.D.**,
KEASORN TANGCHAROENPANICH, M.D.***, SOMBOON CHANSAKULPORN, M.D.****

Abstract

The case report of a congenital nasopharyngeal teratoma presented as an intra-amniotic sac mass was described. Histologically, the tumor was composed of highly differentiated tissue derived from three germ layers. Those tissues were partially developed as infant organs with a central core made up by bone. Haphazardly arrangement of such tissues with disorganized sketal axis helped in differentiating the mass from an asymmetric twins. The evidence of tumor attachment was seen at the retro-uvular region.

Key word : Congenital Teratoma, Nasopharyngeal Teratoma

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Teratoma, a congenital tumor arising from three germ layers, is frequently seen at the sacrococcygeal region of infants[1]. It uncommonly occurs in other sites of the body and is extremely rare in placenta and umbilical cord[2-4]. In this case, it exhibited as a lobulated mass mostly covered by skin, and had departed from its attachment site, to become a separated mass in the amniotic sac resembling an acardiac twin, clinically. Recognition of the tumor at delivery will lead to a serious attempt to find out the base of detachment in order to stop bleeding and then save the infant.

* Department of Pathology, Faculty of Medicine, Srinakharinwirot University, Bangkok 10110,
** Department of Pathology, Faculty of Medicine, Ramathibodi Hospital, Mahidol University, Bangkok 10400,
*** Department of Obstetrics and Gynecology, Bangkok Metropolitan Medical Collage and Vajira Hospital, Bangkok 10300,
**** Department of Pediatrics, Faculty of Medicine, Srinakharinwirot University, Bangkok 10110, Thailand.

CASE REPORT

A preterm, male infant was delivered by a 22-year-old woman, primigravida, who had had no antenatal care. The mother was admitted to the hospital due to labor pain and then had an immediate membrane rupture. The 1,500 g infant was delivered by forceps extraction due to fetal distress. The Apgar scores were 7 and 8 at the first and fifth minute respectively. The infant was estimated as 34 weeks gestational age, small for date and was found to have a high-arch palate with an inverted "V" shaped uvula. Active bleeding in the oral cavity and the characteristic of wide open mouth were noted. No further anomalies or skin lesion were found. A few minutes later, a lobulated fetus-like mass was spontaneously delivered then followed by a normal placenta and umbilical cord.

Gross morphological findings

The placenta with an attached umbilical cord weighed 570 g totally. The placental plate was 23x17x3 cm and revealed smooth, glistening fetal surface with normal vascular pattern. The maternal surface revealed complete cotyledon. No retroplacental hematoma was found. Serial cut sections of the placenta revealed no striking lesion. The attached umbilical cord was 27 cm long and 1.0 - 1.5 cm in diameter. It showed normal insertion 3 cm away from the placental rim. The cut sections revealed three normal vessels. The mass weighed 440 g and measured 12x11x6 cm. It showed a lobulated surface which was mostly covered by skin (Fig. 1). Xerographic study demonstrated a centrally disoriented core of bones (Fig. 2). The cut sections of the tumor revealed solid and cystic areas showing variation of tissue components in different areas. They included epidermis with underlying edematous soft tissue, brain tissue with partially well-formed sulci and gyri, fat, cartilage and bony parts. Only a small area of hemorrhagic raw surface, 1.0 cm in diameter, was noted on the mass. It contained cross cuts of blood vessels, suggestive of an attachment site of the mass to other structures.

Histologic findings

The placenta, umbilical cord and membrane revealed no remarkable change. The lobulated mass was composed of a wide variety of mature tissues, representing all three germ layer derivatives (Fig. 3, 4). Its surface was mostly covered by skin. The tissue components included epidermis with sebaceous glands and hair follicles, intestinal epithelium with incompletely-formed muscular layers, small ducts and acini, clusters of infant hepatocytes with evidence of hematopoiesis, adipose cells, cartilage and bone. Brain tissues, choroid plexuses and ependymal structures were hapha-

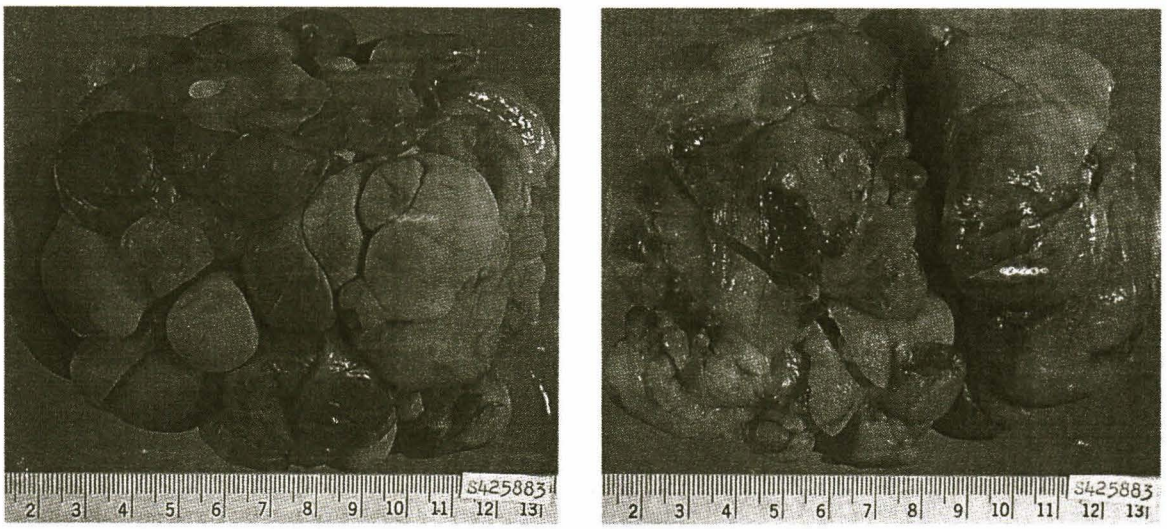
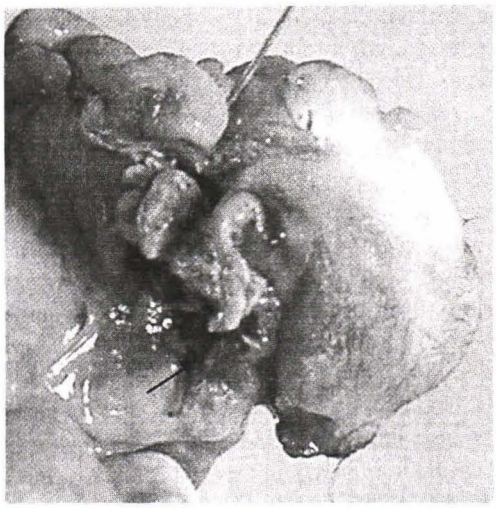


Fig. 1. A large multilobulated mass mostly covered by skin.



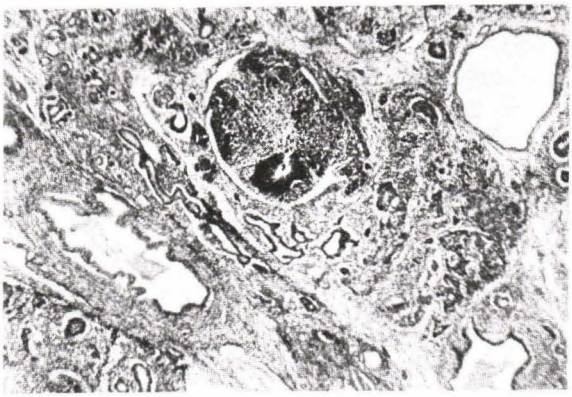


Fig. 3. Tissue components including ependymal structures with surrounding connective tissue and cavities lined by mucin-producing epithelium.

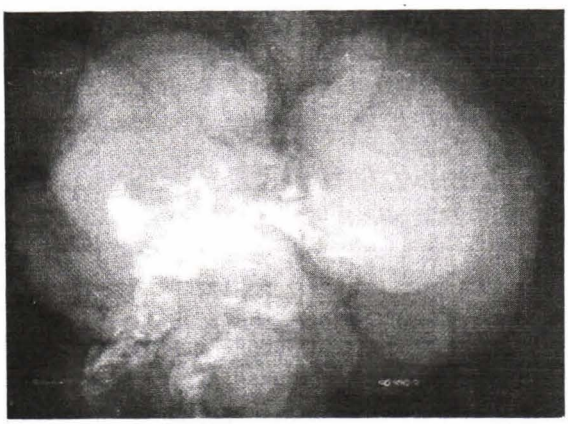


Fig. 2. Xerograph showing disoriented calcified structures.

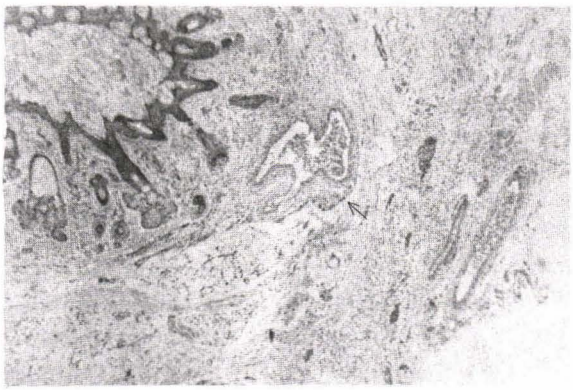


Fig. 4. Epidermis with sebaceous glands and developing hair follicles, mature fatty tissue and proposed feeding vessels (arrow).

zardly present among the previously mentioned tissues.

DISCUSSION

The tumor of this case presented as a separated mass in the amniotic sac with a preterm, male infant who had a high-arch palate and active bleeding per oral. According to the gross appearances, the tumor was misunderstood to be an acardiac twin at the first glance. The diagnosis of teratoma was based on the haphazardly arranged mature tissues derived from all three germ layers with lack of a skeletal axis.

According to Baldwin[5], asymmetric twins were separated as chorangiopagus parasiticus (acardius/acephalus), ectoparasite (heteroparasite), and endoparasite (fetus in fetu). Those were sometimes difficult to distinguish from fetal congenital teratomas or teratomas of placenta and umbilical cord. Only a few cases of placental[6] and umbilical[7,8] teratomas have been reported in the literature. Fox and Elston[2] suggested that an acardius had its own umbilical cord that attached to the placenta of the synchronous infant or to a separate placenta. The umbilical teratoma was recognized as a rudimentary or well-developed mass within the

umbilical cord, while the placental teratoma did not have an umbilical cord and received its arterial blood from a major fetal vessel on the placental surface. In addition, an acardius was usually described as an amorphous mass which was composed of disorganized fetal tissues. Many cases demonstrated cranial and caudal ends of the fetus with evidence of central skeletal bone easily recognizable as vertebral column, ribs, pelvis, and base of the skull. Some of these tumors developed in ovarian tissue and were referred to as a fetiform teratoma or homunculus[9]. Congenital teratomas arising at any part of the body have been discussed in similar directions[5].

For this case, the tumor appeared as a separate mass in the amniotic sac without a trace of its origin from the external parts of the infant, placenta or the umbilical cord. In addition to the histologic and xerographic evidences of teratoma, we saw large, thick and thin walled blood vessels with fresh hemorrhage of surrounding soft tissue around the raw surface of the mass. These were proposed to be feeding vessels from the host. The evidence of active bleeding from the uvular region with uvular defect suggested that the tumor arose from the anterior nasopharyngeal region. The open mouth gesture was the clue that the mass had been in the oral cavity for a long period of time and finally protruded out of the oral cavity, so detachment injury occurred during delivery. The presence of recent blood clot in the infant's mouth and the non-infarcted mass also supported that the detachment developed shortly before birth. The forceps application, without the prenatal recognition of the tumor in the head area, might also have played a role in the tumor detachment.

Congenital teratomas of the nasopharynx (also known as epignathus) have rarely been reported. Tharrington and Bossen[10] could cite only 27 cases from the literature, with addition of their own case. Most cases (81.5%) were detected at birth. Nine of those cases presented with a protruding mass from the oral cavity while the remainder presented with airway obstruction. Polyhydramnios occurred in 60 per cent of the recorded cases[10,11]. It was proposed to be secondary to interference of amniotic swallowing produced by the obstructing mass. As the tumor was composed of varied tissues, including bone and cartilage, the ultrasonographic, computed tomography or even usual radiographic study could yield a prenatal diagnosis of this tumor [12]. Histologically, most cases of congenital teratoma of the nasopharynx are benign tumors[10]. A case of a recurrent tumor with a malignant germ cell differentiation to be an endodermal sinus tumor has been reported[13] which was detected after 3 years of removal of the primary nasopharyngeal teratoma. This incidence suggests an extended follow-up of the patient after removal of the primary mass.

In summary, the congenital nasopharyngeal teratoma is rare, but, with awareness of this lesion, a prenatal diagnosis is possible by clinical investigations, resulting in a well-planned delivery and proper childcare at birth.

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เทอราโตมาของเนื้อเยื่อหลังโพรงจมูกตั้งแต่กำเนิด: รายงานผู้ป่วย 1 ราย

นันทนา แก้วพิลา, พ.บ., มานะ โรจนวุฒนนท์, พ.บ.**,*
*เกสร แต่งเจริญพาณิชย์, พ.บ.***, สมบูรณ์ จันทร์สกุลพร, พ.บ.*****

รายงานผู้ป่วย 1 รายซึ่งมีเนื้องอกเทอราโตมาที่เนื้อเยื่อหลังโพรงจมูกตั้งแต่กำเนิด จากการศึกษาทางด้านพยาธิวิทยาพบว่าเป็นเนื้องอกที่ประกอบด้วยเนื้อเยื่อที่มีพัฒนาการอย่างดีมาจากเนื้อเยื่อก่อกำเนิดทั้ง 3 ชนิด โดยประกอบกันคล้ายเป็นอวัยวะต่าง ๆ ของทารกและมีแนวของกระดูกเป็นแกนกลางของเนื้องอก หากแต่เนื้อเยื่อและกระดูกดังกล่าวกระจายกันอยู่อย่างไม่เป็นระเบียบจึงทำให้สามารถแยกออกจากครรภ์แฝดได้ ส่วนต้นกำเนิดของเนื้องอกรายนี้มีหลักฐานที่สนับสนุนว่าน่าจะมาจากเนื้อเยื่อหลังโพรงจมูกของทารกและหลุดออกมาอยู่ภายในถุงน้ำคร่ำระหว่างการคลอด

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นันทนา แก้วพิลา และคณะ
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* ภาควิชาพยาธิวิทยา, คณะแพทยศาสตร์ มหาวิทยาลัยศรีนครินทรวิโรฒ, กรุงเทพ ฯ 10110
** ภาควิชาพยาธิวิทยา, คณะแพทยศาสตร์ โรงพยาบาลรามาธิบดี, มหาวิทยาลัยมหิดล, กรุงเทพ ฯ 10400
*** ภาควิชาสูติศาสตร์-นรีเวชวิทยา, วิทยาลัยแพทยศาสตร์กรุงเทพมหานครและวชิรพยาบาล, กรุงเทพ ฯ 10300
**** ภาควิชากุมารเวชศาสตร์, คณะแพทยศาสตร์ มหาวิทยาลัยศรีนครินทรวิโรฒ, กรุงเทพ ฯ 10110