



**Society of Radiologists in Ultrasound
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Clinical history: 29 yo G1P0 with dichorionic diamniotic twin pregnancy referred at 25w5d for polyhydramnios and an enlarging left cheek mass in Twin A.

By report, a left cheek mass measuring 2.5 x 1.6 x 2.5 cm with a dominant feeding vessel was identified at 17 weeks, with normal intracranial anatomy. By 22 weeks, the mass had grown to 8.9 x 5.6 x 6.4 cm. The patient presented to our institution with symptoms of preterm labor at 26 weeks. At that time, the mass measured 19 x 12 x 12 cm with extension into the intracranial space. Secondary findings included severe polyhydramnios, enlarged structurally normal fetal heart (without hydrops) and maternal short cervix with funneling. The patient chose to have selective termination of twin A and amniotic fluid reduction in hopes of delaying preterm labor. However, one week after the procedure, the patient's labor progressed and she gave birth. The patient refused fetal autopsy. However, autopsy MR and core biopsy were allowed.

Figures:

FIGURE 1a: Coronal ultrasound image of the fetal calvarium and upper neck shows a large cystic and solid mass which extends from the left neck into the left cranial vault (yellow arrows denote the intracranial portion of the mass).

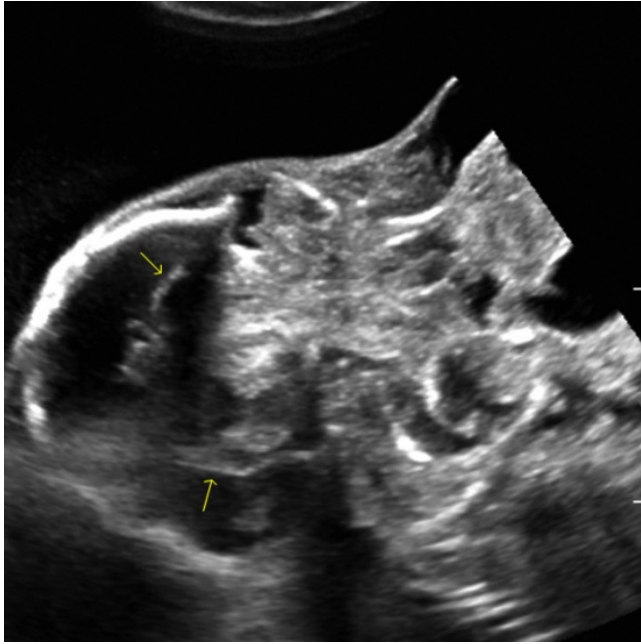


FIGURE 1b: Axial ultrasound image of the superior fetal head rightward shift of the falx (yellow) and compression of the fetal brain.

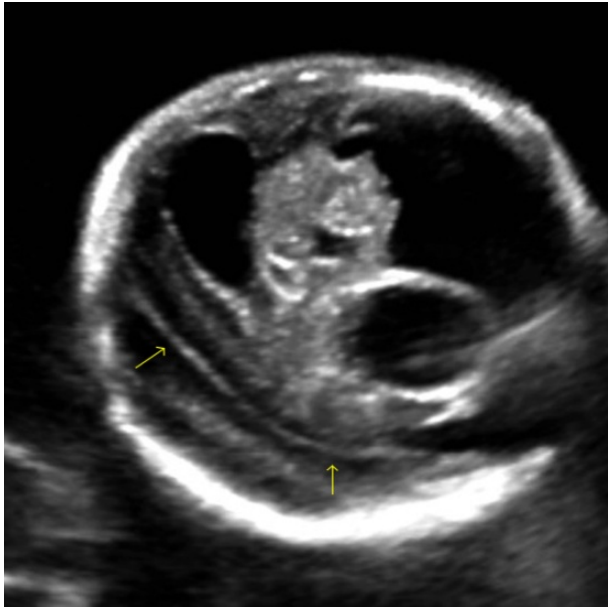


FIGURE 1c: Coronal ultrasound image of the fetal face shows the large cervical component of the mass. (Yellow arrows denote the eyes; pink arrows denote the nasal bones.)

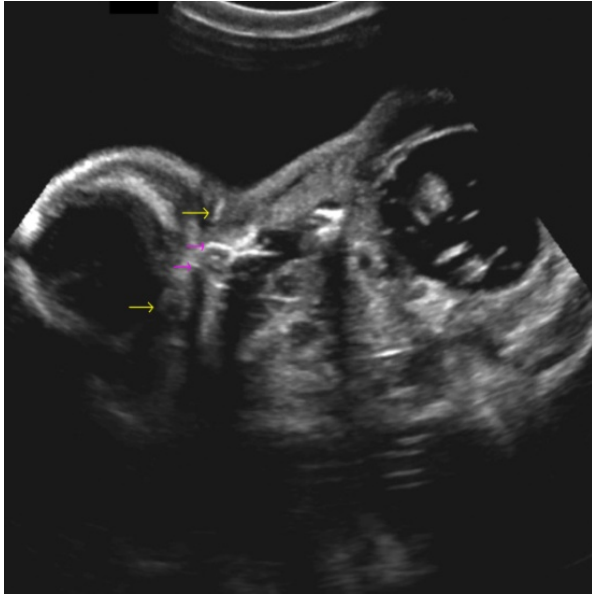


FIGURE 1d: Coronal Doppler ultrasound image demonstrates numerous vessels within the solid portions of the mass.

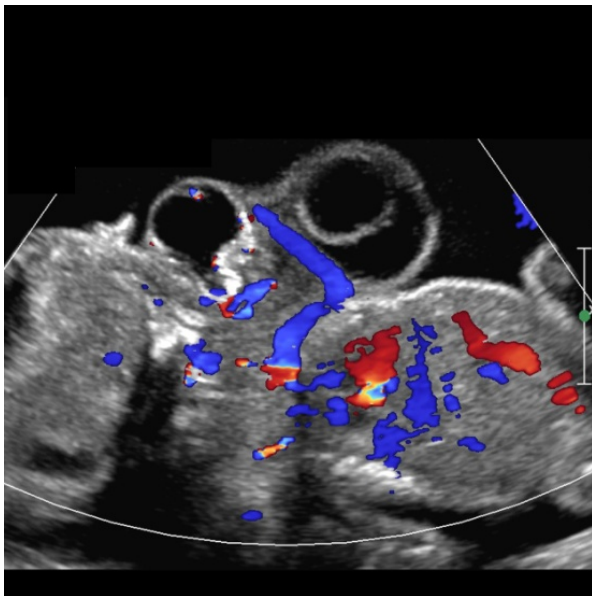


FIGURE 1e: 3D ultrasound image of Twin A's face shows a protruding tongue. The stomach (not shown) contained no fluid, and polyhydramnios was present. These findings suggest impaired fetal swallowing.



FIGURE 2a: After delivery of the twins, postmortem MRI was performed. Sagittal T2 weighted MR image shows a heterogeneous solid and cystic mass extending through a 3 cm defect in the left skull base (pink arrows), just behind the left eye (yellow arrow).

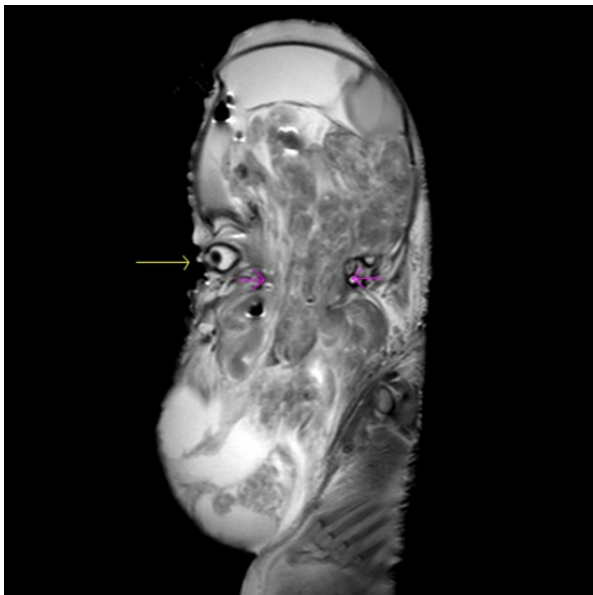


FIGURE 2b: Coronal T2 weighted MR image shows displacement of the brain parenchyma to the right and ventriculomegaly.

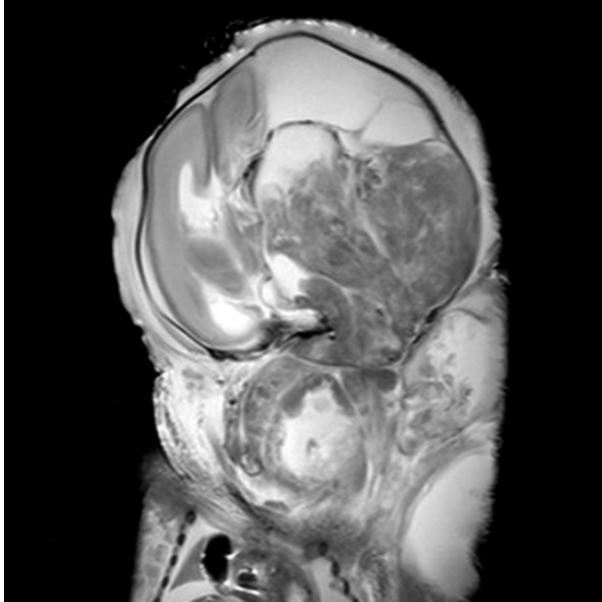


FIGURE 3: Postmortem photograph of Twin A. Note the protruding tongue and lateral displacement of the left eye.



Diagnosis: Congenital Teratoma.

Discussion:

Although teratomas constitute less than 5% of all pediatric neoplasms, they are the most common fetal

tumor and are most frequently sacrococcygeal [1, 2]. In addition, they are the most common congenital intracranial tumor, comprising at least 50% of cases [2].

A teratoma is a neoplasm containing tissues foreign to the site of origin or containing more than one embryonic germ cell layer. Extragonadal teratomas occur more often in children and arise from anatomically misplaced pluripotent germ cells [3, 4]. Most (90%) childhood teratomas contain components of all three germ cell layers, and 20-40% contain immature elements [2, 3, 4]. Up to 95% of head and neck teratomas contain mature or immature neuroectodermal elements, whereas the minority (34%) of sacrococcygeal teratomas contain neuroectodermal elements [3, 4]. While sacrococcygeal teratomas have a clear female predominance, intracranial teratomas occur in a slight male predominance [5]. Pineal region teratomas have an overwhelming male predominance [4]. Most intracranial teratomas diagnosed prenatally are supratentorial, whereas those diagnosed in older children are usually infratentorial [3].

Intracranial teratomas are typically midline and may be massive and locally invasive, replacing the intracranial contents, eroding the skull and extending into the mouth, orbit and neck, as seen in this case [1, 2]. They are most often diagnosed in the third trimester [1]. Often the masses are so large that the precise anatomic point of origin cannot be determined; the most common sites of origin include the pineal gland, suprasellar region, and cerebral hemispheres [1]. Prenatal manifestations of congenital intracranial teratoma include polyhydramnios (from impaired fetal swallowing due to both mass effect and hypothalamic dysfunction), hydrops fetalis (from arteriovenous shunting) and rapidly increasing head circumference or uterine size (due to the enlarging tumor) [1, 3]. Intratumoral hemorrhage may also occur and thus in the setting of any fetal intracranial hemorrhage, underlying neoplasm should be considered [1].

Typical ultrasound features are a mixed solid and cystic mass [1]. On color Doppler, there may be increased vascularity with low-resistance flow, leading to high-output cardiac failure and hydrops fetalis; vascular flow is a helpful finding to distinguish from hemorrhage [1, 2]. CT findings include a large heterogeneous mass with coarse calcifications and hydrocephalus [5]. Although the presence of calcifications on CT or ultrasound is a helpful diagnostic clue they frequently are not present [1]. MR findings are highly variable, but often demonstrate a large multicystic heterogeneous mass [2,3]. MR is helpful to determine the anatomic extent of the tumor, and to differentiate mass from intracranial hemorrhage [1].

The differential diagnoses of congenital supratentorial tumors include primitive neuroectodermal tumor (these contain calcifications but not fat, a finding which may help distinguish from teratoma), glioma, astrocytoma, choroid plexus papilloma, craniopharyngioma and meningioma [1, 2, 3, 4].

The prognosis for congenital intracranial teratoma is extremely poor whether benign or malignant, with an overall mortality rate of 90% rising to 97% if diagnosed prior to 30 weeks gestation [1, 3]. After diagnosis, termination and supportive care are offered [1]. If vaginal delivery is planned, cephalocentesis

may be necessary [1]. Caesarian section may be required to prevent dystocia or, in cases of fetal airway obstruction, to establish a fetal airway [1]. With the ex utero intrapartum delivery procedure (EXIT), the fetus is partially delivered via Caesarian section and tracheostomy is performed while uteroplacental circulation is maintained [1].

References:

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