Abstract: Teratoma originates from multipotent germ cells and consists of all 3 embryological germ layers. It is commonly seen in sacrococcygeal region and is very rarely seen in head and neck region of a neonate. It is even rarer for a cervical teratoma to present as a swelling in the lateral aspect of neck causing airway obstruction. This presentation is about a boy baby who had a large cervical teratoma on the left side of neck with severe respiratory compromise due to the pressure effect. Airway was secured with great difficulty. In spite of good surgical excision of tumour, respiratory failure progressed and the child succumbed. Histopathology of tumour revealed immature teratoma—grade III.

Keyword: Neck swelling, immature teratoma, airway obstruction

Introduction:
Teratomas are complex tumours composed of a collection of heterogeneous cells derived from all three germ cells. Neonatal teratomas occur in about 1:20,000 to 1:40,000 live births 1, 2. Sacrococcygeal teratoma, the commonly occurring tumour, accounts for 40% and only 5% of teratomas occur in head and neck region in a neonate. Congenital cervical teratomas are mostly benign and are said to be amenable to curable resection but the site of their origin and associated respiratory compromise make their treatment challenging. Most congenital cervical teratomas seen in the neonatal period are midline in position and mature in nature.

Case report:
A male infant weighing 1.68 kg was born preterm at 33 weeks gestation to a 26-year-old primigravida mother. The child was delivered by emergency cesarean section in view of polyhydramnios and fetal distress, and was referred from a private hospital at 20 hours of life. Second trimester fetal ultrasound revealed large neck mass and polyhydramnios. The infant did not establish breathing at birth and required resuscitation. The child was intubated with difficulty, and further needed continued respiratory support. On examination, the infant had a mass (6 x 5 cm) on the left lateral aspect of neck (Figure 1) which was predominantly solid and partially cystic in consistency. Transillumination test was negative and no bruit was elicited. Radiological work up showed soft tissue mass (Figure 2) and tracheal shift. Ultra sonogram revealed heteroechogenicity in ultra sonogram. Further evaluation revealed elevated Alpha-fetoprotein, normal human Chorionic Gonadotropin and negative urinary Vanillyl Mandelic Acid. FNAC (Fine needle aspiration cytology) report showed clusters of small round to oval cells (Primitive mesenchymal cells) (Figure 3a) with rosettes (Figure 3b) in the background of eosinophilic material.

In view of increased ventilatory requirements and impending respiratory failure, surgical intervention was planned. The mass was extending from the sub mental region superiorly to the clavicle inferiorly crossing the midline of neck, and peroperatively...
the size of the mass was 7 X 6 cm. Carotid vessels were splayed out laterally. Only subtotal excision was possible due to the adherence of tumour to trachea. Despite surgical intervention and continued respiratory support, the baby succumbed on the 2nd post-operative day. The cut section of the mass revealed predominantly solid interspersed with some cystic areas (Figure 4). Microscopic sections revealed tissues derived from all three germ cell layers. A variety of tissues resembling stratified squamous epithelium (figure 5 a), neuroepithelium with rosette (figure 5 b), mucinous gastric epithelium (figure 5 c), cartilage (figure 5 d), salivary glands and undifferentiated mesenchymal tissue were seen. Many foci of immature neuroepithelial tissues were observed, suggestive of immature teratoma grade III.

Figure 4: Cut section of mass (predominantly solid with cystic areas)

Figure 5 a Figure 5 b

Figure 5 c Figure 5 d

Figure 5: Histopathological examination of tumour: (a) stratified squamous epithelium (b) primitive neuroepithelium with rosette (c) mucinous gastric epithelium (d) mature cartilage

Discussion:
Teratomas are tumours containing components of all the three embryological germ layers originating from multi-potent germ cells 4, 5. Sacrococcygeal area is the most common site (40%) of the tumour in a neonate while the head and neck are unusual sites of presentation accounting for only 5% of the cases. Only three cases of neck teratomas have been reported in India 6. Teratomas of the head and neck are intriguing because of their obscure origin, bizarre microscopic appearance, unpredictable clinical course and often dramatic clinical complications7. Differential diagnosis of congenital neck masses in newborn are cystic hygroma, teratoma and neuroblastoma. Of all these, the most common neck mass in neonates is cystic hygroma. It is cystic in consistency with thin walls, septations and transillumination is usually positive 8. Most patients with cervical neuroblastoma present as a well-defined solid mass with or without intraspinal extension or calcification and urinary Vanillyl Mandelic Acid positivity.

Most of the cervical teratomas are midline9 in position and extend into mediastinum, causing tracheal compression presenting with respiratory distress, requiring ventilation and difficulty in swallowing due to esophageal compression. It is unusual for tumours on the lateral side of neck to present with respiratory distress. Our baby showed midline extension of tumour and was tethered to the trachea causing not only respiratory problem but also difficulty in resection, resulting in subtotal excision only. Ninety percent of cervical teratomas are documented in the pediatric population; however a minority can be seen in adults10. In our case, the mass was detected in fetal life itself and it presented as a lateral neck mass which is an unusual occurrence. Approximately one third of cervical teratomas are reported to be associated with hydramnios and the same was observed in our case. The majority of cervical teratomas presenting in infancy and early childhood are mature in nature and exhibit a benign clinical course, whereas those presenting after the first decade of life often may undergo malignant change.11 Histopathology of the mass in our case revealed immature teratoma Grade III which carries a poor prognosis. Giant neck masses are associated with difficulty in securing the airway and perinatal asphyxia which adversely influence the outcome. Further mortality and morbidity may be increased by complications like pneumonia and hemorrhage into the tumour. Associated tracheal compression sometimes causes tracheomalacia, increasing the post-operative morbidity.12 Head and neck teratomas in children are mostly benign, amenable to resective excision but the time of detection, site and size of the tumour determines the outcome. The retrospective review of reported literature on “Perinatal (fetal and neonatal) germ cell tumors” by Isaac Jr et al, described the following findings; Mass was noted either by antenatal sonography or postnatal clinical examination. Neonates with cervical teratomas generally have good outcome provided, if tumor is resectable. Fetuses have a lower survival rate compared to neonates, 23% versus 85% respectively. Ten percent of the fetuses were stillborn. After surgical resection survival rate was 88.5% and overall survival rate was 68%.3. The natural history of this condition aids the clinician to assist parents in their informed choice. Anticipation and conduct of delivery in a well-equipped center, swift action during delivery to establish breathing and prompt surgical intervention after initial stabilization are the key elements in management of cervical teratoma. A retrospective study done by Children’s Cancer Group on cervicofacial teratomas suggested two techniques for securing airway (EXIT and OOPS) 13-15. The first one is Ex-utero Intrapartum Treatment (EXIT) which involves performing a mini incision with exteriorizing the head or the part of the fetus containing the tumour, while the placenta remains in the uterus. The airway of the fetus is visualized by laryngoscopy or bronchoscopy and child undergoes intubation/tracheostomy. The second technique is an Operation On Placenta Support known as OOPS. In this procedure full delivery of the fetus is performed and the neonate’s airway is secured on surgical table with intact placenta.

Conclusion:
Teratoma should be in the list of differential diagnosis for a neck mass presenting in fetal or neonatal period. Airway compromise is the most serious postnatal complication of cervical teratomas. Prenatal diagnosis helps in anticipation of the airway compromise and effective treatment such as EXIT procedure can be executed during perinatal period. Early anticipation and planned delivery in tertiary level hospital helps in timely management of these cases. A multidisciplinary team should be involved for securing the airway. Tumour needs to be removed completely as soon as the newborn's condition is stabilized. Fetuses with teratomas detected antenatally have 3 times the mortality rate compared with postnatally diagnosed neonates.
Though surgical intervention might be curative in most cases, some may have poor result despite appropriate management, due to severe tracheal compression or non-resectable condition of the mass or immature nature of the tumour.

References: