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Cervical Teratoma and Cystic Hygroma in Nigerian Infants: Case Studies of Two Differential Diagnoses of Neonatal Neck Mass and Review of the Literature

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Abstract

Congenital neck masses include branchial cleft cysts, thyroglossal duct cysts, thymus cysts, dermoid and teratoma, vascular abnormalities, and lymphatic malformations such as cystic hygroma. Cervical Teratomas (CTs) are rare true neoplasm of the neck composed of tissues derived from at least two of the three embryonic germ layers but foreign to the anatomic site of occurrence. CTs occur on the anterolateral surface of the neck, extending midline from the thyroid gland. They are asymmetric and multinodular with a cystic-solid consistency. In contrast, Cystic Hygromas (CHs) are benign multiloculated, compressible, painless lymphatic lesions with a doughy consistency. CHs can occur in the submental triangle, with extension into the floor of mouth. Early neonatal presentation with aero-digestive obstruction is the norm for large CTs and CHs. We present two infants with huge CT and CH delivered in two consecutive years at our facility in south-western Nigeria by unrelated families. Definitive prenatal diagnoses were not made and deliveries were not pre-planned. Both infants had severe respiratory compromise and unfavorable outcome. This report aims to enhance clinical recognition of these rarities, highlight their occurrence in our locale and reiterate the associated management challenges in resource-limited settings. Relevant literatures are also reviewed.

Keywords: Cervical teratoma; Cystic hygroma; Nigerian infants

Introduction

Congenital neck mass comprises anomalies of the branchial arches (branchial cleft cysts, thymic cysts), the thyroid gland (thyroglossal duct cysts), the germ line tissue (dermoids, teratomas) and the cervical neuro-vasculature (cystic hygroma, neurofibromas) [1,2]. Cervical Teratomas (CTs) are rare true neoplasm of the neck composed of tissue which are derived from at least two of the three embryonic germ layers (ectoderm, mesoderm and endoderm) but foreign to the anatomic site of occurrence [3,4]. Only 150 cases of cervical teratoma have been reported in the literature [5]. CTs account for approximately 3% of all teratomas in the neonatal period [6]. They can grow rapidly, resulting in hydrops fetalis, polyhydramnios, and fetal demise. Asphyxia and early neonatal death commonly result from airway compromise among live births [5]. They often occur on the anterolateral surface of the neck, extending midline from the thyroid gland, as far as 12 cm in their longest axis [1]. They are asymmetric and multinodular with a cystic-solid consistency [6,7]. Ectodermal tissue predominates and fine calcifications are present in nearly a half of cases [3,4]. Current evidence suggests that most teratomas are due to abnormal differentiation of fetal germ cells that arise from the fetal yolk sac. Normal migration of these germ cells may cause gonadal tumors, while abnormal migration produces extragonadal tumors [8]. Also, perhaps, CT arises from stem cells within the thyroid gland that grow abnormally into a tumor [7]. There is no apparent relationship to the mother's age [8]. These tumours are mostly benign with favourable outcomes if diagnosed and excised promptly [8]. Total surgical removal is essential to avoid local recurrence and malignant degeneration [9].

Similarly, Cystic Hygroma (CH) is a rare benign cystic lymphatic lesion that can affect any part of the human body [10]. It is synonymous with cystic lymphangioma, which is also known as a macrocystic lymphatic malformation. Nearly three-quarters of CHs occur in the head and neck, with a left-sided predilection [10,11]. Within the neck, the posterior triangle tends to be most frequently affected [10]. Internationally, the incidence of CH is estimated to be 1 case per 6,000-16,000 live births [10]. It affects all races but a reduced incidence has been reported in African Americans [10]. The sex distribution is equal. Most cystic hygromas (50-65%) are evident at birth, with 80-90% of CHs presenting by age 2 years [10,11]. Clinically, CH is a multiloculated, compressive, painless, transluminant mass with a doughy consistency [10,11]. It can occur in the submental triangle, with extension into the floor of mouth [10,11]. CHs are thought to arise from a failure of lymphatics to connect to the venous system, abnormal budding of lymphatic tissue, and/or sequestered lymphatic rests that retain their embryonic growth potential [10,12]. These lymphatic rests can penetrate adjacent structures or dissect along fascial planes and eventually become canalized. These spaces retain their secretions and develop cystic components because of the lack of a venous outflow tract [10-12]. Occasionally, it is acquired due to trauma or obstruction of a lymphatic drainage pathway [13]. On ultrasonography, CH is a multi-cystic lesion with internal septation [11]. Surgical excision is desirable [10,11].

In a 58-year span institutional review by Jordan et al. in Cleveland, only five live newborns with CTs were found [14]. Also, in a recent analysis of over ten thousand deliveries in our sister facility, no single case of CT was seen [15]. Nonetheless, this report aims to enhance clinical recognition of these dual rarities, highlight their occurrence in our locale and reiterate the associated management challenges in resource-limited settings. Relevant literatures are also reviewed.

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Case Description

The following infants with huge cervical masses were delivered in two consecutive years at the Mother and Child Hospital, Ondo, Nigeria by non-consanguineous parents. The two families were unrelated. Definitive prenatal diagnoses were not made and deliveries were not pre-planned. Both infants had severe respiratory compromise and poor outcome as detailed below:

Case 1

A full term female infant was delivered October 10, 2014 to a 24 year old Para 3^{+0} woman via emergency Caesarean section, secondary to fetal bradycardia and intra-partum ultrasound diagnosis of a neck mass. Mother was not booked, but referred in labour from a rural health center to our facility. Besides prenatal history suggestive of polyhydriamnos, the mother was otherwise stable during her antenatal period.

The Baby was delivered limp with a huge cervical mass. She was apneic, centrally cyanosed with a heart rate (HR) of 50 beats per minute. Her APGAR scores were 1^{1min} and 2^{5min}. She was resuscitated in line with standard resuscitation guidelines [16]. An appropriately sized Oro-pharyngeal Airway (OPA) was inserted. Initially, she was ventilated using a bag-and-mask device connected to an oxygen source. At the Neonatal Intensive Care Unit (NICU), she was intubated with an un-cuffed size 2.5 mm ID endotracheal tube with significant difficulties due to gross distortion of her neck anatomy (Figure 1). Intermittent Positive Pressure Ventilation (IPPV) was continued manually and HR increased to 139 beats per minute but percutaneous oxygen saturation (SPO_2) remained 60-67%.

The huge left-sided cervical mass extended from the anterolateral aspect of the face posteriorly to involve the neck almost circumferentially (Figure 1). The mass was irregularly-shaped with visible superficial vessels and areas of dark discoloration. Also, there were areas of cystic and solid consistency. There were no other obvious malformations in this infant.

A diagnosis of giant congenital cervical teratoma with airway compromise was made. A referral to a tertiary institution for a cervicofacial Computerized Tomography (CT) scan and possible surgical excision was planned but unattained due to financial constraints. The baby died at the fourth hour of life.

Case 2

A male infant delivered at a Gestational Age (GA) 34 weeks to a 28 year old Para 2^{+0} woman via emergency lower segment Caesarean section indication being ultrasound diagnosed huge neck mass (?cystic hygroma) with eclampsia and polyhydriamnos. Date of delivery was February 02, 2013. Antenatal period was otherwise uneventful.

He had moderate perinatal asphyxia with APGAR scores of 5^{1min} and 7^{5min}. His anthropometric measurements were as follows: birth weight 4.5 kg, Occipitofrontal Circumference (OFC) =35 cm and length 41 cm. He was in respiratory distress. He had a cystic, soft, transluminant cervical mass extending from the left temporal region to the lower part to the jaw, measuring 29×31 cm without tenderness or bruit. Further systemic examinations were normal. He received oxygen at 3-5 liters/minute via nasal prong with an SPO_2 of 96%, HR=144 beats/minute and respiratory rate=44 cycles/minute. Random blood sugar was 4.0 mM/L and euglycemia was maintained with intravenous fluid 10% dextrose water. Haematocrit was 43%. At the 3^{rd} hour of life, he had intermittent apneas but he was successfully resuscitated (Figure 2).

He was later referred to the pediatric surgical team of the State Specialist Hospital, Akure Nigeria for a definitive management.

Discussion

The most significant clinical manifestation of Cervical Teratoma (CT) and Cystic Hygroma (CH) in early neonatal life is respiration compromise as found in our patients who were in severe respiratory distress at delivery [11,14]. This is due to direct pressure effects of the masses on the underlying neck structures and possible mediastinal involvement [14]. Although small CT and CH may be asymptomatic until early childhood, neonatal presentation is the norm for huge masses [11,14]. Other possible effects or complications of these lesions include dysphagia, infections and heart failure, as well as spontaneous bleeding or rupture of CH [11,14]. Sometimes CT and CH may contribute to fetal demise especially when there are coexisting malformations [11,14]. We uphold the clinical diagnoses of CT and CH in case 1 and 2 respectively based on the classical asymmetric, multinodular, cystic-solid consistency of the former and the compressible, non-tender, brilliantly transluminant nature of the latter [7,11,14]. On ultrasonography, CH is a multi-cystic lesion with internal septation and no blood flow on colour doppler ultrasonography [11].

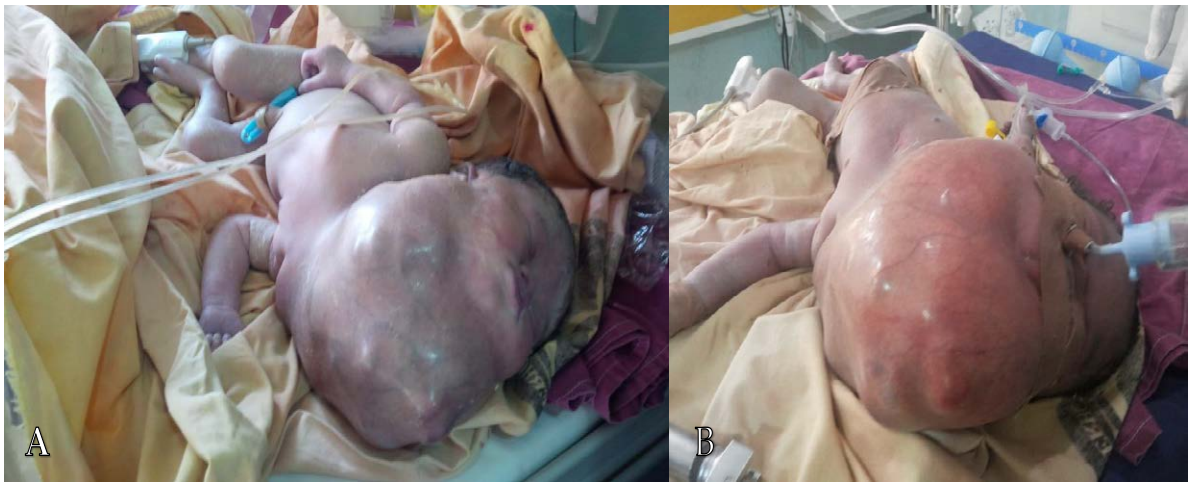

Figure 1: A giant congenital Cervical Teratoma (CT) in a Nigerian infant; (A): the asymmetric multi-nodular mass kept the neck in a hyperextended position with severe airway compromise and cyanosis at birth; (B): shows some improvement in her level of oxygenation post-intubation.

Figure 2: Shows a huge left-sided Cystic Hygroma (CH) in a Nigerian neonate; (A): there is significant respiratory compromise visible as intercostal and subcostal recessions with cyanosis due to its pressure effect on the neck and the ipsilateral chest wall; (B): improved oxygenation achieved with intranasal oxygen and positioning with supports (evidenced by the reduced duskiness of the lips, body and neck mass).

Although histology was unachieved in our CT case, mature teratomas most frequently show adult type tissue derided from the embryonic layers especially skin and adnexa, bronchial mucosa and gland, as well as gastrointestinal mucosa [7,14].

Prenatal diagnosis of CT and CH has been described using ultrasonography. CTs are typically seen as large asymmetric, well defined, one sided neck masses often associated with polyhydriamnos [5,17]. Rarely, open fetal surgery may be necessary if hydrops (in-utero heart failure) develops before 30 week GA [17]. Also, CH is often identified as a multilocular fluid-filled cavity visible as increased nuchal translucency greater than 99th percentile for gestational age in the first trimester [18,19]. However , over 80% of simple CH with normal karyotype resolve before delivery [19]. Moreover it is desirable to diagnose CT prenatally in order to plan delivery and postnatal care in a designated centre to improve outcome [14]. Nonetheless, early prenatal diagnosis was not attained in our patients, apparently due to sub-optimal antenatal care and non-availability of ultrasound scanning machines in the referring rural facilities.

In addition, current trend in the management of CT includes Ex-utero Intrapartum Treatment (EXIT) procedure to secure the newborn airway at delivery while still on placental support and early surgical extirpation of the tumour [5,20]. The tumor should be completely removed in order to prevent recurrence or malignant transformation [14]. Similarly, a complete surgical excision of CH is the treatment of choice [11]. Other management modalities include cauterization, marsupialization and sclerotherapy with bleomycin, triamcinolone or fibrin sealant [11]. Also, radiofrequency ablation may be used in persistent or unresectable CH. CH reoccurs in about 20% of cases after treatment [11]. Pre-operative computerized tomography will ascertain the extent of CT and CH, their association with neurovascular structures, prevent iatrogenic damage during surgical excision and enhance prognosis [7,11].

In conclusion, large CTs and CHs are potentially life-threatening neonatal emergencies due to the associated severe airway obstruction. Prenatal diagnosis via optimal antenatal care including prenatal ultrasonography will ensure well planned deliveries and favorable outcome of these rare congenital disorders in resource-limited settings.

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