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A CASE OF IMMATURE TERATOMA MASQUERADING AS CYSTIC HYGROMA

Jai Durairaj Paramasivam[1], Divya Kandasamy Ramamoorthy[2*], Kumutha Jayaraman[3], Sonti Sulochana[4], Surya Rao Rao Venkata Mahipathy[5], Harish Sudarsanan[6]

[1]Associate Professor, Dept. of Paediatric Surgery, Saveetha Medical College & Hospital, Saveetha Institute of Medical and Technical Sciences, Thandalam, Kanchipuram Dist. – 602105, Tamilnadu, India
[2]Resident, Dept. of Plastic & Reconstructive Surgery, Saveetha Medical College & Hospital, Saveetha Institute of Medical and Technical Sciences, Thandalam, Kanchipuram Dist. – 602105, Tamilnadu, India
[3]Professor & Head, Dept. of Neonatology, Saveetha Medical College & Hospital, Saveetha,Institute of Medical and Technical Sciences, Thandalam, Kanchipuram Dist. – 602105, Tamilnadu, India
[4]Professor, Dept. of Pathology, Saveetha Medical College & Hospital, Saveetha Institute of Medical and Technical Sciences, Thandalam, Kanchipuram Dist. – 602105, Tamilnadu, India
[5]Professor & Head, Dept. of Plastic & Reconstructive Surgery, Saveetha Medical College & Hospital, Saveetha Institute of Medical and Technical Sciences, Thandalam, Kanchipuram Dist. – 602105, Tamilnadu, India
[6]Associate Professor, Dept. of Neonatology, Saveetha Medical College & Hospital, Saveetha Institute of Medical and Technical Sciences, Thandalam, Kanchipuram Dist. – 602105, Tamilnadu, India

ABSTRACT
Teratomas are more common in childhood and fetal life and are histologically complex.Immature teratomas contain combinations of mature epithelial and connective tissues with immature areas of mesenchymal and neuroectodermal tumours. Here we present a case of immature teratoma with clinical picture mimicking cystic hygroma in all the antenatal & postnatal imaging modalities. The gold standard treatment for benign masses is surgical excision with malignant tumours requiring combination of surgery, chemotherapy and radiotherapy.

KEYWORDS: Antenatal, Immature, Teratoma, Surgical excision

INTRODUCTION
Teratomas of head and neck have an obscure origin, varied pathological appearance and have a dramatic presentation. Cervical teratomas are rare extra-gonadal teratomas and constitute a small percentage (1.6% - 9.3%) of pediatric teratomas which equates 1 per 40,000 births.[1] Less than 200 cases of teratoma have been reported globally and only 3 case reports from India with only 1 of them surviving.[2] The majority of infantile and early childhood cervical teratomas are mature and clinically benign while those presenting later exhibit malignant behaviour. A study with 65 cases having childhood teratomas under the age of 7 years revealed only 4 cases to have cervical teratomas (3 mature and 1 immature).

CASE REPORT
A 38 year old primi with a history of In Vitro Fertilisation (IVF) treatment and twin conception came at gestational age of 37 weeks 3 days for safe confinement. The mother had no comorbidities apart from her antenatal scan in the fifth month which showed twin A to have a multiseptated cystic lesion in the anterior aspect of midline of neck in the subcutaneous plane. Colour flow mapping showed no significant vascularity in the lesion. The diagnosis suggested was lymphatic abnormality or cystic lymphangioma. Twin B was normal. Emergency LSCS was done. Both male newborns had adequate birth weight of 3.1kg/3kg and had APGAR score of 8. In view of respiratory distress immediately after birth twin A was intubated. Clinical examination of twin A showed a large swelling in the anterolateral aspect of neck, extending from lower part of neck upto mandible and from lateral aspect of neck upto midline. Skin over the swelling was stretched and swelling was tense cystic in consistency, fluctuant and transilluminant. Routine blood investigations and thyroid profile were normal. X-ray showed soft tissue swelling in the left lateral aspect of neck with tracheal deviation to right. Ultrasonogram revealed a large multiloculated cystic lesion 10x9x7cm in size and 2mm thick in left lateral neck extending upto midline. MRI neck was done and showed large nonenhancing cystic lesion involving left lower neck soft tissue region, causing displacement of adjacent structures and no definite intracranial/intrathoracic/intraorbital extensions with all features suggestive of cystic hygroma. (Fig 1 a,b)

In view of large swelling with respiratory distress excision biopsy was planned 3 days postnatally.(Fig 2a) Under anesthesia elliptical incision was made in lateral aspect of neck. Extension was from trachea anteriorly to spinous process posteriorly. A straw coloured fluid

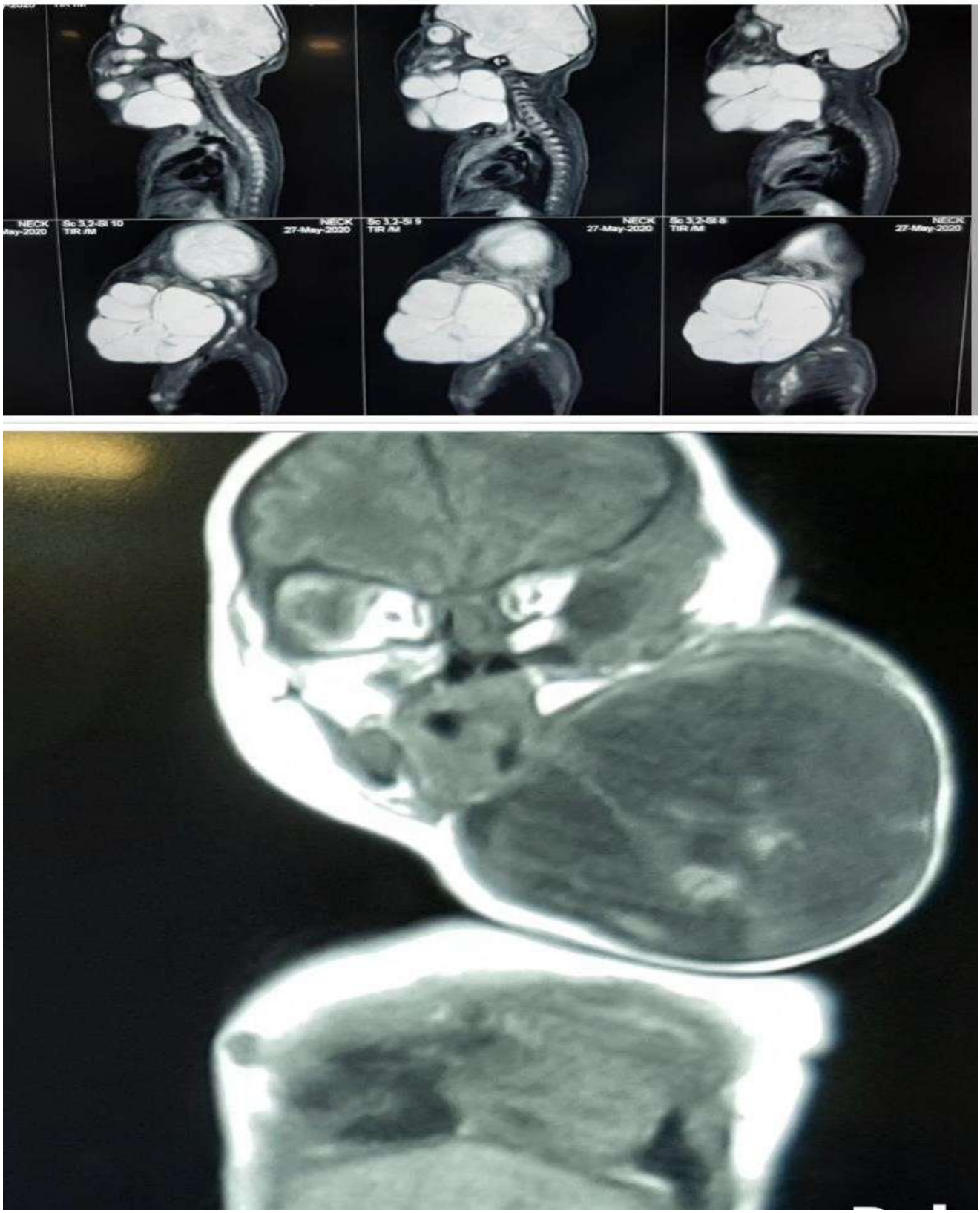

Fig. 1a,b - MRI neck was done and showed large non-enhancing cystic lesion involving left lower neck soft tissue region, causing displacement of adjacent structures and no definite intracranial/intrathoracic/intraorbital extensions

was drained from the multiloculated cyst. There was no adhesion with adjacent structures and they were all preserved. Cyst was excised in toto and sent for biopsy. (Fig 2b) Postoperatively, the baby was managed with elective ventilation for 48 hours. (Fig2c) After extubation baby started nasogastric feeds on 2nd post-operative day. Histopathology revealed a diagnosis of immature teratoma (grade1). Following surgery alpha feto protein (AFP) was raised and thyroid profile revealed hypothyroidism. The child was advised to take thyroxine tablets 12.5mg everyday and was asked to come for follow up with AFP value monitoring and imaging every 3 months.

DISCUSSION

Teratomas are derived from 3 germ layers - ectoderm, mesoderm, endoderm. Thus they have tissues of all germ layers. Most teratomas present in fetal life or childhood and are uncommon in adults. There is preponderance based on gender in childhood teratomas as 80% of all teratomas occur in females. Teratomas may be solid, cystic or mixed and are classified into mature and immature types. Thus many congenital cervical teratomas can be mistaken for cystic hygroma. Mature teratomas contain adult tissues with varied organic development whereas immature type contains embryonic/extra embryonic tissue. Upto 90% contain all 3 germ layers and 20-40% contain immature tissues. 75-85% cervical teratomas have both mature and immature features.

Location has been associated with age, as evidenced by extragonadal tumours occurring primarily in neonates and young children whereas gonadal tumours are most noted in adolescents. Although teratomas are may occur anywhere along the midline they are most commonly found in the sacrococcygeal region (60%) and rarely found in the neck (3.4%). Other sites include mediastinum, retroperitoneum and gonadal locations. Cervical teratomas are said to arise from the embryonic thyroid

anlage and clinical features are mainly due to pressure effects of the tumours. One third of reported cervical teratomas complicate maternal hydraminos.

Cervical teratomas (2%) are less common than cervical lymphatic malformation (6%). Teratomas although are usually benign have a chance of turning malignant while cystic hygromas never turn malignant. Structurally teratomas can be cystic / solid sometimes calcification and mostly occur in the midline while cystic hygromas can be multicystic/macrocystic and occur in the lateral area. Lymphatic malformations require sclerotherapy apart from surgery and the thyroid function remains normal unlike in cervical teratomas where there is a possibility of origin of mass from thyroid tissue giving an altered thyroid profile.[1,3] Both these types of cervical masses can present with respiratory distress but its less acute in cystic hygroma. In summary, there can be virtually no differences between cervical cystic hygroma and cervical teratoma when majority of the mass are cystic with no calcification. Thus careful pathological examination is essential.

Grading of teratomas are important for management but is challenging. There are 4 grades as per Gonzalez-Crussi grading system; 1) Well differentiated tissue, 2) Occasional foci (less than
10%) contain incompletely differentiated tissue, 3) 10-50% of immature tissue. Grade 0 & 1 are considered mature teratoma and grade 2 & 3 are considered immature teratoma. Malignant teratomas contain areas of germ cell tumour, non-germ and immature teratoma with metastasis. Diagnosis between mature and immature teratoma can be challenging if it is not composed of extensive unusual neuroectodermal tissue component. CT scan is superior to MRI only for giving staging of the tumour and for identifying vascular involvement. Surgery is the preferred treatment.[4] Prognosis depends upon factors such as degree of maturation of tissues, completeness of resection, airway obstruction and associated anomalies.[5] Long term follow up is done by monitoring tumour marker levels and MRI scanning.[6]

CONCLUSION

Congenital cervical teratomas are usually benign and have a good prognosis. Due to the various components of these tumours and due to their predominant incidence in childhood they must be considered as a differential diagnosis along with cystic hygroma for congenital lesions presenting as a cystic neck mass.

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