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Case Report

Diagnostic imaging and surgical management of a congenital cervical teratoma

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الملخص

التيراتومات (الأورام المسخية) العنقية الخلقية هي أورام نادرة، تنشأ في الرقبة وتتكون من ثلاث طبقات نسيجية رئيسة من أنسجة الجنين: الأديم الظاهر، والأديم المبطن، والأديم المتوسط. والغالبية العظمى من التيراتومات العنقية هي أورام حميدة. إلا أن الأهمية السريرية لهذه الأورام تنشأ من مضاعفاتها أثناء الحمل بسبب أثر الكتلة بعد الولادة على مجرى الهواء والمريء.

من الممكن تشخيص التيراتومات العنقية الخلقية من خلال الفحص بالأشعة فوق الصوتية في وقت مبكر قبل الولادة. ويعتمد الشكل على حجم الورم، وعادة ما تظهر ككتلة عنقية كبيرة بمحتويات صلبة وتكيّسية، تتسبب في رجوع مفرط للرقبة إلى الخلف، وكثيرا ما تُصحب بالاستسقاء السلوي. تساعد الأشعة فوق الصوتية في فترة ما بعد الولادة في تمييز التيراتومات العنقية الخلقية من الكتل العنقية الخلقية الأخرى الشائعة. وأفضل طريقة لتقييم صلابة الورم، وامتداده للأنسجة المحيطة وتأثيره ككتلة على محتويات الرقبة هي التصوير بالرنين المغناطيسي. في تقرير الحالة لدينا نعرض طفلا نتج عن حمل كامل ووُلد بكتلة عنقية ضخمة. كان التصوير بالرنين المغنطيسي مفيدا في إظهار محتواها، وامتدادها للأنسجة المحيطة، وتأثيرها ككتلة على محتويات الرقبة. والترسيم الواضح للكتلة يسهل الإزالة الجراحية الكاملة دون مضاعفات.

الكلمات المفتاحية: التيراتومات العنقية؛ الأورام المسخية؛ الخلقية؛ كتلة عنقية؛ كتلة عنقية رخوة؛ الاستئصال الجراحي

Abstract

Congenital cervical teratomas are rare tumours arising from the neck and consist of three major tissue layers of an embryo: the ectoderm, endoderm, and mesoderm. A great majority of cervical teratomas are benign tumours. However, the clinical significance of these tumours arises from the complications they can cause during pregnancy due to the postnatal mass effect on the airway and oesophagus of the neonate.

Diagnosis of a congenital cervical teratoma is possible during an early prenatal ultrasound evaluation. The appearance depends on the size of the tumour, but it is typically a large neck mass with solid and cystic components that causes hyperextension of the neck and is frequently associated with polyhydramnios. In the postnatal period, ultrasound helps in differentiating cervical teratoma from other common congenital cervical masses. MRI is the modality of choice to evaluate the consistency of the tumour, surrounding soft tissue extent of the tumour, and any mass effect on other cervical structures. In our case report, we present a case of a full-term baby that was delivered with a large cervical mass. MRI was helpful in demonstrating the complex content of the mass, surrounding soft tissue extension, and mass effect on other major cervical structures. The clear demarcation of the mass facilitated complete surgical removal without complications.

Keywords: Cervical soft tissue mass; Cervical teratoma; Congenital; Neck mass; Surgical excision

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Introduction

Congenital cervical teratomas are rare germ cell tumours arising from the neck. Most of these tumours are benign, though malignant transformation has been rarely described. They are usually diagnosed during the prenatal period and require multidisciplinary management to improve the prognosis. Ultrasound and MRI evaluation are very useful to differentiate these tumours from other common congenital cervical masses, such as lymphatic malformations (cystic hygromas), through evaluation of their vascularity and soft tissue content.

The purpose of this article is to describe the radiological manifestations of congenital cervical teratomas and highlight the main differentiating points compared with other similar masses such as cystic hygromas.

Case report

Our case is a full-term baby girl, born by spontaneous vaginal delivery, who had a large neck mass discovered at birth. On physical examination, the mass was warm and more prominent on the left side of the neck with a firm, multicystic consistency (Figure 1). Initially, the newborn had stable vital parameters and no respiratory compromise. However, she developed respiratory distress and was intubated on the 5th day after birth. Chest radiographs performed at birth showed a large neck mass with faint scattered calcifications. MRI revealed a large left anterolateral multi-loculated complex mass lesion measuring 7 × 8.5 × 7.5 cm and extending from the level of the mandibular angel to the level of the thoracic inlet. The mass displaced the oesophagus and airway to the contralateral side and attenuated the left internal carotid artery and left internal jugular vein. The radiological appearance was characteristic of a congenital cervical teratoma (Figure 2).

The infant underwent surgical excision through a left transverse incision directly over the mass. The tumour surface was exposed in the sub-dermal plane. A capsule was present, allowing a plane of dissection of the mass from the surrounding structures, which were displaced but not infiltrated. It was lobulated grey-white mass (Figure 3). Complete excision of the mass without rupture of the capsule rupture was successfully performed and the specimen was sent for histopathological examination.

Histopathology revealed a lobulated grey-white solid mass with areas of cystic changes that was well-capsulated and measured approximately 10 × 7 × 5 cm. The microscopic findings showed that the tumour was composed of a mixture of predominantly mature as well as embryonal tissue with ectodermal, mesodermal and endodermal components. The immature tissue component was the neuroepithelium, which had areas of necrosis without any evidence of mitosis. The capsular margin was intact without any vascular invasion. The final diagnosis was an immature teratoma (Grade II) with no mitosis, lymphovascular or capsular invasion.

Discussion

Teratomas are classified according to their cellular differentiation as mature, immature or malignant. Mature teratomas usually contain well-differentiated tissues from the three germ cell layers: ectoderm, mesoderm, and endoderm. Immature teratomas also contain tissues from all the germ cell layers, but immature tissues, primarily neuroepithelial tissues, are present. Teratomas can also be graded from 0 to 3 based on the amount of immature tissue found in the tumour specimen. All teratomas that contain malignant foci and those immature teratomas with metastasis are considered malignant.[1]

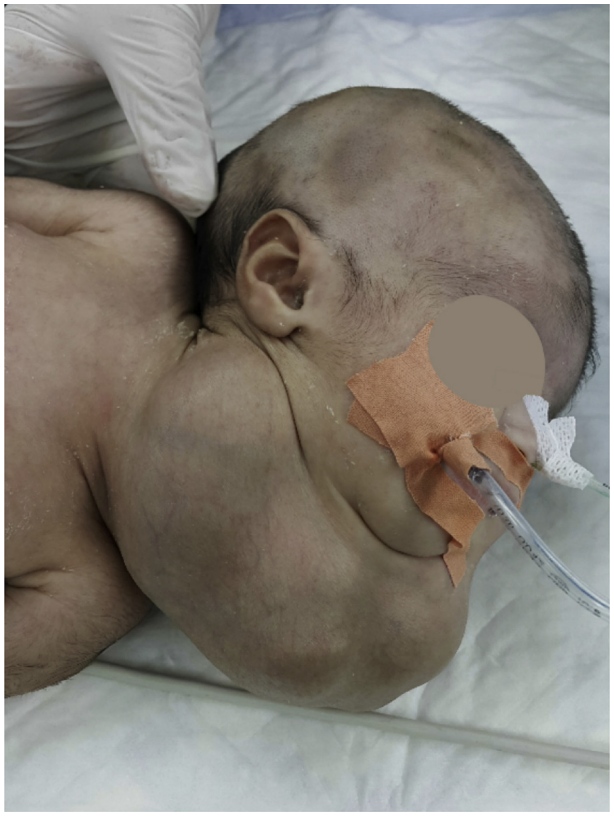

Figure 1: Left huge, lobulated and soft cervical mass with visible dilated vessels.

The main differential diagnoses for foetal neck masses include lymphangiomas or cystic hygromas, cervical teratomas, haemangiomas, branchial cysts, cervical neuroblastomas, soft tissue sarcomas, and congenital cervical thyroid goitres. One differentiating feature of foetal neck masses is on their location, as teratomas are frequently anteriorly located and along the midline, whereas lymphangiomas or cystic hygromas, haemangiomas and branchial cleft cysts are more posterior and lateral in location.[2–4]

Large congenital cervical teratomas can cause hyperextension of the neck and reduced amniotic fluid swallowing leading to polyhydramnios, which is seen in one-third of the cases, along with subsequent pulmonary hypoplasia and hypoventilation.[2,5,6]

Medical imaging is the cornerstone of early detection of congenital cervical teratomas, mainly as a prenatal diagnosis using ultrasound or MRI, and plays an important role minimizing the risk of complications. If the diagnosis is still not definitive, additional radiologic investigations can also be performed postnatally, with the exception of a CT scan, which is rarely indicated due to the hazards of radiation in a newborn.

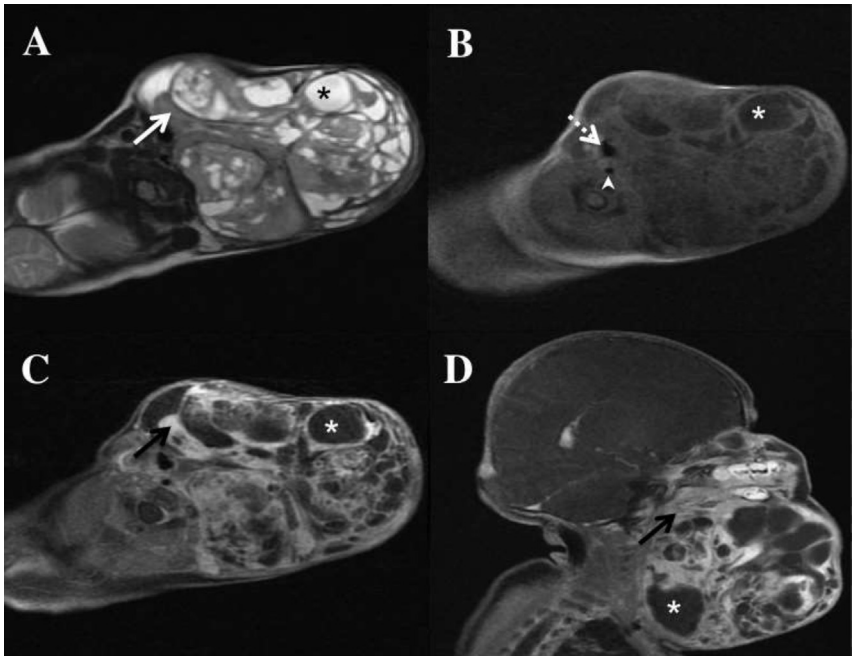

Figure 2: Axial T2 (A), axial T1 pre (B) and post contrast (C), and sagittal T1 post contrast (D) MRI images demonstrate large well-defined multi-loculated complex neck mass with significant mass effect. The solid component (arrows) shows intermediate T2 and T1 signal intensity with avid heterogeneous enhancement while the cystic component (asterisks) shows high T2 and low T1 signal intensity with no enhancement. Oesophagus (arrowhead) and airway (dashed arrow) are displaced.

In our case, we will discuss the radiological appearance of cervical teratomas and cystic hygromas, and emphasize the important differentiating imaging features between these two diagnoses on US, CT and MRI.

Teratomas usually arise along the midline of the body, typically at the sacrococcygeal region. However, congenital teratomas in the cervical region are rare, accounting for 3% of all paediatric teratomas. They appear as single or multiple large bulky masses consisting of cystic and solid components. Calcification is present in 50% of cases. Airway compromise secondary to tumour mass effect is an important prognostic indicator.[5,6]

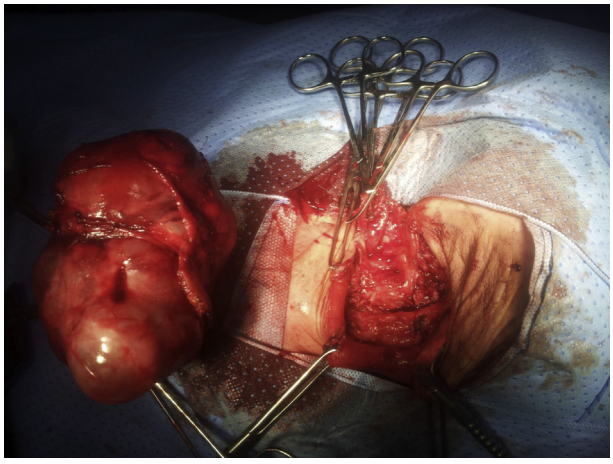

Figure 3: Picture taken during the surgical procedure shows the typical appearance of the lobular soft tissue mass. Surgical management must be as complete as possible to avoid recurrence and malignant transformation.

Cystic hygromas are cystic lesions that are developmental abnormalities of the lymphoid system. These lesions can be single or multiple, and typically appear as multi-septated cystic lesions and rarely have any solid components. They can be complicated by lymphoedema and can rapidly increase in size due to trauma, infection or haemorrhage. Their associations with chromosomal abnormalities such as trisomy 21 and Turner syndrome are responsible factors for poor prognosis.[5,6]

On ultrasound, cervical teratomas appear as large multiloculated, multiseptated mass lesions with cystic and solid components and scattered areas of calcification. By contrast, the sonographic features of cystic hygromas depend on the gestational age of the patient. The features of a cystic hygroma can range from mild oedema in the posterior cervical region in the first trimester, to a large multiloculated predominantly cystic mass with septations of variable thickness later in gestational age. Occasionally, an echogenic area may be identified within the lesion due to clusters of small abnormal lymphatic channels. Fluid–fluid areas can also have a dependent and layered echogenicity if there is a haemorrhagic component.[3,7–9]

On computed tomography, teratomas appear as multiloculated cystic lesions with focal areas of fat attenuation and other areas of calcification. Cystic hygromas are suspected on CT if the lesion is a poorly circumscribed, multiloculated mass with homogeneous fluid attenuation. Higher intra-lesional attenuation is appreciated when secondary infection is present. It is not uncommon for cystic hygromas to extend through fascial planes from one location in the neck into another due to their infiltrative nature.[2,9,10]

It is worth pointing out that MRI is superior to other imaging modalities in delineating soft tissue contrast resolution, in elucidating the relationship of lesions to the surrounding structures, and in facilitating visualization of the internal anatomy of lesions. In addition, MRI provides multiplanar imaging with a large field of view, enabling better examination of foetuses with large or complex masses. However, foetal MRI examination may yield inadequate diagnostic data in the early gestational stages due to foetal movement and the small size of the foetus.[11]

Cervical teratomas are suspected on MRI when a multiloculated multiseptated complex lesion with solid soft tissue and cystic components is identified. Focal areas of high signal intensity on T1- and T2-weighted images that have low signal intensity in fat saturation sequences represent the fatty component of these lesions. When the mass is large, it usually causes contralateral displacement of the surrounding structures, including the airway and carotid sheath. Cystic hygromas usually appear as an infiltrative single or multicystic mass lesion with low or intermediate T1 and high T2 signal intensity. Sometimes, these lesions may demonstrate high T1 signal intensity due to clotted blood or chylous contents. Fluid–fluid levels can be appreciated if haemorrhage is present.[2,9,10]

In our case, the MRI clearly showed the margins of the mass which helped with surgical planning for successful removal. The appearance of a multicystic mass lesion with enhancing solid components and less invasive margins in a newborn was consistent with a diagnosis of congenital cervical teratoma. However, the large amount of enhancing soft tissue and the absence of any fatty component were concerning for possible malignant transformation and therefore, post-surgical histopathologic evaluation was required.

Cervical teratomas are usually treated with prompt complete surgical excision, which is effective and associated with good long-term outcomes. Procedures such as ex utero intrapartum treatment (EXIT) and operation on placental support (OOPS) are used for lesions that have been diagnosed during the prenatal period to prevent airway obstruction at birth and to increase the chances of postnatal survival. Close follow-up is necessary to avoid possible postoperative respiratory distress.[12–16]

Conclusion

Various imaging modalities play a significant role in the early diagnosis of congenital cervical teratomas. The unique imaging characteristics of congenital cervical teratomas help to discriminate these lesions from other congenital neck lesions and consequently allow early surgical excision, ultimately decreasing the risk of complications and improving long-term outcomes.

Authors' contributions

SA wrote the introduction and discussion. AY collected the data from the patient file and lab results and wrote the abstract. AA chose the radiology pictures and wrote their description. FDA is the surgeon who performed the operation and wrote the surgical findings. MA provided the references and organized the article and editing.

Conflict of interest

The authors have no conflict of interest to declare.

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