CASE REPORTS

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Solid ectopic cervical thymus in an infant

Ektopični cervikalni timus čvrste strukture kod odojčeta

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Abstract

Introduction. Ectopic cervical thymus (ECT) occurs as a result of incomplete migration of the thymic primordia during embryogenesis. In the majority of cases, ECT is asymptomatic; however, in 10% of patients, there are different kinds of symptoms. **Case report.** A four-month-old baby boy was referred to our clinic for an evaluation of a growing large mass on the right side of the neck, present since birth. Physical examination revealed a solid, painless, soft, moderately mobile mass of irregular round shape localized on the right side of the neck, in front of the sternocleidomastoid muscle, below the parotid gland, and above the carotid lodge. The skin above the mass was unchanged. The dimensions of the mass were 40 × 32 × 15 mm. Based on the clinical and ultrasonographic findings, as well as the findings of the magnetic resonance imaging, it was suspected that the mass was an ECT. The mass was removed by surgical excision. The pathohistology report confirmed the presence of an ECT, with Hassall's corpuscles in the medulla. The postoperative course went smoothly, and the wound healed well. During the regular clinical, immunological, and ultrasound follow-ups over a period of six months, normal findings were registered. **Conclusion.** Congenital ECT is a rare congenital anomaly that must be, however, taken into account when considering the differential diagnosis of cervical tumor masses.

Key words:
congenital abnormalities; diagnosis, differential; infant; surgical procedures, operative; thymus gland.

Apstrakt

Uvod. Ektopični cervikalni timus (ECT) nastaje kao posledica nepotpune migracije primordijuma timusa tokom embriogeneze. U najvećem broju slučajeva ECT je asimptomatski, međutim kod oko 10% bolesnika postoje različite vrste simptoma. **Prikaz bolesnika.** Četvoromesečni dečak je hospitalizovan u našoj ustanovi zbog prisustva velike izrasline na desnoj strani vrata, prisutne od rođenja, sa tendencijom rasta. Fizikalnim pregledom konstatovana je čvrsta, bezbolna, meka, delimično pokretna masa nepravilnog kružnog oblika na desnoj strani vrata, ispred prednje ivice sternokleidomastoidnog mišića, iza parotidne žlezde i iznad karotidne lože. Koža iznad izrasline bila je nepromenjena. Dimenzije mase bile su 40 × 32 × 15 mm. Na osnovu kliničkog i ultrasonografskog nalaza, kao i nalaza magnetne rezonance, postavljena je sumnja da se radi o tkivu ECT. Promena je odstranjena hirurškim putem. Patohistološkim pregledom potvrđeno je prisustvo ECT, sa Hasalovim korpuskulima u meduli. Postoperativni tok protekao je uredno i rana je dobro zarasla. Na redovnim kliničkim, imunološkim i ultrazvučnim kontrolama u periodu od šest meseci registrovan je uredan nalaz. **Zaključak.** Kongenitalni ECT je retka kongenitalna anomalija koja se, međutim, mora uzeti u obzir prilikom razmatranja diferencijalne dijagnoze cervikalnih tumorskih masa.

Ključne reči:
anomalije; dijagnoza, diferencijalna; odojče; hirurgija, operativne procedure; timus.

Introduction

Ectopic cervical thymus (ECT) is an uncommon cause of neck masses [1, 2]. It results from incomplete migration of the thymic primordia during embryogenesis [3–10]. Most ECT are cystic but they can also be solid (10%) [7, 11]. Symptoms depend on the localization and size of the ectopic tissue [2, 7]. It can be asymptomatic in most (80–90%) cases; however, in a

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small number of patients, there are symptoms such as pain or upper respiratory tract infection, or due to compression symptoms like stridor, dyspnea, dysphagia, and hoarseness [1, 2, 6–9, 11, 12].

The diagnosis of ECT is rarely made before the surgical treatment due to its asymptomatic nature [1, 6, 7]. It is essential to confirm the presence of the normal mediastinal thymus (if total thymectomy is performed, immunodeficiency could occur) [7–10]. Physical examination, ultrasonography (US), computed tomography, and magnetic resonance imaging (MRI) are diagnostic tools that are sufficient to establish an accurate diagnosis in the majority of cases [1, 2, 8, 9–16]. Fine needle aspiration biopsy can be an effective procedure to establish the diagnosis of ECT, and in the case of cystic ECT, this procedure can also be therapeutic [7, 10, 12, 15, 17, 18].

There are a number of neck lesions (congenital or acquired) that occur during childhood [1, 19]. The differential diagnosis includes thyroglossal duct cyst, branchial anomalies cysts, lymphadenopathy, vascular anomalies (infantile hemangioma, lymphatic malformation, venous malformation), inflammatory lesions, benign or malignant tumors, etc. [1–3, 13].

There are opponent opinions about the treatment [1–3, 8, 10]. Surgical excision is recommended based on the risk of severe clinical symptoms or malignant transformation [1, 4, 6–9, 11, 13, 16, 17]. On the other hand, some authors suggest observation with an MRI performed every six months to one year expecting involution (because of high morbidity associated with surgical excision) [10, 15, 17, 19]. Small and asymptomatic ECT requires observation (watch and wait policy); however, ECT causing compressive symptoms demands surgical management [3, 8, 10, 15].

To the best of our knowledge, 150 cases of ECT with sufficient documentation were reported in the pediatric population during the last 20 years (143 patients, seven cases with bilateral localization) [7, 18]. In ten cases, the definitive diagnosis was obtained after the autopsy, and in 106 cases, the surgical excision was performed (the final diagnosis was obtained after pathohistological analysis), while the rest were conservatively managed (observed). There were only 28 cases of surgically excised solid ECT during these 20 years, with the average age of the patients being 3.5 months [18].

Here, we present a case report of a four-month-old baby boy with ECT on the right side of his neck who underwent surgical excision.

Case report

A four-month-old baby boy was referred to the Clinic for Pediatric Surgery at the Institute for Children's Disease in Montenegro for the evaluation of a large mass on the right side of the neck. The mass has been present since birth. The mass was asymptomatic, but it was growing over time. The family history and the previous medical history were unremarkable. Physical examination revealed a painless, soft, moderately mobile, and round solid mass localized on the right side of the neck, in front of the sternocleidomastoid muscle, below the parotid gland, and above the carotid lodge. The mass was 40 × 32 × 15 mm large in diameter. No overlying skin changes or pits were apparent; the patient's neck position and mobility were not limited by the presence of the mass. US and MRI of the neck were performed (Figures 1 and 2). Following the clinical and MRI findings, the diagnosis of an ECT was highly suspected. The dimensions of a normal retrosternal thymus were 64 × 26 × 24 mm. After adequate preoperative preparation, surgical excision was done. The blunt dissection technique was performed, respecting the integrity of the pharyngeal, laryngeal, tracheal wall, and the surrounding neurovascular structures (Figure 3). The final pathohistology report confirmed the presence of an ECT tissue with Hassall's corpuscles in the medulla (Figure 4). The patient's postoperative course was unremarkable, without neurological defects or injuries. The wound healed primarily. Furthermore, clinical, immunological, and US follow-ups over a period of six months were unremarkable (Figure 5).

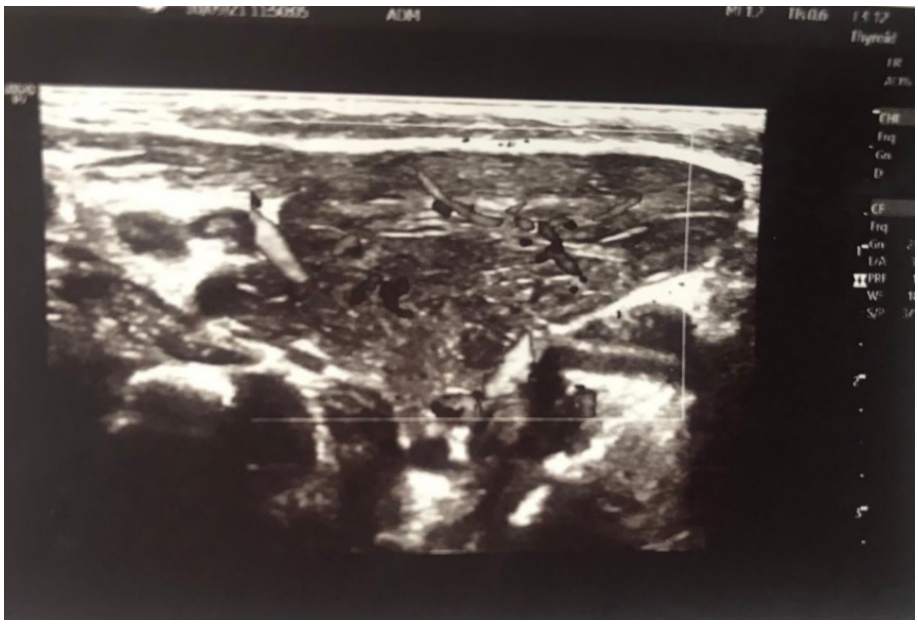

Fig. 1 – Ultrasound revealed a homogenous solid mass on the neck.

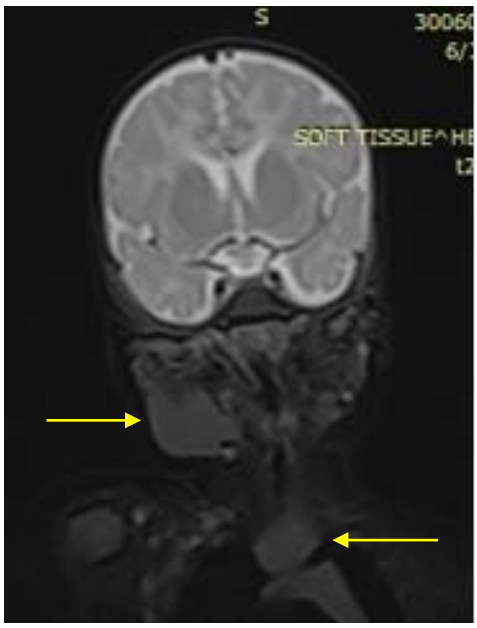

Fig. 2 – Magnetic resonance imaging of the neck. In the right parotid region of the neck (upper arrow), a homogeneous, clearly demarcated change is differentiated, which in all base sequences shows a signal that corresponds to the signal of the thymus (lower arrow).

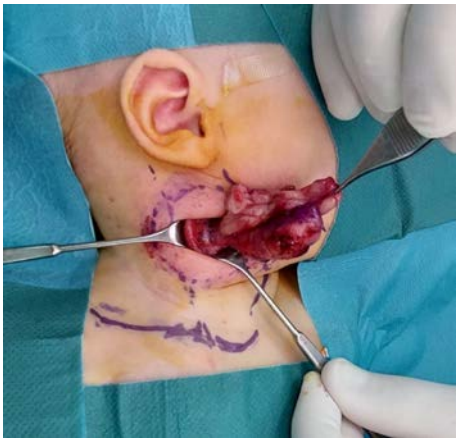

Fig. 3 – Intraoperative view of ectopic cervical thymus.

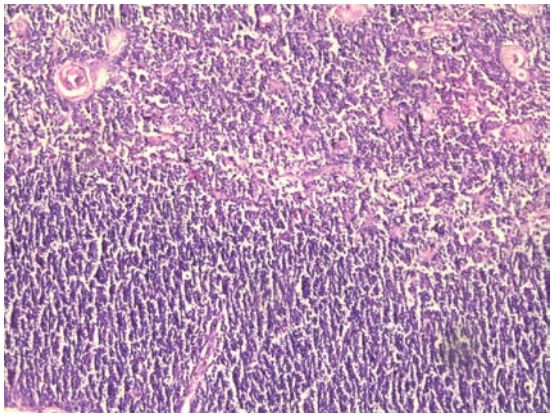

Fig. 4 – Pathohistology revealed ectopic cervical thymic tissue with Hassal's corpuscles in the medulla (hematoxylin and eosin, ×10).

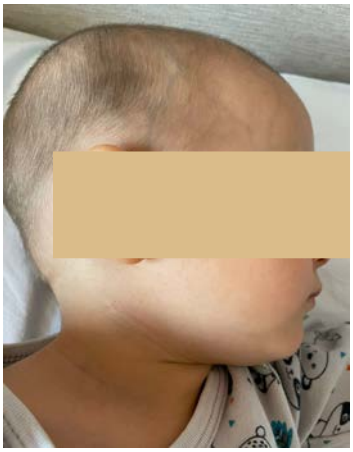

Fig. 5 – Patient six months after surgical treatment.

Discussion

The thymus is a paired organ developed from the third and occasionally fourth pharyngeal pouches during the sixth week of fetal life [2, 4, 7]. By the third month of development, the thymus matures to have a cortex and a medulla with Hassall's corpuscles [2]. Some authors suggest that ECT develops from the persistence of the thymopharyngeal duct or the degeneration of Hassall's corpuscles [1, 2, 6]. Due to its embryology, the ectopic thymus (ET) tissue can be found anywhere along the thymic pathway of descent [1, 2, 5]. For unknown reasons, according to the literature, ECT is more common on the left side (60–70% of total cases) [4, 6, 10, 13]. In our report, the ectopic tissue was on the right side of the neck, above the carotid lodge. Other authors have described ET tissue in the pharynx, subglottis, unilaterally or bilaterally in the neck [4, 5, 11].

The thymus exhibits age-related variation in weight and size. It reaches its maximum weight by puberty; after that, it slowly involutes, and the lymphoid component of the thymus is replaced by fat and connective tissue, with persistent Hassall's corpuscles [2, 7]. In our case, the normally located retrosternal thymus was 27 mm (transverse) and 24 mm (anteroposterior) in size. Çolak and Özkan [14] reported in their study that the average transverse diameter for male infants is 34.6 ± 4.9 mm, and the average anteroposterior diameter is 18.4 ± 4.0 mm.

ET exhibits hyperplasia during the first several years of life or following vaccination or infection [1, 7]. In our case, ET was 40 mm in length, while the average length, according to the literature, is 22.5 mm (ranging from 8 mm to 38 mm) [4]. Neck mass of thymic origin can be solid or, more often, cystic or both solid and cystic [2, 7, 9]. Only 10% of all ET masses have a form of the solid ET [7, 8, 11]. In our case, the presence of thymus tissue with clearly differentiated cortical and medullary zones with Hassall's corpuscles was confirmed on pathohistology.

The thymus plays an important immunological role during childhood [1, 11, 13]. Because of its immunological role, proving the presence of normal thymus tissue to avoid the occurrence of autoimmune diseases is vital. In our case, the normal thymus tissue was present, with a standard number of lymphocytes after surgery.

Cervical masses in children, whether benign or malignant, can be present with heterogeneous clinical signs [1, 4, 13]. Following the physical examination, US, and MRI findings, it can be very difficult sometimes to distinguish between benign and malignant lesions [4, 13]. A fast-growing cervical mass necessitates prompt attention with preferential screening for infectious diseases, vascular anomalies or bleeding, and malignancies due to its treatment [2, 6]. Additionally, an important consideration is the limited space in the neck to accommodate a fast-growing lesion that could have significant, sometimes fatal consequences [4, 13]. Dermoid cysts, lymph nodes, or branchial cysts should also be considered [9, 13]. In our case, an asymptomatic baby boy with normal laboratory findings was presented. According to the literature, after obtaining a complete history and physical examination, the US and the MRI have to be performed [1, 3, 4, 6, 9]. Based on the clinical and MRI findings, the lesion was highly suspected for ECT.

There are different opinions about the therapeutic approach. According to some authors, surgical excision is the treatment of choice for ECT for diagnostic and therapeutic reasons and risk of malignancy [1, 4, 6–9, 11, 13, 16, 17]. On the other hand, some authors suggest observation, especially in the case of small and asymptomatic ECT [8]. Due to the theoretical risk of infantile immunocompromising related to thymectomy, we find it reasonable to follow the mass with regular physical and imaging controls in asymptomatic children with a small mass. On the contrary, a large mass that can cause compressive symptoms has to be completely excised [1, 6, 11]. In our case, we performed surgical treatment because of the growth of the lesion, the risk of compressive symptoms, potential neoplastic degeneration, and the need for a definitive diagnosis. The blunt dissection was performed during the surgical procedure with great care to avoid injury to surrounding vital structures. There is no clear data about solid ECT recurrence (2% for cystic) [1, 2, 11]. In our case, the postoperative period was unremarkable, and there was no difference in the total number of lymphocytes or the occurrence of infection during a six-month follow-up.

Conclusion

Congenital ECT is a rare congenital anomaly that should be, however, taken into account when considering the differential diagnosis of neck masses.

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