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MANAGEMENT OF CONGENITAL NECK MALFORMATIONS - BRANCHIAL CYSTS

LEČENJE UROĐENIH ANOMALIJA VRATA - BRANHIJALNE CISTE

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Ključne reči
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Abstract

Objective: Congenital neck malformations result from incomplete obliteration of the first, second, or third branchial clefts and develop in early fetal period. Second branchial cleft anomalies are by far the most common presenting near the mid- to upper border of the sternocleidomastoid (SCM) muscle. They could be present in several forms: cysts, fistulas and sinuses.

Material and methods: This is a retrospective study of 45 patients hospitalized within 2007-2013 at Department for ENT and Maxillofacial Surgery, Clinical Hospital Center „Zemun" Belgrade, Serbia for second branchial abnormalities, medial or lateral branchial cyst. The parameters that were analyzed are age, gender, clinical examination, surgical procedures and post-surgical evolution. Among the anomalies of the second branchial arch, 27 (60%) patients with lateral branchial cyst and 18 (40%) patients with medial branchial cyst were encountered. Neither patients with branchial sinuses nor with fistulas were diagnosed. Histological examination of the lesions after excision confirmed the diagnosis in all the cases.

All patients were submitted to preoperative imaging diagnostics (ultrasound, CT) which have clearly demonstrated existence of the cystic masses in the upper, mid- and lower part of the neck.

All patients were primarily treated surgicaly - cystic formations were extirpated, with hyoid bone resection and ligation in case of medial branchial cysts.

Conclusion: Second branchial arch anomalies are the most common branchial anomalies. Patients with cysts usually present later with a mass (sterile or infected). Therefore, they are diagnosed by the patients itself or during the routine examination by the doctor.

INTRODUCTION

The differential diagnosis of congenital cervical anomalies is important for head and neck masses pathology, in both children and adults. These lesions could be present as palpable cystic masses, infected masses, draining sinuses, or fistulaes. Appropriate diagnosis and management of these lesions requires a profound understanding of their embryology and anatomy. Correct diagnosis, resolution of infectious issues before definitive therapy, and complete surgical excision are essential for recurrence prevention [(1)]. Branchial anomalies comprise approximately 20%, thus being the second most common pediatric congenital head and neck masses, after thyroglossal duct cysts [(2, 3)].

The anatomic location of a branchial cleft abnormality represents the presumed branchial cleft origin. Branchial system anomalies can manifest as a sinus, fistula, or cyst. Branchial cleft sinuses with external openings are usually associated with the first and second branchial cleft arches. Branchial cleft sinuses with internal openings are usually associated with the third and fourth arches.

More than 90% of branchial cleft anomalies arise from the second branchial cleft system. At least 75% of all second branchial cleft abnormalities are cysts [4], which typically manifest at the age between 10 and 40 years. Second branchial cleft fistulas and sinuses are less common and usually present during the first decade of life [5, 6]. No gender predilection has been reported [7]. Approximately 8% of branchial cleft anomalies arise from the first branchial cleft system. Cysts arising from the third and fourth branchial cleft systems are rare.

Several theories of the genesis of branchial cleft anomalies are proposed.

According to the branchial theory the lateral cervical cysts are the result of imperfect obliteration of the branchial clefts, arches, and pouches. Ascherson, in 1832, was the first investigator who described 11 cases of branchial fistulae, equating the development of lateral cervical cysts with that of branchial fistulae due to their location [8]. He suggested that incomplete obliteration of branchial cleft mucosa, which remains dormant until stimulated to grow later in life, result in cyst formation. Since then, a number of investigators re-emphasized this link between the pharyngeal arches and cervical cysts [9-13].

The second theory, suggested by His W. assumed that branchial fistulas are vestiges of the cervical sinus rather than of the pharyngeal clefts or pouches [14]. Parallel to the branchial theory, this precervical sinus theory was extended to include lateral cervical cysts. Wenglowski suggested an entirely different hypothesis based on human embryo and cadaver studies [15]. He showed that pharyngeal cleft tissue was not represented in any adult structure inferior to the hyoid bone. Thus any cyst lying below this level could not be derived from a pharyngeal cleft. Wenglowski went on to describe the development of the thymus from the third pharyngeal pouch via the thymopharyngeal duct. He suggested that incomplete obliteration of the thymopharyngeal duct resulted in a lateral cervical cyst. However, a major problem with the thymopharyngeal duct theory is that more anomalies are to be found at the lower end of the embryonic thymopharyngeal duct, that is, at the lower end of the neck and mediastinum rather than cephalad near the hyoid bone.

There were no findings of genetic origin documented in literature.

By the end of the 4th week of embryonic life, the branchial arches (derived from neural crest cells) and the mesenchyma (derived from the lateral mesoderm) are easily recognizable. Five pairs of ectodermal clefts (grooves) and five endodermal branchial pouches separate the six arches, with a closing membrane located at the interface between the pouches and the clefts (16,17). During the fifth week of fetal development, major head and neck structures are formed. The five pharyngeal arches (bands of tissue) are important structures that are formed. These arches contain primitive connective tissue that becomes cartilage, bone, muscle and blood vessels. Incomplete, failed or persistent embryonic development of these arches results in several anomalies or defects in the neck.

Bailey H. [21] classified second branchial cleft cysts into four types. The Bailey type I cyst is the most superficial and lies along the anterior surface of the sternocleidomastoid muscle, just deep to the platysma muscle. The type II cyst is the most common and found in the „classic" location for these cysts: along the anterior surface of the sternocleidomastoid muscle, lateral to the carotid space, and posterior to the submandibular gland. A type III cyst extends medially between the bifurcation of the internal and external carotid arteries to the lateral pharyngeal wall. The type IV cyst lies in the pharyngeal mucosal space and is lined with columnar epithelium.

Congenital cervical anomalies are important to consider in the differential diagnosis of head and neck masses in children and adults. They are bilateral in about 1% of the cases, without any proclivity of either left or right side to develop at [18].

MATERIAL AND METHODS

This study comprises the epidemio-clinical aspects, the treatment modalities and the evolution of these congenital pathologies. It is a retrospective study of 45 patients treated at the Department for ENT and Maxillofacial Surgery, Clinical Hospital Center of Zemun, Belgrade, Serbia within a period of 6 years (January 2007 to December 2013). The patients had developmental anomalies of the second branchial arch - branchial cysts (there was no patients with branchial fistulas nor sinuses in the studied group). All patients were submitted to clinical examination and preoperative diagnostic imaging (ultrasound, CT) which demonstrated existence of the cystic masses in the upper, mid- or lower part of the neck. Patients age ranged from 16 to 50 years, with mean age 35 ± 6.8, were predominantly males 32 (71%) and 13 women (29%).

The clinical and paraclinical context of the therapeutically decision, the surgical procedures and postoperative evolution as well as the histopathologica examination of the excised pieces were analyzed.

RESULTS

In our study all the patients had branchial cysts, which coincide with literature data that show a greater incidence of branchial cysts compared to branchial sinuses and fistulas. On the other hand, most of the patients (70%) were in the first or second decade of life (literature data show a higher frequency of branchial cysts in adults - second or third decade). Also, there was almost twice as much men than women (32/13) in our study (in the specialty literature there was no gender proclivity). The structure of the studied group was as follows: the ratio rural/urban was 3/1 (35/10); the ratio lateral/medial branchial cyst 1.5/1 (27/18). The rest of the results we obtained are similar to those in the specialty literature: there was no predisposition of a certain side of the neck (right or left).The main complications were: cystic swelling, infectious complication, recurrence of the disease due to the incomplete resection of hyoid bone.

DISCUSSION

Cysts on the lateral side of the neck are relatively common anomalies. Despite the fact that branchial anomalies, such as sinuses and fistulae, are usually diagnosed during infancy, the lateral cervical cysts are identified most com-

monly between the second and fourth decades of life, when they grow bigger because of infection or other causes [19] (Fig. 1). Males and females are equally affected and there is occasionally a hereditary tendency [20, 21] . Cysts, may be bilateral, are slow-growing and have a duration of weeks to many years. The lesion, usually, is presented as an asymptomatic circumscribed movable mass, usually close to the anterior border of the sternocleidomastoid muscle. Depending on the size and the anatomical extension of the mass, local symptoms, such as dysphagia, dysphonia, dyspnea, and stridor, may occur. Cysts may vary in size and may fluctuate. Infected cysts may develop into abscess, especially during periods of upper respiratory tract infections, due to the lymphoid tissue located beneath the epithelium. Spontaneous rupture of an abscessed branchial cleft cyst may occur, resulting in a purulent draining sinus to the skin or the pharynx. Larger cysts may displace the sternocleidomastoid muscle posterolaterally, and the carotid and internal jugular vein medially. With any further increase in size of the cyst, the deep cervical fascia prevents its expansion between the sternocleidomastoid muscle and the strap muscles of the larynx. As a result, the cyst takes the path of least resistance and extends posterior and medial to the sternocleidomastoid muscule. The preoperative diagnosis is mainly based on clinical and radiological criteria, which also define the topographic relationship. The precise location and course of the cervical cysts depend on the particular branchial pouch or cleft from which they are derived. Most frequently, these cysts are identified along the anterior border of the upper third of the sternocleidomastoid muscle. However, these cysts may develop anywhere along the course of a second branchial fistula, extending from the skin to the lateral neck, between the internal and external carotid arteries, and into the palatine tonsil. Therefore, the second branchial cleft cyst is in the differential of a parapharyngeal mass.

Complete surgical resection, through a wide, transverse cervicotomy under general anaesthesia is the treatment of choice and results in a good prognosis (Fig. 2). Identification, during operation, of the internal and external carotid arteries and the vagus, hypoglossal, glossopharyngeal and superior laryngeal nerves will avoid injury of these structures. Complications of surgical treatment include recurrence, formation of a persistent fistula, and damage to the cranial nerves [22]. Patients with infected cysts receive a full course of antibiotics before surgery to decrease the risk of recurrence and persistent fistula. Alternative treatments, such as percutaneous sclerotherapy, remain unproven [23]. Most branchial cleft cysts are lined with stratified squamous epithelium and contain a brownish fluid with crystals of cholesterol at histopathologic examination[24]. Occasionally, the cyst is lined with respiratory (ciliated columnar) epithelium. The wall may contain lymphoid tissue arranged in a follicular pattern. In infected or ruptured lesions, inflammatory cells are seen within the cyst cavity or the surrounding stroma.

The diagnosis of a branchial cleft anomaly should be considered whenever a child presents with a neck mass, recurrent neck abscesses, or suppurative thyroiditis. The usual workup for a branchial cleft anomaly includes either magnetic resonance imaging or computed tomography to determine if a tract exists.

A problem during cyst preparation may appear since they often adhere to the carotid artery and the internal jugular vein, and bleeding can be a problem that requires careful dissection. Very often cyst may adhere to the hypoglossal nerve, the posterior belly of the digastric muscle, and the stylohoid muscles. Another problem can be proper hemostasis of smaller vessels that supply the cyst wall that could be solved using endoscope and a sophisticated ultrasonically activated scalpel as presented by Matsui et al.[25]. Unfortunately, this was not done with this sophisticated equipment, because our department doesn't posses such one.

As we presented all of our 45 patients are successfully cured and healed, performing mentioned precise surgical procedures. Therefore it can be concluded that: the only appropriate treatment of congenital cervical cysts is surgery providing that the resection is meticulous with complete resection in order to avoid relapse. Complete resection also suppresses the risk of secondary malignant degeneration of amygdaloidal and thyroglossal duct cysts.

Lateral cysts of the neck containing malignant epithelium represent a diagnostic and therapeutic challenge. Much attention in the literature has been focused on the frequent relationship between a solitary cystic cervical metastasis and an occult primary tumour in the tonsil or the tongue base. It has been suggested that metastatic deposits from primaries in these sites have a particular tendency to undergo cystic transformation. Despite rigorous investigation however a high proportion of primary sites remain occult(26). Branchial cleft cyst carcinoma is extremely rare, compared to the far more frequent cystic metastases arising from primary malignancies.

CONCLUSIONS

Branchial cysts are frequently incorrectly diagnosed and forgotten in the differential diagnosis. Branchial cyst should be suspected in any patient with a swelling in the lateral part of the neck, painful or painless. Second branchial cysts have variable sonographic appearances, which may confuse the unexperienced doctors. The CT and MRI are very useful in demonstrating the cystic nature and the anatomical extensions of branchial anomalies (Fig. 3). The histopathological examination establishes definitive diagnosis.

The treatment of branchial anomalies begins with a careful and complete history and physical examination searching for associated systemic anomalies such as craniofacial or branchio-oto-renal syndrome. The definite treatment for branchial anomalies is a complete surgical excision. A resection of a hyoid bone is appropriate treatment in curing medial cyst of the neck. Because of the high incidence of secondary infection of these anomalies, an early excision is recommended. Patients who have undergone acute inflammatory episodes should await subsidence following antibiotic therapy, with incision and drainage of any abscess if indicated. General anesthesia is usually recommended because of the complexity of the dissection.

Properly performed surgical treatment would not allow a recurrence of the branchial anomaly.

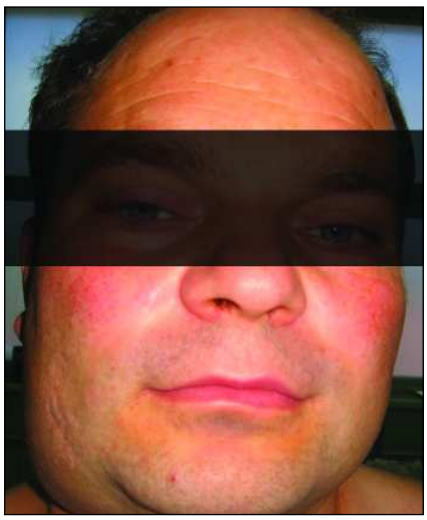

Figure 1

The 36-years-old man was admitted to our department with an asymptomatic mass, located at the right side of the neck. He was referred to our department for diagnostics and treatment of a slowly enlarging mass at the right side of the neck. The swelling appeared 3 years before and progressed gradually to its current size. The examination revealed an asymptomatic, movable mass at the anterior border of the sternocleidomastoid muscle, between its upper and middle third. The CT scan revealed a cystic lesion. Based on these clinical and radiological data, the lateral cervical cyst was preoperatively diagnosed.

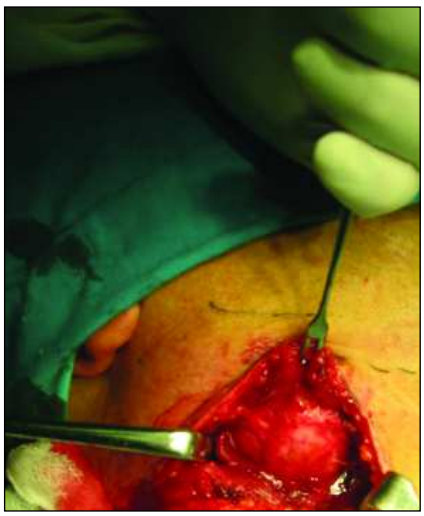

Figure 2

At the operation a cystic lesion was found under the platysma, anterior to the sternocleidomastoid muscle, in contact with the internal jugular vein.

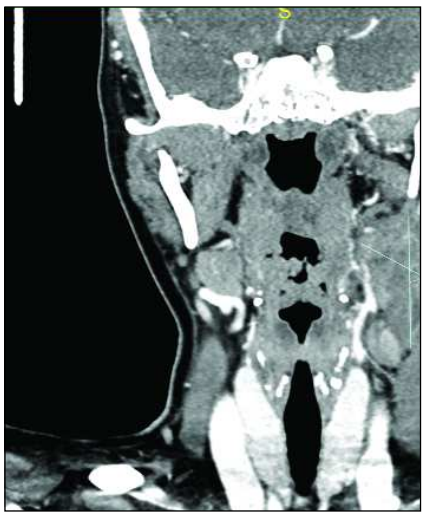

Figure 3

The 26-year-old woman was sent to our department for for diagnostics and treatment of a slowly enlarging mass on the left side of the neck. The swelling which appeared 2 years before progressed gradually to its actual size. The examination revealed a mobile, non-tender mass, located close to the anterior border of the sternocleidomastoid muscle, and the CT-scan demonstrated a cystic lesion. Physical examination showed a movable, painless mass, located close to the anterior border of the sternocleidomastoid muscle. The lesion was excised through a wide, oblique cervicotomy. After incision of a skin an anterior border of sternocleidomastoid was identified, prepared and retracted, cyst usually spontaneously appeared and using blunt preparation cyst was dissected from surrounding tissue. A complete cyst was extirpated, and this was a final maneuver in its removal. And finally after closure by layers, direct intradermal suture has been done. Histopathologic evaluation documented a second branchial cleft cyst. The patient had an uneventful postoperative course.

Sažetak

Uvod: Kongenitalne anomalije vrata najčešće su posledica nepotpunog razvoja prvog, drugog ili trećeg škržnog luka tokom embrionalnog razvoja fetusa. Anomalije drugog škržnog luka se najčešće razvijaju u predelu srednje i gornje trećine sternokleidomastoidnog mišića. One se javljaju u nekoliko različitih formi: kao ciste, sinusi i fistule

Materijal i metode: U retrospektivnoj studiji koja je sprovedena na odeljenju za ORL i maksilofacijalnu hirurgiju, Kliničko-bolničkog centra "Zemun"u Beogradu, Srbija, ukupno je lečeno 45 bolesnika sa dijagnozom medijalna ili lateralna cista vrata u periodu od 2007-2013. godine, u relaciji sa godinama, polom, kliničkim pregledom, hirurškom procedurom i posthirurškim praćenjem. Kod 27 bolesnika (60%) je dijagnostikovana lateralna branhijalna cista, a kod 18 (40%) medijalna branhijalna cista vrata. Pacijenti sa branhijalnim sinusima ili fistulama nisu dijagnostikovani.

Svim našim bolesnicima je sprovedena preoperativna CT i Eho dijagnostika koje su ukazale na prisustvo cističnih masa u gornjim, srednjim i donjim delovima vrata.

Takodje, svi bolesnici su podvrgnuti hirurškom lečenju gde je cistična lezija u potpunosti ekstirpirana, poslata na histopatološku analizu, a u nekim slučajevima (medijalne ciste vrata) resektovana je hioidna kost a njeni fragmenti ligirani.

Zaključak: Anomalije drugog skržnog luka ubrajaju se u najčešće branhijalne anomalije. Potrebna je adekvatna dijagnostika, individualno planiranje operativnog zahvata kao i potpuno uklanjanje promene i njene histopatološke analize, kako ne bi došlo do recidivaranja. U tu svrhu, potrebna je i resekcija hioidne kosti kod medijalnih branhijalnih cista, kao i podvezivanje njenih fragmenata a u cilju izbegavanja komplikacija-disfagija.

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