

ORIGINAL PAPER

Management of second branchial cleft anomalies

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Abstract

Branchial cleft anomalies are developmental disorders of the neck. The aim of this study was to evaluate the data of our patients, who have been diagnosed and treated for second branchial abnormalities in the last six years. We report our clinical experience in second branchial anomalies with a review of the literature. Our study is a retrospective one on a number of 23 patients hospitalized within 2001–2007 in ENT Clinic of Craiova for second branchial abnormalities in relation with age, gender, origin environment, clinical and paraclinical context in which the therapeutic decision was made, surgical procedures, post-surgical evolution. Among the anomalies of the second branchial arch, we encountered 10 (43.47%) patients with branchial cyst and 13 (56.52%) patients with branchial sinus. Twelve (52.17%) of the 23 patients were women and 11 (47.83%) were men; 9 (39.13%) patients were diagnosed and treated within the first age decade, seven (30.43%) within the second age decade, five (39.13%) within the third age decade and two (8.71%) in the fourth age decade. Histological examination of the lesions after excision established the diagnosis in all the cases. Second branchial arches anomalies are the most common branchial anomalies. Sinuses are more frequently than cysts and branchial fistulae are extremely rare. There is no gender predilection. The majority of patients (approximately 70%) were diagnosed and treated during their childhood. Treatment for these lesions is complete surgical excision for prevent recurrences.

Keywords: second branchial cleft, branchial fistulae, branchial cysts, branchial sinus, abnormalities.

Introduction

The term branchial cyst was first used by Ascherson in 1832. He suggested that these cysts were results of impaired obliteration of branchial clefts [1].

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 [2, 3].

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.

Congenital cervical anomalies are important to consider in the differential diagnosis of head and neck masses in children and adults. These lesions can present as palpable cystic masses, infected masses or fistulae [4].

Branchial cleft cysts and fistulae represent about

20% of cervical masses in children [5]. They are bilateral in about 1% of the cases, without any proclivity to one side (right or left) in which they develop [6]. They are located in the cervical region, parotid and mediastinum.

The branchial cysts were classified into four types:

▪ First branchial cleft cysts are divided into type I and type II. Type I cysts are located near the external auditory canal. Most commonly, they are inferior and posterior to the tragus (base of the ear), but they may also be in the parotid gland or at the angle of the mandible. They may be difficult to distinguish from a solid parotid mass on clinical examination. Type II cysts are associated with the submandibular gland or found in the anterior triangle of the neck.

▪ The second branchial cleft accounts for 95% of branchial anomalies [7–9]. Most frequently, these cysts are identified along the anterior border of the upper third of the sternocleidomastoid muscle, adjacent to the muscle. However, these cysts may present anywhere along the course of a second branchial fistula, which proceeds from the skin of the lateral neck, between the internal and external carotid arteries, and into the palatine tonsil [10]. Therefore, second branchial cleft cyst is in the differential of a parapharyngeal mass.

▪ The third and the fourth branchial cleft cyst are rare.

☐ Material and methods

The study material consisted of the patients' observation files, surgical reports and histopathologic examinations.

The present study is a retrospective one, on a number of 23 patients hospitalized in ENT Clinic of The Clinical County Hospital of Emergency Craiova within a period of six years (April 2001–April 2007).

The patients were suffering from development anomalies of the second branchial arch: branchial cysts and sinuses (there was no patient with branchial fistulae in the studied group).

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

☐ Results

The structure of the studied group was as follows (Figure 1): the ratio men/women was of approximately 1/1 (11/12); the ratio rural/urban = 2/1 (15/8); the ratio branchial cysts/branchial sinuses/branchial fistulae = 10/13/0.

Case no. 1

The patient S.F., 26-years-old, rural environment, was hospitalized in ENT Clinic Craiova (Observation File no. 57795) within 18.11.2004–25.11.2004 for a distortion of the right lateral-cervical region.

At local examination there was found a tumoral formation of 10/7 cm in the right lateral-cervical region. The tumor was oval, soft, fluctuant, painless spontaneously and while palpated, mobile in profound planes and unadherent to tegument (Figure 2). The cervical echography depicted the presence of a cystic formation in the right lateral-cervical region adherent to the right submandibular gland.

The diagnosis was of cystic right lateral-cervical formation and cervicotomy practiced with extirpation of the tumoral formation (C.O. 1578/18.11.2004), (Figure 3, a–c). The excised piece (Figure 4, a and b) was sent for histopathological examination and diagnosed as branchial cyst (BHP 264835/19.09.2004, Figures 5–7).

The anatomo-pathological exam was surprising – clinical and echographic diagnoses were of cyst of submandibular gland. The shown images are suggestive for the diagnosis of infected branchial cyst:

- Figure 5 – parotid gland with alterations of unspecified chronic parotiditis; the peripheral part of a branchial cyst can be noticed nearby (HE staining, ob ×40);

- Figure 6 – infected branchial cyst (internal part): lymphoid tissue, walled by stratified squamous epithelium, in exuded leucocitary lumen (HE staining, ob ×40);

- Figure 7 – infected branchial cyst, with follicles formation in lymphoid tissue (HE staining, ob ×40).

Histopathologically, the cyst was formed from the exterior to interior, from a fibro-collagen wall, lymphoid tissue with follicle structures and a stratified

squamous epithelium, allowing the diagnosis of branchial cyst. There was abundant leucocitary exudate in the cyst lumen, and collagen sclerosis zones in the lymphoid tissue, suggesting the lesion age and overinfection. We noticed the parotid gland with alterations of unspecified chronic parotiditis adjacent to the cyst.

Case no. 2

The patient C.A.L., 18-years-old, rural environment, was hospitalized in ENT Clinic Craiova (Observation File no. 1539) within 10.09.2001–20.09.2001 as she presented a tegumentary orifice latero-cervical and seromucous secretions that drain through it.

At local examination, we could see a fistulous orifice in the inferior third of right cervical region, with mucous secretions. The fistulous tract was imagistically depicted by fistulography (Figure 8, a and b). The diagnosis was of right branchial sinus.

The patient was operated on (C.O. 1097/18.09.2001), the fistulous tract was excised (Figure 9) and the piece sent to histopathological examination where stratified squamous epithelium, subjacent fibrosis and chronic inflammation were discovered (BHP 225398/19.09.2001, Figures 10 and 11).

Histopathologically, the branchial sinus was made up from a fibro-collagen wall, lymphoid tissue with follicle structures and a stratified squamous epithelium. We noticed the presence of collagen sclerosis zones adjacent to the squamous walled epithelium, suggesting an old lesion:

- Figure 10 – branchial sinus (internal part): lymphoid tissue with lymphoid follicles formation walled with stratified squamous epithelium, adjacent collagen fibrosis (HE staining, ob ×40);

- Figure 11 – branchial sinus (detail): stratified squamous epithelium, adjacent lymphoid tissue and collagen fibrosis (HE staining, ob ×40).

☐ Discussions

Congenital abnormalities of the branchial apparatus can result in various abnormal conditions in the neck, including a cyst (a small, partial malformation with no external opening), a sinus (a malformation with one superficial opening) or a fistula (a complete malformation with both internal and external communications).

Bailey H [11] 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.

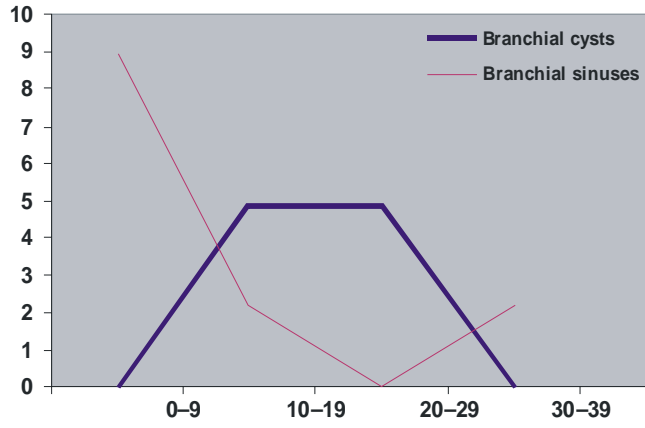
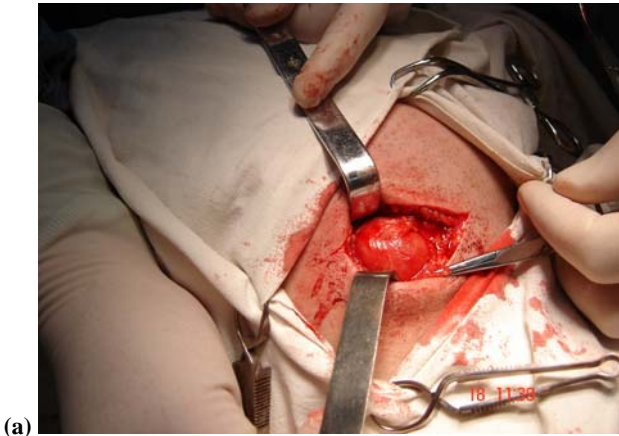


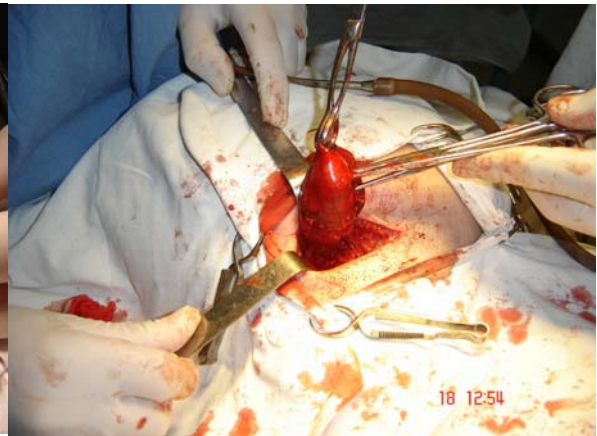
Figure 1 – Patients’ distribution on age decades



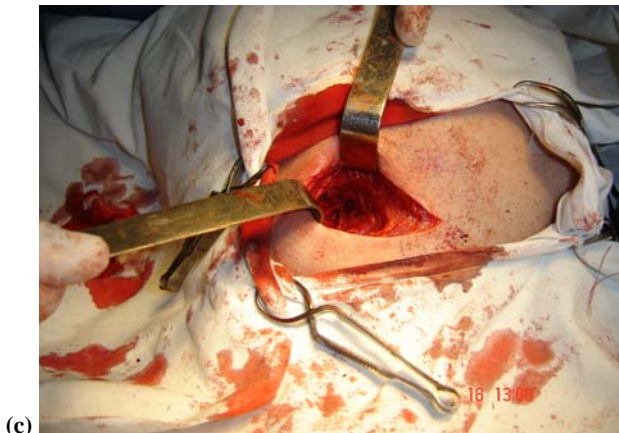
Figure 2 – The aspect of tumor at inspection



(a)



(b)



(c)

Figure 3 – (a-c) Intra-surgical aspects



(a)



(b)

Figure 4 – (a-b) Excised piece

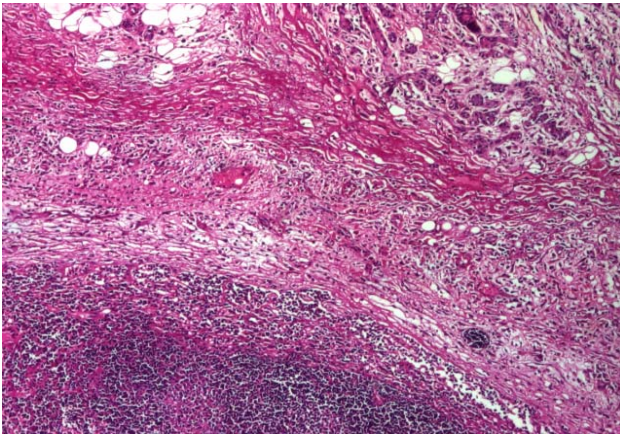


Figure 5 – Parotid gland with alterations of unspecified chronic parotiditis; the peripheral part of a branchial cyst can be noticed nearby (HE staining, ob $\times 40$)

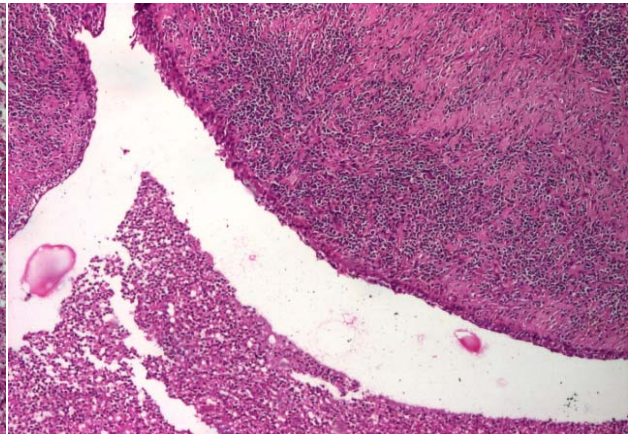


Figure 6 – Infected branchial cyst: lymphoid tissue, walled by stratified squamous epithelium, in exuded leucocitary lumen (HE staining, ob $\times 40$)

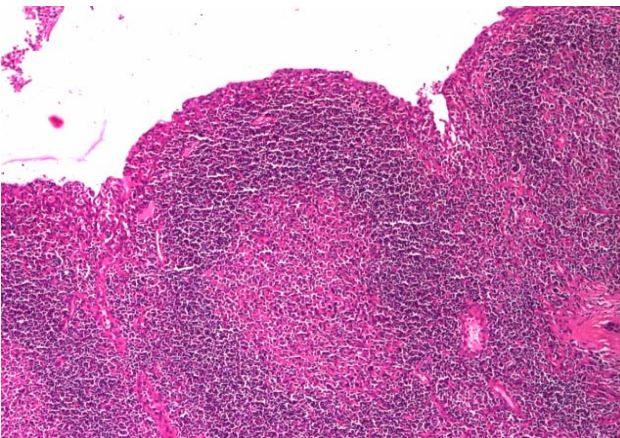


Figure 7 – Infected branchial cyst, with follicles formation in lymphoid tissue (HE staining, ob $\times 40$)



(a)



(b)

Figure 8 – Radiological aspect: (a) frontal and (b) profile



Figure 9 – Excised piece

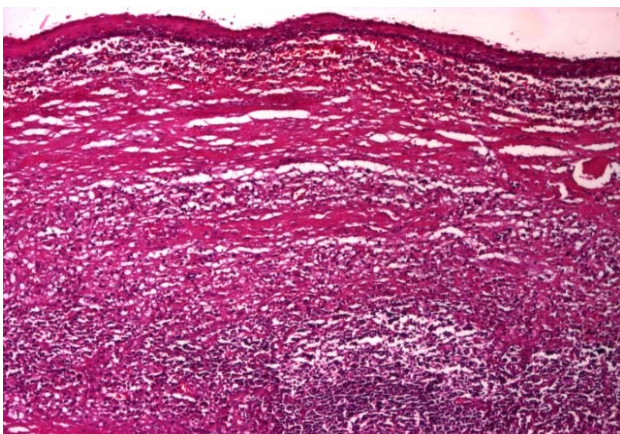


Figure 10 – Branchial sinus (internal part): lymphoid tissue with lymphoid follicles formation walled with stratified squamous epithelium, adjacent collagen fibrosis (HE staining, ob $\times 40$)

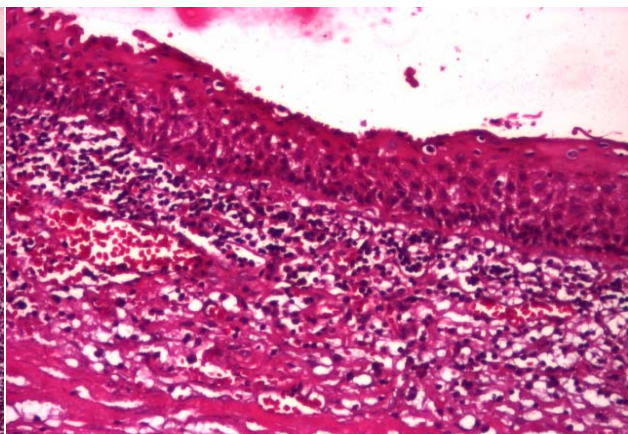


Figure 11 – Branchial sinus (detail): stratified squamous epithelium, adjacent lymphoid tissue and collagen fibrosis (HE staining, ob $\times 40$)

Ninety-five percent of abnormalities of the branchial cleft apparatus arise from the second cleft. At least 75% of all second branchial cleft abnormalities are cysts [12], which typically present when an individual is between 10 and 40-years-old. Second branchial cleft fistulas and sinuses are less common and usually present during the first decade of life [13, 14]. No gender predilection has been reported [15].

In our study approximately 40% of the patients had branchial cysts and the rest of 60% had branchial sinuses (there was no case of branchial fistulae), contradicting thus the specialty literature data that show a greater incidence of branchial cysts than of branchial sinuses and fistulae. Besides, most of the patients belonged to the first and second decades – 70% – (specialty literature data show a higher frequency of branchial cysts at adults – decade two and three). The rest of the results we obtained are similar to those in the specialty literature: there was no gender proclivity and no predisposition of a certain side of the neck (right or left).

Branchial cleft abnormalities are usually small, but can enlarge enough to cause difficulty swallowing and breathing. The most common symptoms of a second branchial cleft abnormality are following:

- Small lump or fluctuant masses in the lateral portion of the neck adjacent to the anteromedial border of the sternocleidomastoid muscle at the mandibular angle [13, 16, 17] (usually only on one side of the neck, rarely on both sides; usually painless, unless infected). There is also another symptoms: hoarseness (attributed to palsies of cranial nerves IX, X, and XII [18]), a bulge in the overlying pharynx [19]. Palsy of cranial nerves IX, X, XI, or XII is extremely rare in benign tumors or cysts, and usually suggests direct infiltration of local malignancies or metastatic lymph nodes [20, 21].

- Small opening in the skin that drains mucus or fluid near the front edge of the sternocleidomastoid muscle. The ostium is usually noted at birth just above the clavicle in the anterior neck [22].

At the studied group the symptomatology was the classical one, the patients had cervical distortions or a cervical tegumentary aperture.

Besides anamnesis and complete clinical examination, paraclinical explorations – echography and fistulography – were needed to obtain an accurate positive diagnosis.

At echography, a second branchial cleft cyst is seen as a sharply marginated, round to ovoid, centrally anechoic mass with a thin peripheral wall that displaces the surrounding soft tissues. The mass is compressible and shows distinct acoustic enhancement. Occasionally, fine, indistinct internal echoes, representing debris, may be seen.

At CT, these cysts are typically well circumscribed, homogeneously hypoattenuated masses surrounded by a uniformly thin wall [17]. The mural thickness may increase after infection. The cyst typically displaces the sternocleidomastoid muscle posteriorly or posterolaterally, pushes the vessels of the carotid space medially or posteromedially, and displaces the submandibular gland anteriorly [13, 22]. It may also be

seen more medially within the parapharyngeal space after extending through the stylomandibular tunnel and middle constrictor muscle [2, 23].

MR imaging better depicts the deep tissue extent of a second branchial cleft cyst, which allows accurate preoperative planning. The cyst fluid varies from hypointense to slightly hyperintense relative to muscle on T1-weighted images and is usually hyperintense on T2-weighted images [15].

For the studied group, as paraclinical exploration we practised fistulography – 13 cases (the patients diagnosed with branchial sinuses) and echography – 10 cases (the patients with branchial cysts). The echographic aspect of the cervical tumor, that of cystic formation, facilitated the diagnosis very much. Some authors [24–26] recommend radiological studies (CT, MRI). At the studied group, we did not make such complex radiological examinations as the clinical data; echography and respectively, fistulography were considered sufficient in diagnosing the patients accurately.

Pathologic characteristic – branchial cysts are usually filled with a turbid, yellowish fluid that may contain cholesterol crystals. Their walls are thin and lined with stratified squamous epithelium overlying lymphoid tissue [13, 27]. Columnar respiratory epithelium is occasionally present.

The anatomo-pathologic exams of the operated patients with second branchial cleft pathology have accurately determined the diagnosis, specifying the presence of stratified squamous epithelium.

Management of second branchial cleft anomalies is surgical excision [28]. Surgery should ideally be performed on the uninfected neck.

The studied patients were performed complete excisions of fistulous tracts (13 cases) – and as a proof stands the absence of recurrence within 1–5 years – and respectively, cervicotomies with branchial cysts excision (10 cases).

☒ Conclusions

Branchial cysts are frequently incorrectly diagnosed and forgotten in the differential diagnosis. Branchial cyst should be suspected in any patient with a swelling or a tumor in the lateral part of the neck, painful or painless.

Second branchial anomalies are frequently diagnosed at persons belonging to the first and second age decades. There is no proclivity of the position of branchial anomalies (right or left) or gender predisposition. Second branchial cysts have variable sonographic appearances, which may confuse the inexperienced; for accurate diagnosis, the doctor performing the exploration shall be highly accustomed with this aspect. In the absence of a clear result, CT and MRI are very useful in demonstrating the cystic nature and the anatomical extensions of branchial anomalies.

The histopathological examination establishes positive 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. 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.

Correct surgical operation will not allow a recurrence of the branchial anomaly.

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