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*Original Article*

# Clinicopathological characteristics of cervical chondrocutaneous branchial remnant: a single-institutional experience

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**Abstract:** Cervical chondrocutaneous branchial remnant (CCBR) is an uncommon developmental anomaly typically seen on the lateral neck. We recently experienced four cases of CCBR and initiated a comprehensive review of previously published cases. During a 10-year period, four (0.4%) of the 1,096 patients who underwent excision of branchial cleft anomalies were diagnosed as having CCBR at our institution. Patient age ranged from 2-6 years and patients presented with asymptomatic cutaneous masses present since birth measuring approximately 1 cm on the lateral neck. Three patients had congenital thyroid hemiagenesis, subependymal cyst, and tongue tie, respectively. We identified 76 previously published cases of CCBR. The median age of these patients was 18 months. CCBR developed more often in males (48/80; 60.0%). Most of the masses were located on the left (34/80; 42.5%) or right (18/80; 22.5%) lateral neck, whereas 23 (28.75%) involved bilateral lesions. Lesion size ranged from 0.3-3.5 cm. Grossly, the overlying skin of the masses was similar to the surrounding skin of the neck. Histologically, the lesions were covered by keratinizing squamous epithelium and had skin appendages and cartilage. Thirty-nine (48.75%) and 12 (15.0%) patients were found to have elastic and hyaline cartilage, respectively. Twenty-eight patients had single (13/28; 46.4%) or multiple (15/28; 53.6%) congenital anomalies. Forty-four different types of anomalies were reported. The most frequent anomalies were problems with cardiovascular and auditory systems. Our observations suggest that CCBR is a visible marker for more serious associated congenital anomalies. We recommend that clinicians and pediatricians further evaluate patients with CCBR through complete physical examination, abdominal and cardiac ultrasound, karyotyping, and biochemical marker analysis.

**Keywords:** Cervical chondrocutaneous branchial remnant, congenital cartilaginous rest of the neck, cervical accessory tragus, branchial cleft anomaly, congenital malformation

## Introduction

The branchial arches represent the embryological precursors of the face, neck, and pharynx. Anomalies of branchial arches are the second most common congenital lesions of the head and neck in children, accounting for approximately 20% of pediatric congenital head and neck lesions and occasionally forming parts of complex syndromes, especially those of the first and second branchial arches [1]. During the third to fifth week of embryonic development, the second branchial arch grows caudally and covers the third, fourth, and sixth branchial arches. When it fuses to the skin caudal to these arches, the cervical sinus is formed. Eventually, the edges of cervical sinus fuse and the ectoderm within the tube disappears. Persistence of the branchial cleft or pouch results in cervical anomalies along the anterior border of the sternocleidomastoid muscle from the tragus to the clavicle. They may present as cysts, sinuses, fistulae, or cartilaginous remnants.

Accessory tragi are fairly common, congenital anomalies of the external ear. They usually appear as small, skin-colored, preauricular tags or nodules and consist of skin, subcutaneous fat, and/or cartilage [2]. During the fifth to sixth week of embryonic development, the formation of the auricle of the ear is initiated from the first and second branchial arches. These arches then form the six hillocks (mesenchymal tubercles), and subsequently fuse to form the auricle structures [3]. The formation of accessory tragi

is due to errors during this period. The diverse clinical manifestations of accessory tragi may be unilateral or bilateral, single or multiple, and soft or firm. These anomalies may be isolated or associated with other congenital anomalies of the first and second branchial arches.

Similar to preauricular accessory tragi in appearance but located in the lower neck, cervical chondrocutaneous branchial remnants (CCBRs) are rather less common congenital lesions. They are among the rarest of branchial cleft anomalies. Owing to lack of consensus, there are numerous synonyms for CCBRs including branchial cartilages, cervical auricles, cervical accessory tragi, cervical skin tags, cervical vestiges, choristomas, papillomas, fibromas, wattles, and congenital cartilaginous rests of the neck [4-32]. In 1997, however, Atlan and colleagues [8] suggested the term CCBRs as a clear, widely acceptable name for these anomalies.

At present, 76 cases of CCBR have been documented in the English literature [4-32]. Existing confusion regarding the origin and the correct nomenclature is caused by the difficulty of clinical differential diagnosis of CCBRs, which mainly include sentinel tags next to the branchial sinus tracts or fistulae, acrochordons, and benign papillomas. Recently, some patients presented to our institution with CCBRs. Upon review of the previous literature and standard textbooks, we found very little detailed, clinically useful information concerning these lesions. Therefore, we decided to examine our institutional experience and review previously published cases thoroughly with the goal of clarifying their clinical and pathological characteristics, anatomical and surgical characteristics, and any associated local or distant congenital anomalies. Comprehensive analyses of cases may expand our knowledge regarding CCBRs.

## Materials and methods

### *Case selection*

The cases were selected from the computerized files of Department of Pathology, Severance Hospital, Yonsei University College of Medicine, Seoul, Republic of Korea. A thorough search was performed using the key words “branchial”, “branchial cleft cyst”, “branchial cleft fistula”, “branchial cleft sinus”, “cervical chondrocutaneous branchial remnants”, “congenital cartilaginous rests of the neck”, “cervical accessory tragus”, “cervical auricles”, “neck auricles”, and “wattle” among archival surgical pathology cases. Clinical and pathological information were obtained from the electrical medical record system and pathology reports. The clinical details that were reviewed included age at the time of diagnosis, sex, presenting symptom, side of lesion, number of lesion, size of lesion, associated congenital anomaly, histological type of cartilage, and coexisting unusual histopathological finding. This study was reviewed and approved by the Institutional Review Board at Severance Hospital, Yonsei University Health System, Seoul, Republic of Korea (2016-1518-001).

### *Histopathology and histochemistry*

The resected specimens were fixed in 10% neutral-buffered formalin and embedded in paraffin blocks. Each formalin-fixed, paraffin-embedded block was sectioned at 4 μm on a standard rotary microtome, and slices were brought from a water bath on cleaned slides and stained with hematoxylin and eosin. Masson trichrome staining and elastic van Gieson staining were also performed. The final histopathological diagnoses of the lesions were made by a board-certified pathologist.

### *Literature review*

The Medline database was thoroughly searched using the PubMed retrieval service. Searches were performed at August 2016, using the key words “branchial cartilages”, “cervical accessory tragus”, “cervical auricle”, “cervical chondrocutaneous branchial remnant”, “cervical skin tag”, “congenital cartilaginous rest of the neck”, “neck auricle”, and “wattle”.

## Results

### *Patient demographics*

During the period from August 2007 to July 2016, 1,096 patients underwent excision for branchial cleft anomalies. The ages of the 1,096 patients ranged between 1 month and 83 years (median, 12 years). The majority (972/1,096; 88.7%) of these patients were diagnosed with first or second branchial cleft

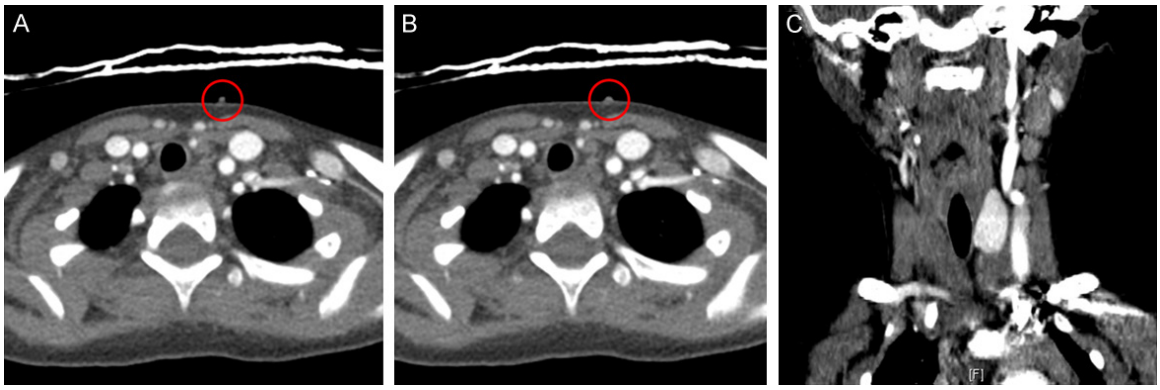


**Figure 1.** Image findings of cervical chondrocutaneous branchial remnants (case 2). A: Axial view of neck computed tomography reveals a polypoid mass (red circle) in the left neck. B: The mass (red circle) appears to extend to the subcutaneous layer, but there is no connection to other deep structures of the neck. C: A coronal view reveals right thyroid hemiagenesis.

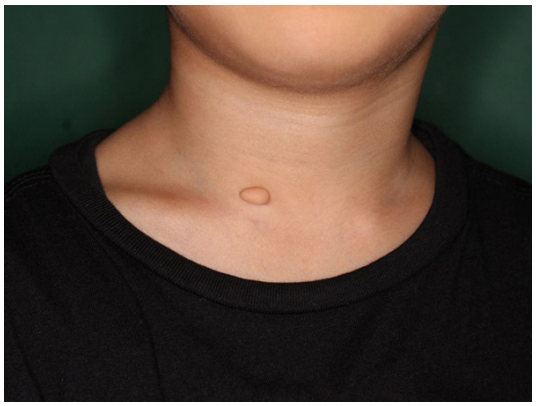

**Figure 2.** Gross findings of cervical chondrocutaneous branchial remnants (case 1). An ovoid, pedunculated mass is located at the right anterior neck. It is covered with normal-looking skin.

anomalies. Four (0.4%) patients were diagnosed with CCBR.

*Case presentation*

The patients' ages at the time of diagnosis were three (case 1), five (case 2), six (case 3), and two (case 4) years. There were two (50.0%) boys and two (50.0%) girls. All patients presented with asymptomatic cutaneous masses that had been present since birth measuring approximately 1 cm on the cervical area. In three (75.0%) cases the masses were located on the right side, and the remaining one (25.0%) mass was identified on the left lateral neck. None of the cases had bilateral lesions or median locations. Three (75.0%) patients had associated congenital anomalies: thyroid hemiagenesis (case 2), subependymal cyst (case 3), and tongue tie (case 4). Neck computed tomography (CT) was performed in one (25.0%) case (case 2) and revealed a tiny pedunculated mass on the left neck. The mass extended to the subcutaneous layer, but did not appear to be connected with deep underlying structures (**Figure 1A**, **1B**). In this case, the right thyroid was not identified (**Figure 1C**). All patients underwent surgical excision. The masses and underlying cartilage were removed with some subcutaneous tissue. There were no connections between the lesions and the deep soft tissue of the neck. Grossly, they were pedunculated masses covered with normal-appearing skin (**Figure 2**). The cut surfaces showed centrally located cartilaginous tissues. Histologically, the lesions consisted of overlying skin, a central core of mature cartilage, and surrounding subcutaneous tissue. The subcutaneous tissue contained normal skin appendages including hair follicles, pilosebaceous units, and eccrine glands. The overlying epidermis and subcutaneous fat were unremarkable. Masson trichrome and elastic van Gieson stain confirmed the presence of elastic cartilage in all cases. Representative photomicrographs are shown in **Figure 3**. No definitive evidence of epidermal alterations such as dysplasia or inflammation was noted. Brief case reports with detailed clinical information are provided below.

Case 1: A three-year-old boy presented with a painless skin tag measuring 1.0 cm on the right neck since birth. The mass was pedunculated and mobile in the subcutaneous soft tissue. It was covered with normal-looking skin without any epidermal alterations. He had neither a

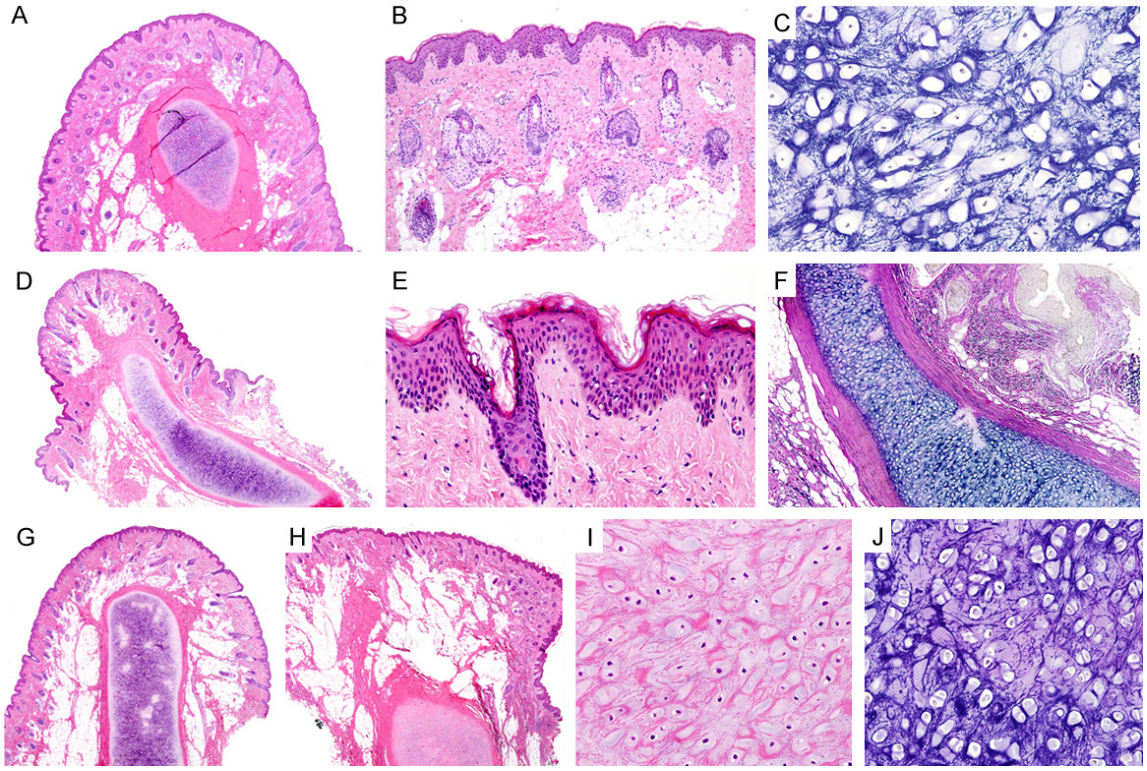


**Figure 3.** Histopathological and histochemical findings of cervical chondrocutaneous branchial remnants. (A) Case 1. Scanning view of the lateral neck mass reveals a central core of mature cartilage with surrounding fibrosis in the subcutaneous tissue (B). The epidermis and dermal pilosebaceous units are unremarkable. (C) Van Gieson stain highlights the elastic fibers surrounding the individual chondrocytes. (D) Case 2. The mass has a cartilaginous core with overlying skin and subcutaneous tissue. (E) The epidermis shows mild papillomatosis and hyperkeratosis. (F) Van Gieson stain confirms the presence of elastic cartilage. (G) Case 3. Scanning view reveals a polypoid lesion covered with unremarkable skin and subcutis and containing mature cartilage. (H) Case 4. An exophytic cutaneous nodule consists of overlying skin, a cartilaginous core and subcutaneous fat. (I) High-power view of the cartilaginous core reveals thin, pink-to-glassy red streaks surrounding individual chondrocytes. (J) Van Gieson stain highlights the elastic fibers.

familial history of congenital anomaly nor perceived malformations of other organs. He underwent surgical excision of the right neck mass.

Case 2: A five-year-old girl presented with an asymptomatic skin nodule measuring 1.3 cm on the left neck that had been present since birth. On physical examination, the mass seemed to be connected to the deep structures of the neck, forming a fistula, and the clinical impression was branchial cleft cyst. Neck CT revealed no connections between deep structures and right thyroid hemiagenesis. She underwent surgical excision of the left neck mass.

Case 3: A six-year-old girl presented with an asymptomatic skin tag measuring 1.2 cm on the right neck that had been present since birth. She suffered from respiratory distress and hyperbilirubinemia immediately after birth and was hospitalized for two months. Brain ultrasonography revealed a cystic lesion in the left caudothalamic notch. The ultrasonographic findings were interpreted as subependymal cyst. The right neck mass was completely excised.

Case 4: A two-year-old boy presented with tongue tie and an asymptomatic skin tag measuring 1.2 cm on the right neck that had been present since birth. The clinical diagnosis was branchial cleft cyst. The mass was a pedunculated, mobile mass covered with a normal-looking skin without any epidermal alterations. Complete surgical excision of the mass was performed.

**Table 1.** Summary of patient demographics and clinical characteristics

| Characteristic | | |
|---|---|---|
| Year reported | Range | 1985-2016 |
| No. of patient | Total | 80 |
| Age | Range | 7 days-51 years |
| | Median | 18 months |
| Sex | Male | 48 (60.0%) |
| | Female | 30 (37.5%) |
| | Unknown | 2 (2.5%) |
| Presenting symptom | None | 24 (30.0%) |
| | Unknown | 56 (70.0%) |
| Side of lesion | Left | 34 (42.5%) |
| | Bilateral | 23 (28.75%) |
| | Right | 18 (22.5%) |
| | Midline | 1 (1.25%) |
| | Unknown | 4 (5.0%) |
| No. of lesion | 1 | 57 (71.25%) |
| | 2 | 23 (28.75%) |
| Size of lesion | Range | 3-35 mm |
| | Median | 15 mm |
| No. of associated anomaly | Range | 0-10 |
| | None | 47 (58.75%) |
| | Multiple | 15 (18.75%) |
| | Single | 13 (16.25%) |
| | Unknown | 5 (6.25%) |
| Histological type of cartilage | Elastic | 39 (48.75%) |
| | Hyaline | 12 (15.0%) |
| | Unknown | 29 (36.25%) |

*Clinicopathological characteristics of cervical chondrocutaneous branchial remnants*

**Table 1** summarizes the patient demographics and clinicopathological characteristics of CCBRs. There are 76 previously published cases and four cases of CCBRs presented herein. The ages of the 80 patients ranged from 7 days to 51 years, with a median age of 18 months. CCBR developed more often in males (60.0%). Fifty-six (70.0%) cases did not include information about clinical symptoms. All remaining patients whose clinical presentations were available had asymptomatic neck masses. Most of the masses were located on the left (34/80; 42.5%) or right (18/80; 22.5%) lateral neck, while 23 (28.75%) cases were bilateral lesions. A single (1/80; 1.25%) patient had a CCBR arising from the midline of the neck. The lesion size was available for 45 cases and ranged from 3 to 35 mm, with a median size of 15 mm. Thirty-seven (82.2%) patients had CCBRs of 20 mm or less. Among the 75 cases available regarding the presence of associated congenital anomaly, 28 (37.3%) patients had single (13/28; 46.4%) or multiple (15/28; 53.6%) congenital anomalies. Grossly, the overlying skin was similar to the surrounding skin of the neck, without any differences in color or pilosity. Histologically, the lesions were covered by keratinizing squamous epithelium and had skin appendages with most containing hair follicles beneath. The deeper layers revealed fibroadipose connective tissue and cartilage. Thirty-nine (48.75%) and 12 (15.0%) patients were found to have elastic and hyaline cartilage, respectively. The details of clinical and pathological characteristics of CCBRs are shown in **Table 2**.

**Table 3** summarizes congenital anomalies associated with CCBRs. Forty-four different types of anomalies were reported. Among the associated conditions, some were as minor as serous otitis media, and others were more serious, including severe cardiac malformations. The most frequent anomalies were problems with cardiovascular (14 patients) and auditory (14 patients) systems, followed by those of head and neck (12 patients) and genitourinary systems (8 patients). The most common anomaly was cardiac ventricular septal defect (5 patients), followed by cardiac atrial septal defect (4 patients) and serous otitis media (4 patients). We herein report for the first time cases of subependymal cyst (1 patient) and thyroid hemiagenesis (1 patient) associated with CCBRs.

## Discussion

A choristoma is the result of displaced anlage and is a mass of tissue that is histologically normal for a tissue or an organ, but foreign to the tissue or site at which it is normally located [33]. Choristomas of the head and neck region have been reported in the oral mucosa, pharynx, middle ear, and cervical skin and soft tissue. Bhargava et al. [34] reported two cases of cartilaginous choristomas occurring in the tonsils of patients with chronic recurrent tonsillitis. Trowbridge et al. [35] described an asymptomatic cartilaginous choristoma of the tongue. Malis et al. [36] reported a case of nasopharyngeal cartilaginous choristomas associated with

**Table 2.** Patient demographics and clinicopathological characteristics of cervical chondrocutaneous branchial remnants

| Year Reported | Author and reference | No. of patient | Age | Sex | Presenting symptom | Side of lesion | No. of lesion | Size of lesion | No. of associated anomaly | Histological type of cartilage | Unusual histopathological finding |
|---|---|---|---|---|---|---|---|---|---|---|---|
| 1985 | Christensen et al. [4] | 1 | 12 years | Male | None | Right | 1 | 15 mm | None | Elastic | None |
| 1986 | Sperling [5] | 1 | 13 years | Female | None | Bilateral | 2 | 5 mm | None | Unknown | Pacinian corpuscle |
| 1991 | Vaughan et al. [6] | 1 | 11 years | Male | None | Left | 1 | 10 mm | None | Unknown | None |
| 1996 | Kim et al. [7] | 1 | 6 months | Unknown | None | Midline | 1 | 15 mm | None | Unknown | None |
| 1997 | Atlan et al. [8] | 17 | Unknown | Male | Unknown | Right | 1 | 30 mm | 10 | Unknown | None |
| | | | 2 months | Male | Unknown | Right | 1 | 16 mm | 10 | Elastic | None |
| | | | Unknown | Female | Unknown | Left | 1 | Unknown | 1 | Unknown | None |
| | | | 15 months | Female | Unknown | Left | 1 | 16 mm | None | Elastic | None |
| | | | 60 months | Male | Unknown | Left | 1 | 7 mm | 3 | Elastic | None |
| | | | 20 months | Male | Unknown | Left | 1 | 16 mm | 1 | Elastic | None |
| | | | 11 months | Male | Unknown | Left | 1 | 15 mm | 1 | Elastic | None |
| | | | 14 months | Female | Unknown | Left | 1 | 17 mm | 7 | Elastic | None |
| | | | 7 months | Female | Unknown | Right | 1 | 17 mm | 1 | Elastic | None |
| | | | 3 months | Male | Unknown | Right | 1 | 25 mm | 2 | Elastic | None |
| | | | 11 months | Male | Unknown | Left | 1 | 15 mm | 3 | Elastic | None |
| | | | 11 months | Male | Unknown | Left | 1 | 15 mm | None | Elastic | None |
| | | | 18 months | Female | Unknown | Right | 1 | 12 mm | 2 | Elastic | None |
| | | | 7 months | Male | Unknown | Bilateral | 2 | 12 mm | None | Elastic | None |
| | | | 16 months | Male | Unknown | Left | 1 | 22 mm | None | Elastic | None |
| | | | 13 months | Male | Unknown | Left | 1 | 8 mm | 2 | Elastic | None |
| | | | 9 months | Female | Unknown | Left | 1 | 13 mm | 3 | Elastic | None |
| 1997 | Dunlevy et al. [9] | 1 | 6 years | Female | None | Left | 1 | 25 mm | None | Unknown | None |
| 1997 | Kim et al. [10] | 1 | 25 years | Male | None | Bilateral | 2 | Unknown | None | Elastic | None |
| 1999 | Bendet [11] | 1 | 10 months | Male | Unknown | Left | 1 | 25 mm | 1 | Elastic | None |
| 2003 | Braun et al. [12] | 1 | 4 months | Male | Unknown | Bilateral | 2 | 25 mm | None | Elastic | None |
| 2003 | Fuad et al. [13] | 1 | 22 years | Male | None | Bilateral | 2 | 35 mm | 2 | Hyaline | None |
| 2003 | Rai et al. [14] | 1 | 6 years | Male | None | Right | 1 | 20 mm | None | Unknown | None |
| 2004 | Rund et al. [15] | 1 | 7 days | Female | Unknown | Left | 1 | 15 mm | None | Elastic | None |
| 2005 | Coras et al. [16] | 1 | 4 years | Unknown | Unknown | Bilateral | 2 | 5 mm | None | Elastic | None |
| 2006 | Konas et al. [17] | 1 | 6 years | Male | None | Left | 1 | 15 mm | None | Elastic | None |
| 2006 | Ozturk et al. [18] | 1 | 4 years | Male | Unknown | Bilateral | 2 | 15 mm | None | Elastic | None |
| 2007 | Gilboa et al. [19] | 3 | Prenatal | Male | Unknown | Left | 1 | Unknown | 1 | Unknown | Unknown |
| | | | Prenatal | Female | Unknown | Bilateral | 2 | Unknown | 2 | Unknown | Unknown |
| | | | Prenatal | Female | Unknown | Left | 1 | Unknown | 1 | Unknown | Unknown |
| 2007 | Rameh et al. [20] | 1 | 1 year | Male | None | Bilateral | 2 | 17 mm | 4 | Elastic | None |
| 2007 | Shin et al. [21] | 1 | 4 years | Male | None | Left | 1 | Unknown | None | Unknown | None |
| 2008 | Dayal et al. [22] | 1 | 4 months | Male | Unknown | Bilateral | 2 | 15 mm | None | Hyaline | None |
| 2011 | Nasser et al. [23] | 1 | 1 month | Female | Unknown | Bilateral | 2 | 25 mm | None | Unknown | None |
| 2012 | Choi et al. [24] | 1 | 4 years | Female | None | Left | 1 | 5 mm | None | Hyaline | None |

Clinicopathological characteristics of CCBR

| | | | | | | | | | | | |
|---|---|---|---|---|---|---|---|---|---|---|---|
| 2012 | Oiso et al. [25] | 2 | 4 years | Man | None | Bilateral | 2 | 7 mm | None | Unknown | Unknown |
| | | | 51 years | Man | None | Right | 1 | 6 mm | None | Unknown | Unknown |
| 2013 | Pham Dang et al. [26] | 6 | 10 years | Female | None | Bilateral | 2 | 20 mm | None | Unknown | None |
| | | | Unknown | Female | Unknown | Unknown | 1 | Unknown | Unknown | Unknown | Unknown |
| | | | Unknown | Male | Unknown | Unknown | 1 | Unknown | Unknown | Unknown | Unknown |
| | | | Unknown | Female | Unknown | Bilateral | 2 | Unknown | Unknown | Unknown | Unknown |
| | | | Unknown | Female | Unknown | Bilateral | 2 | Unknown | Unknown | Unknown | Unknown |
| | | | Unknown | Female | Unknown | Unknown | 1 | Unknown | Unknown | Unknown | Unknown |
| 2014 | Begovic et al. [27] | 17 | 2 months | Male | Unknown | Left | 1 | Unknown | None | Hyaline | None |
| | | | 5 months | Female | Unknown | Left | 1 | Unknown | None | Hyaline | None |
| | | | 6 months | Female | Unknown | Right | 1 | Unknown | None | Hyaline | None |
| | | | 6 months | Male | Unknown | Left | 1 | Unknown | None | Elastic | None |
| | | | 7 months | Female | Unknown | Left | 1 | Unknown | None | Elastic | None |
| | | | 7 months | Female | Unknown | Left | 1 | Unknown | None | Elastic | None |
| | | | 7 months | Female | Unknown | Bilateral | 2 | Unknown | 1 | Elastic | None |
| | | | 8 months | Male | Unknown | Bilateral | 2 | Unknown | None | Elastic | None |
| | | | 9 months | Male | Unknown | Right | 1 | Unknown | 3 | Elastic | None |
| | | | 13 months | Male | Unknown | Left | 1 | Unknown | None | Hyaline | None |
| | | | 15 months | Male | Unknown | Right | 1 | Unknown | 1 | Hyaline | None |
| | | | 54 months | Female | Unknown | Right | 1 | Unknown | None | Hyaline | None |
| | | | 7 years | Male | Unknown | Left | 1 | Unknown | None | Hyaline | None |
| | | | 7 years | Male | Unknown | Bilateral | 2 | Unknown | 1 | Elastic | None |
| | | | 15 years | Male | Unknown | Right | 1 | Unknown | None | Hyaline | None |
| | | | Unknown | Male | Unknown | Bilateral | 2 | Unknown | None | Unknown | None |
| | | | Unknown | Female | Unknown | Left | 1 | Unknown | 2 | Unknown | None |
| 2014 | Chander et al. [28] | 2 | 6 years | Male | None | Bilateral | 2 | 6 mm | None | Elastic | None |
| | | | 5 years | Male | None | Unknown | 1 | 10 mm | None | Elastic | None |
| 2014 | Lowry [29] | 1 | 3 years | Female | None | Left | 1 | 20 mm | None | Elastic | None |
| 2015 | Klockars et al. [30] | 7 | 11 months | Male | Unknown | Left | 1 | 5 mm | None | Unknown | Unknown |
| | | | 10 months | Male | Unknown | Left | 1 | Unknown | None | Unknown | Unknown |
| | | | 3 years | Male | Unknown | Left | 1 | Unknown | None | Unknown | Unknown |
| | | | 3 years | Male | Unknown | Right | 1 | Unknown | None | Unknown | Unknown |
| | | | Unknown | Male | Unknown | Bilateral | 2 | Unknown | None | Unknown | Unknown |
| | | | 2 years | Male | Unknown | Left | 1 | Unknown | None | Unknown | Unknown |
| | | | Unknown | Female | Unknown | Bilateral | 2 | Unknown | 3 | Unknown | Unknown |
| 2016 | Feito et al. [31] | 1 | 5 years | Female | None | Right | 1 | 8 mm | None | Elastic | Pacinian corpuscle |
| 2016 | Nielsen et al. [32] | 1 | 5 years | Male | None | Bilateral | 2 | Unknown | None | Hyaline | None |
| 2017 | Woo et al. [this study] | 4 | 3 years | Male | None | Right | 1 | 10 mm | None | Elastic | None |
| | | | 5 years | Female | None | Left | 1 | 13 mm | 1 | Elastic | None |
| | | | 6 years | Female | None | Right | 1 | 12 mm | 1 | Elastic | None |
| | | | 2 years | Male | None | Right | 1 | 12 mm | 1 | Elastic | None |

**Table 3.** Congenital anomalies associated with cervical chondrocutaneous branchial remnants

| Category | Associated congenital anomaly | No. of patient | Reference |
|---|---|---|---|
| Auditory system | Serous otitis media | 4 | [8] |
| | External auditory canal stenosis/aural atresia | 3 | [8, 11, 30] |
| | Microtia | 3 | [11, 13, 30] |
| | External ear malformation | 2 | [8] |
| | Low-set ear | 1 | [13] |
| | Sensorineural deafness | 1 | [8] |
| Cardiovascular system | Ventricular septal defect | 5 | [8, 19, 20, 27] |
| | Atrial septal defect | 4 | [8, 20, 27] |
| | Complete situs inversus | 1 | [19] |
| | Mitral regurgitation | 1 | [20] |
| | Patent ductus arteriosis | 1 | [8] |
| | Persistant left superior vena cava draining into the coronary sinus | 1 | [19] |
| | Tricuspid regurgitation | 1 | [20] |
| Central nervous system | Epilepsy | 1 | [8] |
| | Subependymal cyst | 1 | This study |
| Chromosomal disorder | XO mosaicism | 1 | [19] |
| Endocrine system | Thyroid hemiagenesis | 1 | This study |
| Gastrointestinal system | Inguinal hernia | 2 | [8] |
| | Umblical hernia | 1 | [8] |
| Genitourinary system | Hydronephrosis | 3 | [8] |
| | Vesicoureteral reflux | 2 | [8, 27] |
| | Cryptorchidism | 1 | [8] |
| | Hydrocele | 1 | [8] |
| | Hypospadia | 1 | [8] |
| Head and neck | Preauricular sinus/fistula | 3 | [8, 27, 30] |
| | Oronasal reflux | 2 | [8] |
| | Tongue tie | 2 | This study, [8] |
| | Branchial-oto-renal syndrome | 1 | [27] |
| | Cleft palate | 1 | [8] |
| | Occipital dermoid cyst | 1 | [30] |
| | Preauricular accessory tragus | 1 | [8] |
| | Retroauricular dermoid cyst | 1 | [8] |
| Musculoskeletal system | Arthrogryposis | 1 | [8] |
| | Club foot | 1 | [8] |
| | Congenital hand malformation | 1 | [8] |
| | Congenital hip dislocation | 1 | [8] |
| | Equinovarus | 1 | [8] |
| Respiratory system | Tracheomalacia | 2 | [8] |
| | Arytenoid dislocation | 1 | [8] |
| | Pulmonary atelectasis | 1 | [8] |
| Visual system | Duane syndrome | 1 | [8] |
| | Lacrimal duct stenosis | 1 | [8] |
| | Lateral eyebrow dermoid cyst | 1 | [8] |
| | Strabismus | 1 | [8] |

persistent adenoiditis. Anderhuber et al. [37] described salivary gland choristomas arising in the middle ear of a 4-year-old boy with conductive hearing loss. Thymic choristomas have

been documented in the surface of the cervical skin [38] and middle ear [39].

In 1985, Christensen et al. [4] first reported a case of CCBR occurring in a 12-year-old boy and designated it as a 'wattle' or 'cutaneous cervical tag'. The lesion had distinctive histological features and a cartilaginous core. The authors believed that the lesion was of auricular cartilage origin and noted that it contained a core of elastic cartilage, similar to the tragus. CCBRs are considered a type of choristomas arising from the first or second branchial arch. Primitive branchial structures contribute to the embryological formation of the head and neck. Most congenital anomalies in this region originate during their transformation into adult derivatives such that branchial anomalies frequently result from the persistence of parts of the branchial apparatus that would normally disappear [8]. The branchial arches begin to develop early in the fourth week of intrauterine life as neural crest cells migrate into the future head and neck. Six arches are formed, the fifth and sixth being rudimentary. By the end of the seventh week, each arch has completed its planned transformation such that any structural anomalies will already be present, including CCBRs.

There is disagreement as to the derivation of CCBR. In discussing the embryonic source of CCBRs, most authors subscribe to one of two theories: that they originate from ectopic auricular tissue [40], or that they arise from branchial tissues contributing to the formation of most cervical structures. Clues to their origin center around the histological type of cartilage found within the lesion. Since only the ear, epiglottis, corniculate cartilage, and part of the arytenoid cartilage are elastic in nature, the presence of elastic cartilage would suggest either an auricular (first or second branchial arch) source or perhaps derivation from the lower neck (fourth through sixth branchial arches). Clarke [40] suggested that CCBR represents ectopic external ear cartilage. This is based on the finding that CCBRs possess a central core of elastic cartilage and numerous telogen hair follicles containing vellus hair, as does the normal auricular cartilage. The external ear anlage is seen as a series of tubercles running along the hypomandibular cleft that extends anterolaterally to either side of the midline of the neck [41]. Since these tubercles produce the elastic cartilage of the external ear, they can be a source of a CCBRs occurring in the lateral neck. In this study, only elastic cartilage was found upon examination of all specimens prepared with van Gieson stain. The external ear forms from three hillocks derived from the first branchial arch, while the rest of the auricle originates from three more caudal hillocks of second branchial arch origin. These six hillocks first appear located in a relatively ventral position and then gradually coalesce and migrate along the general direction of the sternocleidomastoid muscle to their final lateral facial position. It is therefore possible that the CCBRs are left behind by the auricular hillocks during this migratory process. This hypothesis is also compatible with their relatively superficial position and location in relation to the sternocleidomastoid muscle.

However, this explanation is not likely to be correct for CCBRs of the ventral midline area. Nieden and Asbeck [42] observed small epithelial rests in two cases and believed that these rests were remnants of incompletely obliterated clefts, supporting the hypothesis that CCBRs are thus of branchial origin. The epiglottis and some of the laryngeal cartilages are of branchial origin and are normally composed of elastic cartilage. The epiglottis is first distinctly noted as a prominence in the midventral area of the pharynx between the third and fourth branchial arches. The epiglottis is embryologically a strictly ventral midline structure. Therefore, it is likely that a displaced portion of epiglottic of laryngeal anlage is the source of CCBRs occurring in the ventral midline area. It remains unclear whether CCBR originates from auricular cartilage or as a remnant of branchial cartilage.

CCBRs have proven to be visible markers for more serious anomalies. Our review confirmed that approximately one-third of patients with CCBRs have associated congenital anomalies, although the prevalence of each anomaly varies. Anomalies reported include those involving auditory, cardiovascular, nervous, gastrointestinal, genitourinary, musculoskeletal, respiratory, visual, and endocrine systems. In 1997, Atlan et al. [8] performed the largest case study of CCBRs and described a high incidence (76.5%) of associated congenital anomalies. Similarly, Begovic et al. [27] documented that 29.4% (5/17) of cases had associated anomalies; two

had ventricular septal defect and atrial septal defect, respectively, and the others each had vesicoureteral reflux, sinus preauricularis, and branchio-oto-renal syndrome. In our study, 3 of the 4 children had associated congenital anomalies, representing a relatively high incidence (75.0%) of anomalies. In contrast, there have been several case reports describing CCBRs that were not associated with other congenital anomalies or malformations. We hypothesize that this discordance is attributable to lack of thorough surveys for the presence of anomalies by clinicians and pediatricians. Clinical surveillance for congenital anomalies should be performed for patients with CCBRs, especially for the auditory system, cardiovascular system, genitourinary system, and head and neck region. Atlan et al. [8] stated that systematic investigations, except for ultrasound examination for the genitourinary tract, did not provide useful additional information beyond that obtained from a complete physical examination. We suggest that a meticulous physical examination and abdominopelvic ultrasound by qualified pediatricians and/or radiologists should be undertaken in patients diagnosed as having CCBRs.

The differential diagnosis of CCBR includes branchial cleft and thyroglossal duct cysts, thymic cyst, and congenital midline hamartoma. Branchial cleft cysts are located laterally and are lined by upper respiratory epithelium; they contain seromucinous glands and have a shallow epidermal pore. However, thyroglossal duct cysts are located anteriorly and may contain thyroid follicles. Thymic cysts are usually not congenital, are fluid filled, and are lined with stratified squamous epithelium. Congenital midline hamartomas are found on the chin, and they have a prominent skeletal muscle component. Epidermoid cysts, fibroepithelial polyps, and squamous papillomas may mimic the gross appearance of CCBRs, but they do not usually present difficulties in histopathological diagnosis.

Surgical treatment of CCBR requires only simple excision extending no deeper than the superficial neck musculature. Consistent with previous studies, in our cases, no associated fistulous tracts or gross connections with deeper structures were identified. It is not recommended to shave CCBRs, but to remove the lesion completely. Treatment is recommended before school age for social reasons and for histological verification, but can be postponed until a suitable and safe age. Because of their straightforward excision, some surgeons advocate performing surgery under local anesthesia with sedation in adults [8]. Our patients received surgery under general anesthesia because of their young ages.

In conclusion, it is important to recognize that CCBRs may be markers for other serious congenital malformations and/or anomalies. In the clinical setting of CCBR, clinicians and pediatricians should initiate thorough physical examinations, full anamnesis including family history and patient medical history, and ultrasonography of the abdomen, pelvis, and heart.

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## Disclosure of conflict of interest

None.

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