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ORIGINAL ARTICLE

Importance and Impact of Appropriate Radiology in the Management of Branchial Cleft Anomalies

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Abstract Branchial cleft anomalies are common differential diagnosis of neck masses. However, depending on the origin (1st, 2nd, 3rd or 4th arch), this pathology presents at different locations as cyst, sinus and fistula. Incomplete excision or incision and drainage of infected branchial cleft anomalies (cystic presentation) can lead to multiple recurrences. Appropriate radiology is imperative to make a correct diagnosis and to achieve complete excision to prevent recurrence. Our case series highlights the mode of presentation, appropriate radiology and management for each type of branchial cleft anomalies. Data of 27 patients with the diagnosis of branchial cleft anomaly and treated in the department of Otolaryngology—Head and Neck Surgery in a tertiary care referral centre in last 5 years was analysed retrospectively. Demographic data in terms of age, sex, laterality, clinical presentation, duration of symptoms and radiological investigations if any were recorded. The mean age at presentation was 22.1 years in this series of 27 cases including six (22.2%) recurrent cases. Most common clinical presentation was discharging sinus (59.25%) followed by cystic neck swelling (33.3%). Second branchial cleft anomalies were commonest (51.8%) followed by first branchial cleft anomalies (29.6%). Appropriate radiology was available for 17 (62.9%) patients. Branchial cleft anomaly is an important differential diagnosis of neck mass. Appropriate radiology helps in complete excision and prevents recurrences. Recurrent cases pose surgical challenge owing to fibrosis from previous surgery which further increases the chances of incomplete excision.

Keywords Branchial cleft anomalies · Sinus · Fistula · Cyst · Facial nerve monitoring

Introduction

Branchial cleft anomalies (BCA) are the third most common neck masses after lymphadenopathy and thyroglossal duct anomalies [1]. BCA usually present as either cyst, sinuses, or fistulas. Second branchial cleft anomalies are the most commonly seen followed by first, third and fourth BCA in that order [2]. Although congenital in nature, presentation in second and third decades of life is frequent due to the asymptomatic (mass) or occasionally discharging (sinus/fistula) status of the lesions. It is important to establish correct diagnosis with regard to the type of BCA to achieve complete excision and prevent recurrence. This is imperative because recurrent cases are associated with surgery induced scarring and fibrosis which make complete re-excision very challenging. The importance of appropriate radiology and comprehensive management for each type of BCA is discussed in this case series along with brief review of literature.

Methods

A retrospective search and retrieval of medical records of all patients operated for BCA in the department of Otolaryngology and Head Neck Surgery was done from January 2011 to June 2018 was done. Various demographic,

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Table 1 Age distribution

Age group	No. of patients
< 10	4
10–20	9
21–30	10
>30	4

Table 2 Incidence of individual anomalies and their modes of presentation

Branchial arch anomalies	Presentation		
	Cyst	Fistula	Sinus
1st Branchial arch n = 8 (29.6%)	2	0	6
2nd Branchial arch n = 14 (51.8%)	7	2	5
3rd Branchial arch n = 2	0	0	2
4th Branchial arch n = 3	0	0	3

clinical, radiological and surgical data were further analysed for a total of 27 patients of BCA.

Results

Out of 27 patients, complete demographic details was unavailable for five patients who were operated in outpatient basis. There was slight male preponderance (male: 16; female: 11). The mean age at presentation was 22.1 years with an age range of 5–50 years. Most commonly patients with BCA presented in third decade of life (n = 10) followed by second decade of life (n = 9). Table 1 summarizes age distribution for analysed group.

The different modes of presentation as external skin pit with discharge (sinus), neck swelling (cyst) and fistula (radiological diagnosis) is summarized in Table 2. Most common presentation in our series for BCA was a discharging sinus (n = 16/27; 59.2%) followed by cystic neck swelling (n = 9/27; 33.3%). Sixteen BCA were on right side while 11 were left sided.

Diagnosis for BCA was usually established clinically supplemented by radiology. Fine needle aspiration cytology (FNAC) was done in six cases of cystic neck swelling to establish cytological diagnosis (Table 3). CT sinogram was done in 8 cases for tract delineation; CECT Neck was done in 6 cystic neck swelling for evaluating extent and relation to great vessels. MRI Neck was done in 4 cases. Two patients had dual imaging of both CT sinogram and MRI Neck. For ten patients no radiological details were available. The radiological findings are summarized in Table 4.

Discussion

BCA are commonly seen in first three decades of life. Discharging sinuses are usually reported by parents in first decade of life while cysts are usually reported in second and third decade of life. There is no sex predilection reported however our series has shown slight male preponderance [3]. The modes of presentation for BCA can either be sinus, cyst or fistulas. In our series, discharging sinus was most common presentation (n = 59.2%) followed by cysts.

Second BCA are most commonly reported anomaly in literature. In our series, incidence of second arch anomalies (51.8%) was also highest followed by first arch anomalies (29.6%).

Clinical Features

First Branchial Cleft Anomalies

First BCA constitutes 1% of all branchial cleft anomalies [4]. They can present in close relation to parotid, external auditory canal (EAC) and anterior neck near angle of mandible. Patients usually present in first two decades of life. Delayed presentation is often seen in misdiagnosed cases as seen in our series. Presentation can be either a cystic parotid mass or discharging sinus in anterior neck or a fistula with internal opening in EAC.In our series of eight cases, six had sinus presentation and two were cystic. Acierno et al. classified first BCA into type I and type II. Type I anomalies are lateral to facial nerve and presents as parotid swelling whereas Type II anomalies are deep to facial nerve and located in between EAC and hyoid bone [3]. First BCA with cystic presentation is usually misdiagnosed on initial presentation due to infection. Infected cyst mimics abscess and usually managed with incision and drainage. This lead to recurrence and often multiple I&D before correct diagnosis are made. Correct diagnosis can be made with proper clinical history, examining EAC for fistulous opening and high index of suspicion for first BCA. Authors has previously reported a series of four cases of first BCA emphasizing the role of facial nerve monitoring while operating on recurrent first BCA [5]. CECT is usually done for cystic parotid anomalies (Fig. 1) whereas CT sinogram is recommended for sinus presentation. Few authors has advocated CEMRI for tract delineation within the parotid parenchyma as tract contained some amount of fluid [6] as shown in Fig. 2. Superficial parotidectomy is recommended to achieve complete excision along with cuff of cartilaginous EAC if tract extends till floor of EAC.

Table 3 Summarizes FNAC findings for six patients presented with cystic neck swelling

Sr nos.	Age/sex	Complaints	Examination	FNAC findings
1.	50/M	Right side neck swelling for 2.5 years; recurrent	3 × 2.5 cm swelling right level II, non fluctuant	Branchial cyst
2.	21/F	Recurrent swelling left side of neck for 16 years	3.5 × 3.5 cm left parotid region swelling, well defined	Epidermal inclusion cyst
3.	34/M	Swelling left side of neck for 10 years	Swelling left side of neck 1 × 2 cm, mobile	Branchial cyst
4.	39/F	Swelling left side of neck for 2 years	Swelling left side of neck along anterior border of SCM muscle upper 1/3rd	Branchial cyst
5.	28/F	Swelling right side neck for 2 years	Swelling right side neck upper 1/3rd	Branchial cyst
6.	23/F	Swelling right side neck for 2 years	Swelling right side neck upper 1/3rd	Branchial cyst

Second Branchial Cleft Anomalies

Second BCA are most common of all branchial cleft malformations. The most common presentation is either a cystic swelling in relation to anterior border of sternocleidomastoid muscle (SCM) (Fig. 3) or discharging sinus along lower third of SCM muscle. In our series, cystic and sinus presentations were most common mode of presentation (n = 12/14; 85.7%). Fistulous presentation is rarer and usual internal opening is in superior aspect of tonsillar fossa. Figure 4 shows X-ray sinogram delineating the tract till right tonsillar fossa. However CT sinogram is better imaging as it shows anatomical details for operating surgeon. Presentation is usually in third to fourth decade of life for cystic anomalies where sinuses/fistulae usually present early in life as seen in our series. Upper respiratory tract infections often lead to increase in size of swelling, purulent discharge and abscess formation. CECT neck and/or CT sinogram is usually done for pre-operatively depending on type of presentation (cystic vs. sinus). FNAC of cystic presentation confirms diagnosis by presence of either respiratory or squamous epithelium.

Third and Fourth Branchial Cleft Anomalies

The presentation of third and fourth BCA have similar clinical presentations but anatomically third BCA tract originates from base of pyriform sinus and crosses above in relation to superior laryngeal nerve whereas fourth BCA commonly originates from apex of pyriform sinus and crosses below superior laryngeal nerve [4]. Third BCA commonly presents as cystic swelling but in our series sinus was present in both cases. Third BCA can also cause thyroiditis as tract in this anomaly lies in close relation to thyroid gland (Fig. 5) [7]. Treatment involves surgical excision of sinus tract along with partial thyroidectomy if thyroid gland is involved [8].

Fourth BCA can presents as lateral neck mass, abscess or suppurative thyroiditis in lower third of neck [9]. In our series, all three cases had sinus presentation and were recurrent (Fig. 5). Fourth BCA on right side loops around subclavian artery and on left side it loops around aortic arch then ascends in similar course on both sides [3]. Fourth BCA tract has close relationship with thyroid cartilage and trachea, surgery is the mainstay of treatment and it is necessary to remove portion of thyroid gland as done in one of our case [10].

Radiology

Diagnosis is usually made by history and clinical presentation. However, significance of appropriate radiology has not been highlighted in the management of BCA. For sinus presentation, CT sinogram is an apt radiology which delineates the tract completely and shows relation of the tract to great vessels. CT sinogram also gives information regarding entire course of tract and also regarding inner opening in mucosal surface. Thus CT sinogram changes clinical diagnosis of sinus into fistula. On other hand, for cystic neck masses both CECT/MRI are suitable. MRI is superior in terms of delineating contents of cystic masses and relation with surrounding structures in neck. For 1st BCA with sinus or fistulous presentation, MRI is better imaging as fluid present in tract can be traced and relation to facial nerve can be made (keeping retromandibular vein as surrogate marker for facial nerve). In our series, CT sinogram (N = 8) was done in patients with sinus presentation and CECT Neck (n = 6) was done in cystic neck masses. MRI Neck was done in four cases only. In the series by Choi and Zalzal [11], CT scan was performed on 15.38% of patients, sinogram/contrast study was performed on 7.69% of patients, and ultrasound/MRI was done on 1.92% of patients. Computed tomography (CT) is usually the first modality done for cystic BCA. The usual findings for cystic 2nd BCA are low density (10–25 HU) cystic masses with a thin wall which are well circumscribed (Fig. 3). In the background of repeated infections the cyst wall become thickened, irregular and show enhancement

Table 4 Summarizing radiological investigations and findings done for patients

S. nos	Age/sex	Complaints	Clinical findings	Radiological investigations	Type
1	19M	Rt submandibular region neck swelling for 2 years	2.5 × 1.5 cm right submandibular region neck swelling	USG neck—4.9 × 4.8 × 4.2 cm unilocular cyst with internal echoes at right submandibular deep to SCM MRI—suggesting 2nd branchial cyst	Right IInd branchial cyst
2	14 F	Mucoid discharge left side of neck 1.5 years	Pin point opening at ant border of left SCM at upper 2/3rd and lower 1/3rd, mucoid discharge	Sinogram-sinus tract for a distance of 28 mm from external opening	Left IInd branchial sinus
3	12 M	Mucoid discharge right side of neck since birth	Pin point opening at ant border of right SCM at upper 2/3rd and lower 1/3rd	CT sinogram-2nd branchial sinus, no internal opening, tract in subcutaneous tissue lateral to CCA and IJV on right side	IInd branchial sinus
4	19 F	Pus discharge right side of neck for 8 years; operated twice		CT sinogram—blind tract going post and medially deep to SCM	3rd Branchial sinus
5	50 M	Right side neck swelling for 2.5 years; recurrent	3 × 2.5 cm swelling right level II, non fluctuant	CT neck—homogenous soft tissue density, capsulated	Right IInd branchial cyst
6	11 F	Pus discharge left side of neck for 7 years	No opening seen; 4 × 2 cm hypertrophic irregular scar + left thyroidal region	CT sinogram—Mild STD in mid lower neck on left side with contrast filled blind sinus	Left recurrent Branchial sinus type IV
7	5 F	Mucoid discharge right side of neck since birth	Pin point opening 2 cm above medial end of clavicle rt	CT sinogram—Type II Branchial sinus extending up to hyoid bone	Right type II branchial sinus
8	9 F	Pus discharge right side of neck since childhood	Pin point opening at ant border of rt SCM at upper 2/3 rd and lower 1/3 rd	CT sinogram—Right branchial fistula; outer opening at midpoint of SCM and internal opening in right tonsillar fossa	Right II branchial fistula
9	6 M	Mucoid discharge right side of neck since birth	Pin point opening at ant border of rt SCM at upper 2/3rd and lower 1/3rd	CT sinogram—linear tract extending ant to Right SCM, ant to carotid sheath extending to lateral wall of oropharynx	Right type II branchial sinus
10	21 F	Recurrent swelling left side of neck for 16 years	3.5 × 3.5 cm left parotid region swelling, well defined	CT scan-s-48179-well defined cystic lesion in parotid space extending in deep lobe abutting ICA, ? 1st branchial cleft cyst	Right brachial cyst
11	20 M	Swelling left side of neck for 8 years; recurrent	Puckered scar left neck extending from midline	CECT neck-thin air filled tract from abscess cavity to left pyriform sinus	Left type IV branchial sinus
12	19 F	Mucoid discharge from right side of neck since childhood	Pin point opening at ant border of right SCM at upper 2/3rd and lower 1/3rd	Sinogram—IInd branchial fistula with internal opening in right tonsillar fossa	Right type II branchial fistula
13	24 M	Pus discharge left side of neck since childhood, recurred after surgery twice	Pin point opening anterior to sternal end of lower 1/3 rd of left SCM, without surrounding skin scarring	MRI neck and CT sinogram-Nov 2016—2.5 cm long tract with cutaneous opening lying at lower end of left SCM, passing deep to SCM, superficial to strap muscles and passing across left thyroid lobe. B/l PFS normal	Left type IV branchial sinus
14	20 M	Pus discharge left preauricular sinus since childhood	Pin point opening left pre auricular sinus	MRI neck and temporal bone—first branchial anomaly type 1b HRCT—first arch anomaly	Left branchial arch type 1 anomaly sinus
15	39 F	Swelling left side of neck 2 years	Swelling left side of neck along anterior border of SCM upper 1/3rd	CECT—left sided 2nd branchial cleft cyst	Type II branchial anomaly cystic
16	28/F	Swelling right side neck × 2 years	Swelling right side neck upper 1/3rd	CECT: right sided 2nd branchial cleft cyst	Type II Branchial anomaly cystic
17	23/M		Swelling right side neck upper 1/3rd	CT neck/MRI Neck—homogenous soft tissue density, capsulated	Type II branchial anomaly cystic

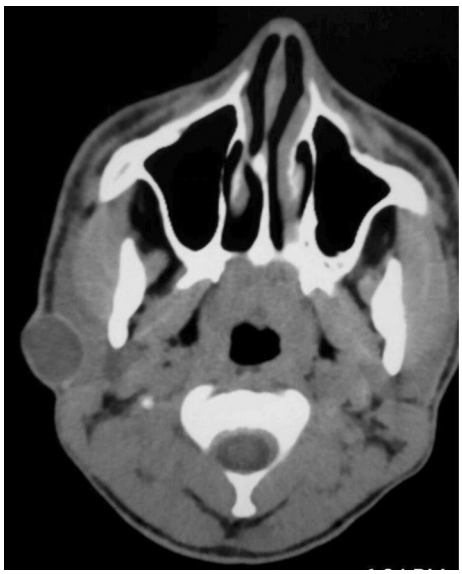

Fig. 1 Showing well circumscribed, thin walled cystic lesion (hypodense) in right parotid type I BCA (cystic presentation)

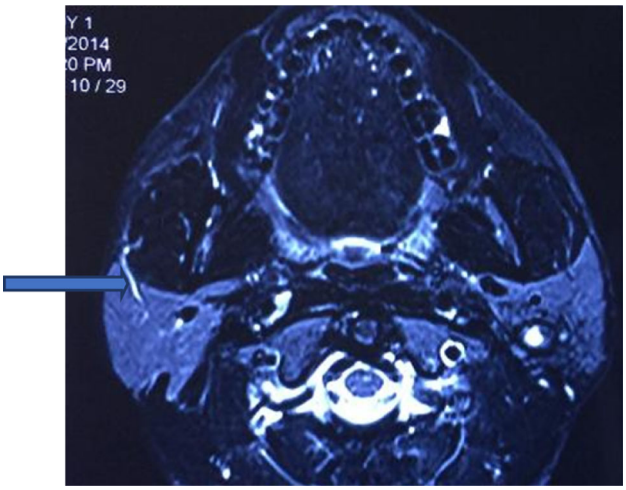

Fig. 2 Showing axial cut of MRI parotid gland with a fluid filled tract traversing in parotid parenchyma (bold arrow) in superficial lobe lateral to retromandibular vein (Type I BCA with sinus presentation)

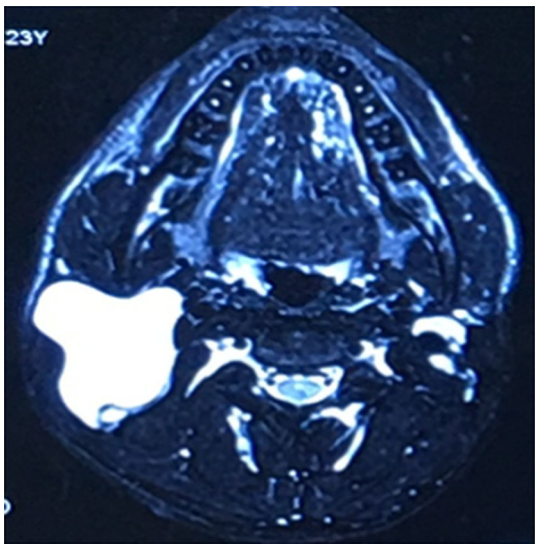

Fig. 3 T2 weighted MRI neck (axial cut) showing hyperintese fluid filled Type II BCA (cystic presentation)

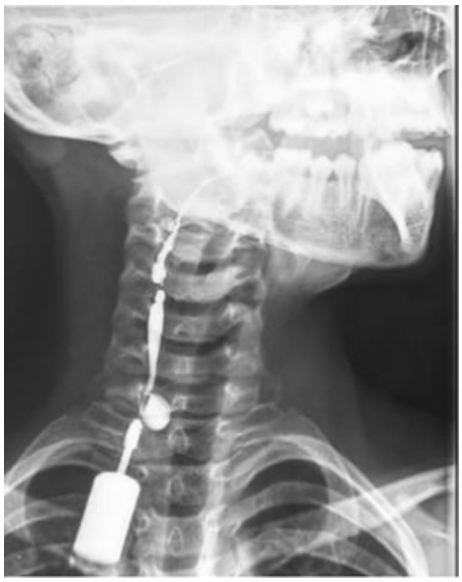

Fig. 4 Showing X-ray sinogram for 2nd branchial sinus with tract delineation up to right tonsillar fossa

with contrast. Peri-lesional fat stranding can also be seen in some cases [7, 12]. MRI is additional modality to look for relation with great vessels and deeper structures especially in recurrent cases or cases with repeated history of infection leading to abscess formation. MRI shows low—intermediate signal intensity on T1—weighted and high intensity on T2 weighted sequences. T1—weighted can be of high signal intensity if protein content of mucous increases within the cystic BCA. High cost and general anaesthesia for paediatric age group makes it less favourable initial modality of choice [7, 12]. Few authors have also advocated use of Ultrasound (US) as initial imaging as it is readily available, rapid and low cost without any radiation exposure. However, operator dependency and lack of anatomical details are limitations for US. Cystic BCA are usually seen as round lesions with low echogenicity and lack of internal septations. There is no acoustic enhancement in cases of BCA [7].

Differential Diagnosis

BCA are usually diagnosed on history and clinical examination. Presence of pit like opening with discharge since birth usually clinches diagnosis for sinus presentation of

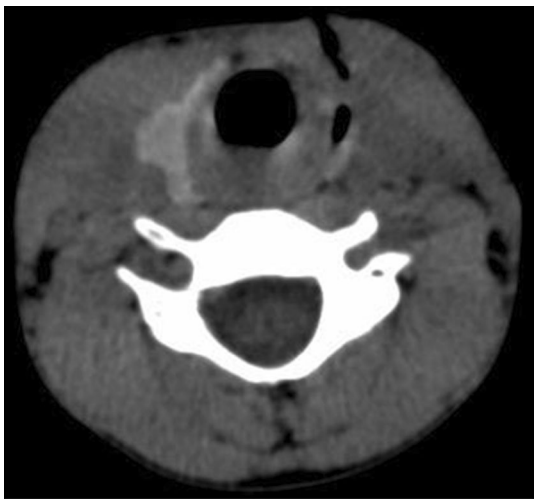

Fig. 5 CECT Neck (axial cut) showing sinus tract traversing through left thyroid lobe (Third arch sinus)

BCA. However, many conditions mimic BCA and poses challenge for diagnosis. Guldfred et al. [13] in their series of 126 patients showed positive predictive value of 86% and sensitivity of 94% for accurately diagnosing BCA pre-operatively. They also concluded that any cystic neck swelling should be considered carcinomatous until proved otherwise.

For 1st BCA, the differential diagnosis includes parotitis with abscess formation, lymphatic malformation and sialocoele [12]. Strong suspicion for 1st BCA is required along with appropriate radiology (preferably CEMRI) for accurate diagnosis. For 2nd BCA (cystic presentation) the differential diagnosis includes cystic nodal metastasis, cystic schwanoma, lymphatic malformation and lymphadenopathy [7, 12, 13]. Pre-operative FNAC should be done in all cases along with radiology (CECT/CEMRI) for diagnosis. FNAC usually yields fluid with cholesterol crystals and lining epithelium of stratified squamous nature. Cystic nodal metastasis can occur from papillary carcinoma thyroid, tonsillar carcinoma and carcinoma nasopharynx. Through clinical examination of all ENT sites should be performed in routine work-up. The differential diagnosis for 3rd BCA includes cervical thymic cyst, cystic nodal metastasis, lymphatic malformation, infrahyoid thyroglossal cyst and laryngocele [12]. History, clinical examination and appropriate radiology along with FNAC are helpful in differentiating these conditions from each other. The differential diagnosis for 4th BCA also includes cervical thymic cyst, thyroid colloid cyst and lymphatic malformation [12]. Initial imaging with US helps to differentiate thyroid colloid cyst and lymphatic malformation from BCA. Endoscopic examination is helpful in locating opening in pyriform sinus if there is no external skin opening.

Treatment

Definitive treatment for brachial cyst, sinus and fistula is surgical excision. These lesions do not spontaneously regress and can have recurrent infections. In our series, all 27 cases underwent surgical excision. Recurrent cases pose difficulty in complete excision especially 1st BCA where facial nerve is at risk owing to fibrosis. Facial nerve monitoring is recommended for recurrent 1st BCA especially sinus/fistulous presentation. Some authors has suggested purely endoscopic electrocauterization with trichloroacetic acid (TCA) for third and fourth BCA with internal opening only [14, 15]. Success rates up to 78% has been reported by authors however in recurrent cases open surgical approach with hemithyroidectomy was performed.

Conclusion

Branchial cleft anomalies are important differential diagnosis of cervical masses. Surgical excision is the treatment of choice. Tract delineation in cases of sinuous/fistulous presentation by appropriate radiology is important to achieve complete excision.

Compliance with Ethical Standards

Conflict of interest Rajeev Kumar, Prem Sagar, Chirom Amit Singh, Rakesh Kumar, Alok Thakar, Suresh C Sharma declare that they have no conflict of interest.

Ethical Approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

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