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Experience of managing Branchial cleft anomalies at civil hospital, Karachi

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Abstract:
Objective: To evaluate the management of different Branchial cleft anomalies encountered in our department.
Study Design: Retrospective analytical.
Settings: Department of ENT, Head & Neck surgery, Dow Medical College & Dr. K.M. Ruth Pauf Civil Hospital, Karachi.
Duration: January 2013 to January 2018.
Material & Methods: A total of 26 cases of branchial cleft anomalies were identified who visited outpatient department. They were diagnosed on the basis of history, clinical examination including location of the lesion and radiological investigation including sino-fistulogram and or CT scan for cystic lesion.
Results: Out of total 26 subjects, 17(65.38%) were males and 9(34.61%) were females, with male to female ratio of 1.75: 1. On presentation majority of the cases (n-18) 69.23% were having cervical sinus, 4(15.38%) cases presented with cyst out of which 03(11.53%) were infected with history of incision and drainage, 4(15.38%) cases of clear cut fistula were identified. The age range was between 15- 35 years with a mean of 19.4 years. Majority were second arch defect with cervical sinus or cyst with only 2(7.69%) cases having third arch defect with fistulous communication between pyriform fossa and cervical skin. First arch defect was not identified in present series. All the cases of branchial sinuses and fistula were managed surgically with standard stepladder technique while the branchial cysts were removed with standard cervical incision. Recurrence wasrecorded2(7.69%) on long term follow-up.
Conclusion: Branchial cleft anomalies are not very common and not confined to pediatric population. Second brachial cleft anomalies are the commonest of the true Branchial cleft abnormalities and the diagnoses is usually straight forward, sinuses are more frequent than cyst and fistulae are extremely rare. The management of this rare lesion is challenging and requires knowledge of the embryological anatomy of branchial apparatus. With the classical approach complete excision must be ensured.

Keywords: Branchial cleft anomalies, branchial sinus, branchial fistula, branchial cyst.

Introduction:
The branchial apparatus appears between the fourth and fifth week of fetal development. It consists of six paired branchial arches separated by brachial clefts externally and branchial pouches internally. In human branchial cleft contribute to the formation of variety of head and neck structures including cervical skin, jaw, middle and external ear. Different developmental anomalies are associated with these branchial cleft anomalies managed at our department.

The term branchial cyst was first used by Ascheron in 1832. He suggested that these cysts were results of impaired obliteration of branchial clefts. Incomplete, failed or persistent embryonic development of these arches results in several anomalies or defectsin the neck.[1] The most

widely held belief is that incomplete obliteration of the cervical sinus plays an important role in this process.[2] Different theories of origin of branchial cleft anomalies had been put forward viz

- Branchial apparatus theory suggests that branchial cysts represent the remains of pharyngeal pouches or bronchial clefts or a fusion of these two elements.
- The cervical sinus theory suggests that branchial cysts represent the remains of the cervical sinus of His.
- The thymopharyngeal duct theory suggests the cysts are remnants of the original connection between the thymus and third branchial pouch.
- The inclusion theory is widely accepted. This theory suggests that branchial cysts are epithelial inclusions within a lymph node.[3,4]

Branchial cleft anomalies are the second most common head and neck congenital lesions seen in children.[5] They are account for up to 17 percent in all peadiatric cervical masses.[6] They are bilateral in about 1 percent of the cases, without any preponderance to one side right or left in which they develop.[7] They are located in the cervical region, parotid and mediastinum. Many developmental anomalies of the branchial apparatus have been identified: cysts, fistulas, sinuses, ectopic glands, and malformations of head and neck structures. Branchial cleft cysts are congenital lesions often presenting as lateral neck masses.

Branchial cleft anomalies were classified into four types. Type-I anomalies are located near external auditory canal. Most commonly, they are inferior and posterior to the tragus. Type-II anomalies are associated with the submandibular gland or found in the anterior triangle of the neck. The second branchial cleft anomalies accounts for 95 percent of branchial anomalies. Most frequently, these are identified along the anterior border of the upper third of sternocleidomastoid muscle, adjacent to the muscle. However, these may presents anywhere along the course of the second branchial fistula, which proceeds from the skin of the lateral neck, between the internal and external carotid arteries, and into the palatine tonsil.[8] Second branchial cleft anomalies are the commonest of the branchial cleft abnormalities, and the diagnosis is usually straightforward.[9] Good knowledge of anatomy and embryology are necessary for proper treatment.[10] Of the second branchial cleft anomalies, sinuses are more frequent than cysts, and fistulae are extremely rare.[11] The III and IV branchial cleft anomalies are rare.[12]

Anomalies that originate from the second or third cleft are in close intimacy with vital neck structures such as the carotid artery, jugular vein and hypoglossal nerve.[13] Branchial cleft cysts are benign; however, superinfection, mass effect, and surgical complications account for morbidity. Surgical excision is the definitive treatment for branchial cleft anomalies.[14,15] The rationale of this retrospective review is to analyzedpresentation and surgical treatment of the patients admitted with branchial cleft anomalies in our setup.

Material and Method:

Study was conducted at department of ENT, Head & Neck Surgery, Dow University of Health Sciences & Civil Hospital, Karachi. Twenty sixpatients of suspicious branchial cleft anomalies of any age and sex were admitted in ENT ward of Civil Hospital, Karachi were included in the study from January 2013 to January 2018. All who were treated elsewhere and having no documentary proof, recurrent or residual diseases after surgery were excluded from the study.

Demographic data, clinical findings of branchial cleft anomalies (site whether right or left side of neck, location on sternocleidomastoid muscle, site of internal and external opening of fistulous tract) & sinogram findings if needed were recorded. All cysts were excisied externally through neck while sinuses and fistulae were excisied with standard step-ladder incision. The extent of neck incision for branchial cyst depends upon its size while number of incisions

Figure. 1: Gender distribution

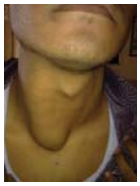

Figure. 2: Front view of Branchial cyst on right side of neck

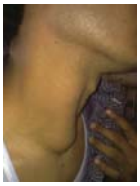

Figure. 2: Side view of Branchial cyst on right side of neck

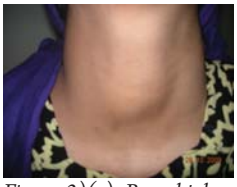

Figure. 3)(a): Branchial sinus

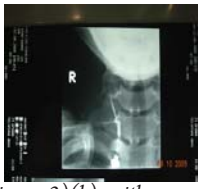

Figure. 3)(b): with sonogram of the same patient (a) on right side of neck

for branchial sinus and fistulae depends upon their vertical length.

Results:

Out of total 26 cases, 17(65.38%) were males and 9(34.61%) were females with male to female ratio of 1.7: 1(Fig 1). The age range was between 15- 35 years with a mean of 19.4 years. On presentation majority of the cases (18) 69.23% were having cervical sinus; 4(15.38%) cases presented with cyst out of which 03 cases were infected with history of incision and drainage. Clear cut fistula were identified in 4(15.38%) cases. Majority 24 cases (92.30%) were second arch defect with cervical sinus or cyst. Location of 4(15.38%) cases presented with branchial cyst was at upper 1/3rd of anterior border of SCM (Fig 2). History of intermittent discharge from sinus located at junction of upper 2/3rd & lower 1/3rd of anterior border of SCM was present in 18(69.23%) cases(Fig 3). Only 2(7.69%) cases have third arch defect with fistulous communication between pyriform fossa and cervical skin. First arch one defect was not identified in present series. All the cases were managed surgically. Cysts with its track excised completely through neck. Sinuses & fistulas were excised with standard stepladder surgical technique. Recurrence was recorded in 02(7.69%) cases on long term follow-up.

Discussion:

95 percent of abnormalities of the branchial cleft apparatus arise from the second cleft. At least 75% of all second branchial cleft abnormaities are cysts,[16] 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.[17]

In our study approximately 81.81% of the patients had branchial sinuses (there was one case of branchial fistulae) and the rest of 18.19% had branchial cyst. These findings are at odd with literature data that show a greater incidence of branchial cysts than branchial sinuses and fistulae. As per the age of presentation most of the patients belonged to the first and second decades (70%). This is in concordance to literature that shows a higher frequency of branchial cysts at adults between decade two and three decades. No gender prediction has been reported.[18] Our data showed more male predominance (male female ratio 1.7: 1).

Vast majority of branchial cleft anomalies are of second cleft type. These may be cystic swelling, sinuses and/or fistula. 60 percent of branchial cysts are located in the upper third of neck, at the anterior margin of sternocleidomastoid muscle.[19] 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 arouse the suspicion.[20,21] They are usually unilateral and very rarely bilateral. They usually present as cystic painless mass, unless infected. The symptom of hoarseness and dyspnoea attributed to palsies of cranial nerves IX, X and XII[22] and a bulge in overlying pharynx respectively.[23] 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.[24] There may be communication with hypopharnyx whereby patient may complain of leakage of fluid diet. Third and fourth branchial sinus and fistulae are rare and may open low in the neck and pyriform fossa.[25,26] Symptomatology in the present study were in accord with reported literature.

Besides complete clinical examination, ultrasound neck and sino fistulograpy may be needed to obtain an accurate diagnosis. In cystic lesions some studies recommend CT scan and MRI for accurate diagnosis and to see the extent of lesion for better planning the surgical exploration beforehand. Simmilarly FNAC of the cystic lesions were suggested by few studies in the literature.[19,21,25,26] In the present study we did not adviced imaging FNAC of cystic lesion as clinical examination, ultrasonography and sinogram were considered sufficient in diagnosing the patients accurately.

Management of second branchial cleft anomalies is surgical excision.[27] Surgery should ideally be performed on the uninfected neck. Previous authors have described the oral approach for the management of a second branchial fistula. Stripping of the fistula was first described by Heanley in 1976, but this requires a correct diagnosis and complete fistula with both internal and external openings, an extremely rare clinical occurrence.[28] Surgical management of second bronchial sinus presents challenge, partly because, unlike the complete branchial fistula, there is difficulty in visualizing the cephalic end of the sinus. The combined approach technique overcomes this limitation by ensuring complete excision of the bronchial sinus tract. With the classical approach, complete excision must be ensured, but this is doubtful in some cases as recurrence rates following complete excision can be as high as 22 percent.[29] Olusesi also described the combined approach branchial sinusectomy which ensures the complete excision of the entire tract of the branchial sinus.[30] We performed complete excisions of fistulous or sinus tracts in 22(84.61%)cases with standard stepladder andcervical skin crease incision. We found no recurrence on 1-5 year follow-up.

Conclusion:

Branchial cleft anomalies are less common and not confined to peadiatric population. Second branchial cleft anomalies are the commonest of the true Branchial cleft abnormalities and the diagnosis is usually straight forward. Amongst second branchial cleft anomalies, sinuses are more frequent than cyst and fistulae are rare. The management of this rare lesion is challenging and requires knowledge of embryological anatomy of the branchial apparatus. With the classical approach complete excision must be ensured.

Conflict of interest: None

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Role and contribution of authors:

Dr Atif Hafeez Siddiqui, conceived, designed, did data collection, data analysis & writing of manuscript

Dr Danish ur Rahim, review and final editing of manuscript

Dr Iqbal A. Muhammad Khayani, literature review, data analysis and final editing of manuscript

Dr Muhammad Umar Farooq, data analysis and final review of manuscript

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