Original Article ISSN (O):2395-2822; ISSN (P):2395-2814

Congenital Anomalies and Anatomical Aberrations in Head & Neck Oncology– A Single Institution Experience.

S. Subbiah[1], G. Gopu[2], Syed Afroze Hussain[3], Bright Singh.R.S[4]

[1]Professor, Centre for Oncology, GRH, Kilpauk Medical College, Chennai, Tamilnadu, India
[2]Associate Professor, Centre for Oncology, GRH, Kilpauk Medical College, Chennai, Tamilnadu, India
[3]Assistant Professor, Centre for Oncology, GRH, Kilpauk Medical College, Chennai, India
[4]Resident, Centre for Oncology, GRH, Kilpauk Medical College, Chennai, India.

Received: February 2018
Accepted: February 2018

ABSTRACT

Background: Congenital anomalies are defined as structural or functional anomalies that occur during intrauterine life. Awareness of such variations will decrease intraoperative dilemma and complications. In this article, we have presented our experience in congenital anomalies and anatomical aberrations in head and neck malignancies. We have reviewed the literature and discussed relevant embryology and their clinical significance. **Methods:** All patients with head and neck malignancies operated at our institution in the past three years, who had congenital anomalies and anatomical aberrations detected during preoperative evaluation or intraoperatively were included in this study. **Results:** Seven types of congenital anomalies were encountered, with most common anomaly being the aberrations in the pattern of facial nerve branches (47.6%). Thyroglossal cyst and non-recurrent laryngeal nerves were the next common anomalies noted (14.2%). Preoperative suspicion and diagnosis were evident in three patients. **Conclusion:** Though rare, congenital anomalies may pose significant challenges to the surgeon. Since most of them are noticed intraoperatively, a thorough knowledge of anatomy and its variation will help prevent injury to these structures. When promptly recognized and appropriately dealt, congenital anomalies do not produce surgical morbidity.

Keywords: Congenital anomalies, Head and Neck Oncology.

INTRODUCTION

Congenital anomalies are also known as birth defects, congenital disorders or congenital malformations. Congenital anomalies can be defined as structural or functional anomalies that occur during intrauterine life and can be identified prenatally, at birth, or sometimes may only be detected later in infancy (WHO fact sheet, updated Sep 2016).

Although approximately 50% of all congenital anomalies cannot be linked to a specific cause, there are some known genetic, environmental and other causes or risk factors. Their spectrum of clinical implication may range from innocuous coexistence, through diagnostic dilemma or therapeutic challenges, to life-threatening defects. Though case reports on congenital anomalies are widely reported in pediatric malignancies, literature is scanty on the prevalence and clinical significance of such anomalies in adult malignancies.[1,2]

Name & Address of Corresponding Author
Dr. S. Subbiah MS., MCh
Professor of Surgical Oncology,
Centre for Oncology,
GRH, Kilpauk Medical College,
Chennai, Tamilnadu,
India.

Aim

In this article, we present our experience in congenital anomalies of head and neck. We have discussed relevant embryology and their clinical significance.

MATERIALS AND METHODS

All patients who underwent surgery for head and neck malignancies at our institution between 2014 and 2017, who had congenital anomalies or anatomical aberrations were included in the study. We have reviewed literature, discussed relevant embryology and their clinical significance.

RESULTS

Seven different types of congenital anomalies were observed among 21 patients (Table 1) with most common anomaly being the aberrations in the terminal branching pattern of facial N (47.6%). Non-recurrent laryngeal nerves and Thyroglossal cyst were the next common anomalies noted (14.2% each). Preoperative suspicion and diagnosis were evident in three patients (Thyroglossal cyst). Surgical challenges posed by these anomalies were

handled effectively by meticulous dissection and hence no anomaly specific morbidity was encountered.

Table 1: Anomaly and aberrations

Anomaly and aberrations	Primary malignancy	Number of cases
Facial nerve	Parotid tumour	10
Non-recurrent laryngeal nerve	Carcinoma thyroid	3
Thyroglossal cyst	Carcinoma thyroid	3
Accessory nerve anomaly	Carcinoma alveolus	2
Retrosternal thyroid	Follicular neoplasm of thyroid	1
Right subclavian artery in neck	Carcinoma tongue	1
Abnormal stylopharyngeus attachment	Carcinoma buccal mucosa	1

DISCUSSION

Table 2: The branching patterns were classified into six types based on the description of Davis et.al[7]

Branching patterns	Percentage of Patients	Description
Type I	13%	No anastomosis occurred between branches of the facial nerve
Type II	20%	Presence of an anastomotic connection between branches of the temporofacial division.
Type III	28%	A single anastomosis between the temporofacial and cervicofacial divisions
Type IV	24%	A combination of type II and type III
Type V	9%	Two anastomotic rami passed from the cervicofacial division to intertwine with the branches of the temporofacial division
Type VI	6%	Plexiform arrangement, the mandibular branch sent a twig to join any members of the temporofacial division

Facial nerve aberrations

Though not strictly considered as congenital anomalies, aberrations in facial nerve branching pattern are quite commonly encountered in surgical practice. Awareness of such variations is essential in anticipating, identifying and preserving them during surgery. There are 23 facial muscles, most of which are paired. In facial expressions, 17 muscles are activated (Freilinger et al., 1990).3 The knowledge of the surgical anatomy of the facial nerve and its correlations with the parotid gland and facial muscles are very important for adequate preservation in the cases of surgery in this area. The iatrogenic injury (21%) is very common.[4] The choice of the surgical approach is very relevant in the parotid surgery because of the extreme anatomic variability of the parotid area and the functional importance of the branches of the facial nerve.[5] The facial nerve emerged from the stylomastoid foramen, which is located immediately dorsolaterally to the base of the styloid process. From there, the facial nerve took a downward and ventrolateral course and entered the retromandibular fossa, which was filled with parotid parenchyma. The branching pattern of the facial nerve varies among individuals. The classification of the peripheral distribution of the facial nerve is based on the type and number of anastomoses between the peripheral branches. McCormack et al.[6] reported on 100 cases, classifying eight types of variations in facial nerve branching. These were arranged in order of increasing complexity, beginning with the simple type and ending with those exhibiting a markedly plexiform arrangement. The classification reported by Davis et al.[7] followed the scheme described by McCormack et al.

We came across 3 out of the 6 described variations in facial nerve branching aberrations (Type: II, III, IV). [Figure 1,2,3] With delicate dissection, branches of facial nerve were preserved in all patients with no postoperative morbidity.

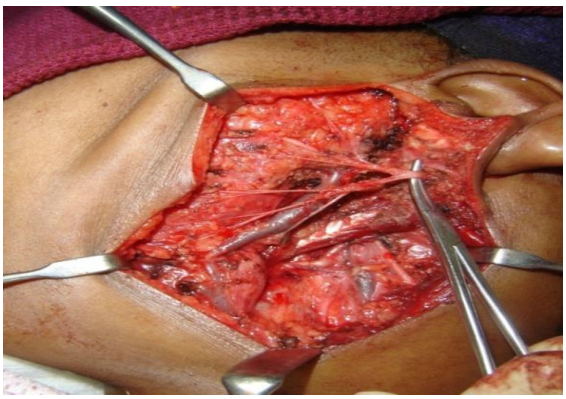

Figure 1: Type II Facial nerve aberration

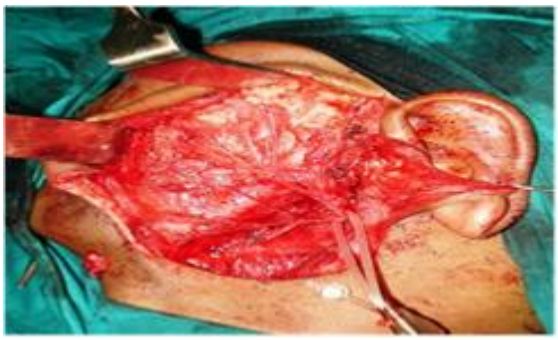

Figure 2: Type III Facial nerve aberration

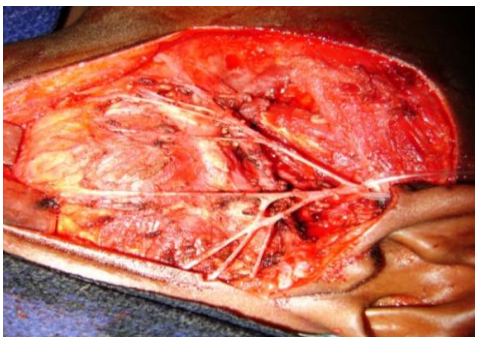

Figure 3: Type IV Facial nerve aberration

Non-Recurrent Laryngeal Nerve (NRLN)

The non-recurrent laryngeal nerve is a rare anomaly, passes transversely into larynx with or without its recurrent branches directly arising from the vagus nerve. Due to deviating from its normal course, the non-recurrent laryngeal nerve is vulnerable to injury during thyroid surgery. The incidence on the right side is 0.3 - 0.8% and extremely rare on the left side 0.004%. When the 4th pair of aortic arch generates the common carotid artery and the right subclavian artery, disappears during embryologic development, the right subclavian artery arises from the right dorsal aorta and absence of innominate artery. With the absence of the normal anatomical position of the right subclavian artery, the right RLN will traverse directly from the vagus nerve before entering the larynx. The left NRLN is associated with situs inversus or loss of ductus arteriosus during the fetal life.[8] Three types can be distinguished. Toniato et al.[9] classified the RLN anomalies into the following: Type A: Non-recurrent laryngeal nerve arising directly from the cervical vagus and coursing along the branches of the superior thyroid peduncle. Type B: Follows a course over and parallel to inferior thyroid artery. Type C: Follows a course parallel to and under or between branches of the inferior thyroid artery [Figure 4] Preoperative diagnosis of NRLN is extremely difficult but may be suspected in situs inversus on a preoperative chest x-ray and in patients with dysphagia lusoria due to an aberrant retro esophageal right subclavian artery. Failure to recognize may result in severe iatrogenic morbidity. An NRLN should be anticipated in the absence of RLN in its usual anatomical position, and a systematic diligent dissection can spare this anomalous vital structure. We found 3 cases of NRLN [Figure 5] in papillary carcinoma thyroid, in which all had type 2A course, identified promptly and preserved. Left RLN had a normal course.

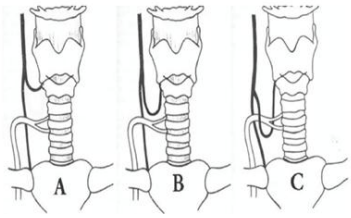

Figure 4: Schematic presentation of Non Recurrent Laryngeal Nerve (NRLN)

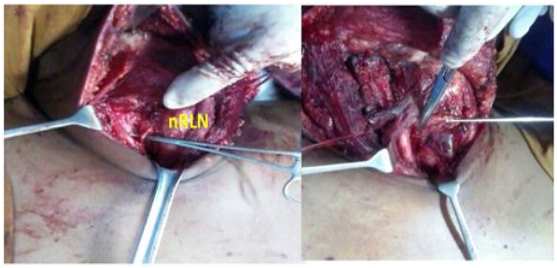

Figure 5: NRLN arising from the vagus nerve at the cervical level

Thyroglossal Duct Cyst (TGDC)

A TGDC is the most common anomaly in the development of the thyroid gland.[10] Seventy percent of midline masses diagnosed during childhood and 7% in adults had TGDCs.[11] Only 1% of thyroid carcinomas arise from a TGDC.[12] Brentano in 1911 and Uchermann in 1915 are credited as being among the first to describe a neoplasm in a thyroglossal duct (TGD) remnant; the median age at presentation is 40 years, and most patients are asymptomatic.[13]

There is still controversy regarding the need to remove the thyroid gland in the case of a papillary carcinoma of the TGDC.[12] Thyroidectomy is recommended in cases where (a) the thyroid gland is multinodular, with a cold nodule in a thyroid iodine uptake scan; (b) enlarged lymph nodes are present, or (c) a history of neck irradiation exists.[14]

There are two theories to explain the thyrogenic origin of TGDC adenocarcinomas. Firstly, the de novo theory is based on the fact that in 62% of cases, ectopic thyroid tissue can be identified histopathologically, and this is supported by the absence of medullary carcinoma in the TGDC as it arises from parafollicular cells. The second is the metastatic theory which suggests that Thyroglossal cyst carcinoma is metastatic from an occult primary thyroid gland, as papillary carcinoma is multifocal.[15] Although Crile believed that the TGDC could act as a natural conduit for the spread of thyroid carcinoma, the metastatic theory seems less likely.[16,17] Mobini et al. conceded that squamous cell carcinoma is probably the only true carcinoma of the TGDC since the other malignancies develop in ectopic thyroid tissue.[18] Thus, squamous cell carcinoma can be considered the only primary thyroglossal cyst tumor, being very rare and having a poor prognosis with a mortality rate of 30–40%.[19]

A previous study proposed an algorithm for treatment of papillary carcinoma in TGDC which involved a simple Sistrunk procedure in patients less than 45 years of age with tumors less than 1.5 cm that are confined to the cyst and who show an ultrasonographically normal thyroid gland and no suspicious lymph nodes. Total thyroidectomy with neck dissection is performed only when lymph node metastases are found on ultrasound or during surgery.[20]

We came across well-differentiated thyroid cancer of TGDC in three patients. Two patients, one each with papillary carcinoma [Figure 6] and follicular neoplasm underwent total thyroidectomy, bilateral paratracheal dissection, and excision of the mass.

Spinal accessory nerve (San)

Relationship of SAN with IJV is variable with an attendant risk of injury during level II nodal dissection. The postoperative morbidity is substantial and impairs the quality of life. Two arrangements of

the nerve about the jugular vein are seen during neck dissections. In most cases (56–90%), the nerve crosses the lateral side of the vein,[21] but in 10–44 % of cases the nerve courses medial to it.[22] On very rare occasions, the nerve may be enclosed by the vein.[23,24] In its medial course, SAN crossed the IJV ventrally in 39.8% of cases, dorsally in 57.4% of cases and passed through the IJV in 2.8% of cases.

We found quite a rare medial relationship of SAN to IJV with ventral coursing in a case of carcinoma tongue [Figure 7]. Awareness of such anomalous relationship is essential to prevent this vital nerve from being injured in the background of altered anatomy.

Figure 6: Thyroglossal duct cyst (TGDC)

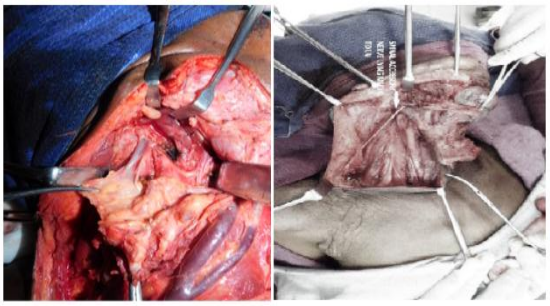

Figure 7: Spinal accessory nerve lying medial to internal jugular vein

Retrosternal Goitre (RG)

Initially, goiter was generically considered as retro-sternal when extended below the thoracic inlet.25 Later, RG was defined by DeSouza and Smith as goiter with a portion of its mass ≥ 50% situated in the mediastinum. However, this definition lacks anatomic precision and can be misinterpreted. More precise definitions of RG have been suggested, namely: a goiter lying two fingerbreadths below the thoracic inlet with the patient in a supine position,[26] a goiter reaching the aortic arch,[27] or the carinae tracheae,[28] a goiter with its inferior pole passing through the cervico-thoracic isthmus below the subclavian vessels. Several classification systems have also been developed to better classify RG. Cohen and Cho divide goiters into four grades, depending on the percentage of goitre mass located in the mediastinum.[29] Huins et al. proposed a classification of RG based on the relationship of goitre with anatomical structures of the mediastinum: they defined three grades of goitre depending upon mediastinal extension, namely, to the level of the aortic arch, to the level of the pericardium or below the level of the right atrium.[30] Four definitions of RG are described: (i) a thyroid in which any part of the gland extends below the thoracic inlet with the patient in the surgical position; (ii) a gland reaching the level of the aortic arch; (iii) a thyroid reaching the level of T4 (on chest x-ray) and (iv) greater than 50% (or ‘‘the majority") of the gland residing below the thoracic inlet. DeSouza and Smith classified retrosternal thyroid as grade I where thyroid located above aortic arch, grade II between aortic arch and pericardium and grade III where thyroid extend below right atrium.[30] We encountered one retrosternal thyroid with follicular carcinoma, which was above the aortic arch (grade I), total thyroidectomy with paratracheal dissection was accomplished comfortably through cervical approach [Figure 8]. Though not an anatomical anomaly, this aberration should not be overlooked since technical implications in surgery differ depending on its grade.

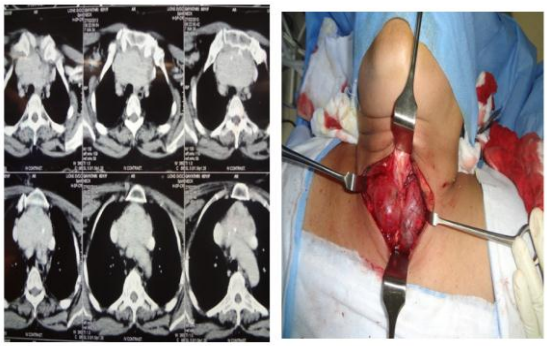

Figure 8: Retrosternal goitre (RG)

High right subclavian artery

Highly placed right subclavian artery in the neck is uncommon in surgical practice. The developmental abnormalities which cause this malformation are unknown. The supra-aortic trunks originate from the aortic sac and from the 6 pairs of aortic arches corresponding to the 6 branchial arches, most of which become partially or completely obliterated near the eighth week of gestation. One possible explanation for a high location of the right subclavian artery could be the persistence of a portion of the proximal segment of the fourth right arch. A high bifurcation of the subclavian artery is associated with changes in the right recurrent nerve, which surrounds the right subclavian artery near its origin Farmery et al. in their study on the ascent of subclavian into the neck found a mean (SD) excursion of the right subclavian artery above the clavicle was 10.4 (11.4) mm. The mean (SD) distance from the cricoid cartilage to the right subclavian artery was 30.6 (14.3) mm, and the data showed a high degree of variance. There was a linear relationship between neck length and cricoid -

subclavian distance. This anomaly might pose lethal complications when injured while attempting techniques like percutaneous tracheostomy.[31] We encountered a high arching right subclavian artery in the neck, 3 cm above the clavicle with the higher take-off of right RLN in the neck [Figure 9].

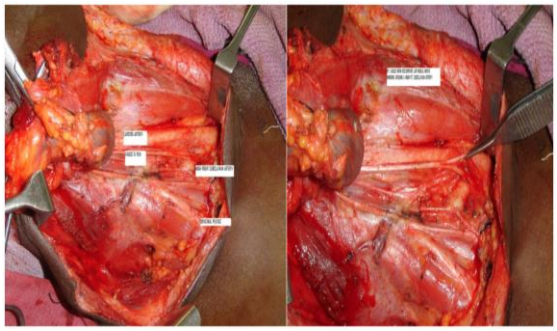

Figure 9: High right subclavian artery

Abnormal stylopharyngeus

The stylopharyngeus is a long, slender muscle, cylindrical above, flattened below. It arises from the medial side of the base of the temporal styloid process, passes downward along the side of the pharynx between the superior pharyngeal constrictor and the middle pharyngeal constrictor, and spreads out beneath the mucous membrane. Some of its fibers are lost in the constrictor muscles while others, joining the palatopharyngeus muscle, are inserted into the posterior border of the thyroid cartilage.[32]

It usually courses deep to posterior belly of digastric muscle. In one of our neck dissections, we noticed anomalous coursing of stylopharyngeus anterior to digastric muscle [Figure 10]. Though it appears innocuous, this might pose a problem for trainees in identifying the posterior belly of digastric muscle which acts as an important landmark for many other vital structures deep to it.

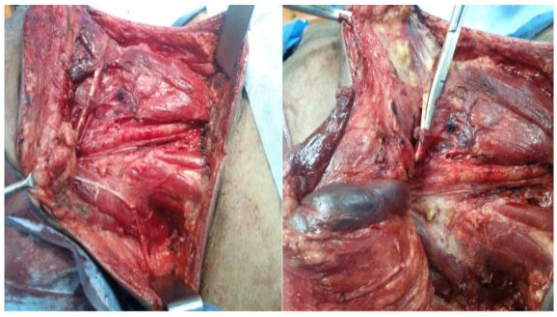

Figure 10: Abnormal stylopharyngeus

CONCLUSION

Though rare, congenital anomalies may pose significant challenges to the surgeon. Since most of them are noticed intraoperatively, a thorough knowledge of anatomy and its variation will help prevent injury to these structures. When promptly recognized and appropriately dealt, congenital anomalies do not produce surgical morbidity.

REFERENCES

1. Patel ZM, Adhia RA. Birth defects surveillance study. Indian J Pediatr 2005 Jun;72(6):489–91.
2. Oosterhuis JW, Looijenga LH. Testicular germ-cell tumours in a broader perspective. Nat Rev Cancer 2005 Mar;5(3):210–22.
3. Freilinger G, Happak W, Burggasser G, Gruber H. Histochemical Mapping and Fiber Size Analysis of Mimic Muscles. Plastic and Reconstructive Surgery. 1990;86(3):422-428.
4. Babademez MA, Acar B, Gunbey E, Karabulut H, Karasen RM. Anomalous relationship of the retromandibular vein to the facial nerve as a potential risk factor for facial nerve injury during parotidectomy. In: Journal of Craniofacial Surgery. Vol 21. ; 2010:801-802. doi:10.1097/SCS.0b013e3181d84027.
5. Rodrigues ADC, Andreo JC, Menezes LDF, Nior RÚ, Anatomy GM. Anatomy of the Facial Nerve and its Implication in the Surgical Procedures. Int J Morphol. 2009;27(1):183-186. doi:10.4067/S0717-95022009000100031.
6. McCormack, L.J., Cauldwell, E.W., Anson BJ. The Surgical Anatomy of the Facial Nerve. Surg Gynecol Obs. 1945;80:620-630.
7. Davis RA, Anson BJ, Budinger JM, Kurth LR. Surgical anatomy of the facial nerve and parotid gland based upon a study of 350 cervicofacial halves. Surg Gynecol Obstet. 1956;102(4):385-412. doi:10.1002/ca.20011.
8. Li X, Wang Z, Lu X, et al. Non-recurrent laryngeal nerve related to thyroid surgery: a report of 5 cases and literature review. Med Sci Monit. 2010;16(6):CS71-5. http://www.becker-hickl.de/handbook.htm.
9. Toniato A, Mazzarotto R, Piotto A, Bernante P, Pagetta C, Pelizzo MR. Identification of the nonrecurrent laryngeal nerve during thyroid surgery: 20-year experience. World J Surg. 2004;28(7):659-661. doi:10.1007/s00268-004-7197-7.
10. Vera-Sempere F, Tur J, Jaén J, Perolada JM, Morera C. Papillary thyroid carcinoma arising in the wall of a thyroglossal duct cyst. Acta Otorhinolaryngol Belg. 1998;52(1):49-54. doi:10.1007/PL00020131.
11. Yang YJ, Haghir S, Wanamaker JR, Powers CN. Diagnosis of papillary carcinoma in a thyroglossal duct cyst by fine-needle aspiration biopsy. Arch Pathol Lab Med. 2000;124(1):139-142. doi:10.1043/0003-9985(2000)124<0139:DOPCIA>2.0.CO;2.
12. Dedivitis RA, Guimarães A V. Papillary thyroid carcinoma in thyroglossal duct cyst. Int Surg. 2013;85(3):198-201. doi:10.1103/PhysRevLett.110.165504.
13. Weiss SD, Orlich CC. Primary papillary carcinoma of a thyroglossal duct cyst: Report of a case and literature review. Br J Surg. 1991;78(1):87-89. doi:10.1002/bjs.1800780127.
14. Kazemi M, Assadi M, Kazemi AA, Ghazvini LA. Primary papillary carcinoma in a thyroglossal duct cyst. Hell J Nucl Med. 2012;9(1):39-40. doi:10.1021/ct200908r.
15. Tew S, Reeve TS, Poole AG, Delbridge L. Papillary thyroid carcinoma arising in thyroglossal duct cysts: incidence and management. Aust N Z J Surg. 1995;65(10):717-718. doi:10.1016/j.jcps.2013.01.002.
16. Crile G. Papillary carcinoma of the thyroid and lateral cervical region; so called lateral aberrant thyroid. Surg Gynecol Obstet. 1947;85(6):757-766. doi:10.1073/pnas.0800367105.
17. Kennedy TL, Whitaker M, Wadih G. Thyroglossal duct carcinoma: a rational approach to management. Laryngoscope. 1998;108(8 Pt 1):1154-1158. doi:10.1177/0300985813511128.
18. Mobini J, Krouse TB, Klinghoffer JF. Squamous cell carcinoma arising in a thyroglossal duct cyst. Am Surg. 1974;40(5):290-294. doi:10.1016/j.virusres.2014.12.019.

19. Boswell WC, Zoller M, Williams JS, Lord SA, Check W. Thyroglossal duct carcinoma. Am Surg. 1994;60(9):650-655. doi:10.1021/cr00101a006.
20. Plaza CPR, López MED, Carrasco CE-G, Meseguer LM, Perucho A de la F. Management of well-differentiated thyroglossal remnant thyroid carcinoma: time to close the debate? Report of five new cases and proposal of a definitive algorithm for treatment. Ann Surg Oncol. 2006;13(5):745-752. doi:10.1245/ASO.2006.05.022.
21. Caliot P, Bousquet V, Midy D, Cabanié P. A contribution to the study of the accessory nerve: surgical implications. Surg Radiol Anat. 1989;11(1):11-15. doi:10.1148/rg.282075064.
22. Soo KC, Hamlyn PJ, Pegington J, Westbury G. Anatomy of the accessory nerve and its cervical contributions in the neck. Head Neck Surg. 1986;9(2):111-115. doi:10.1002/hed.2890090207.
23. Krause HR, Bremerich A, Herrmann M. The innervation of the trapezius muscle in connection with radical neck-dissection: An anatomical study. J Cranio-Maxillofacial Surg. 1991;19(2):87-89. doi:10.1016/S1010-5182(05)80613-4.
24. Gardiner KJ, Irvine BW, Murray a. Anomalous relationship of the spinal accessory nerve to the internal jugular vein. Clin Anat. 2002;15(1):62-63. doi:10.1002/ca.1094.
25. Cougard P, Matet P, Goudet P, et al. [Substernal goiters. 218 operated cases]. Ann Endocrinol (Paris). 1992;53(5-6):230-235. doi:10.1177/019459988509300202.
26. Kanzaki R, Higashiyama M, Oda K, et al. Surgical management of primary intrathoracic goiters. Gen Thorac Cardiovasc Surg. 2012;60(3):171-174. doi:10.1007/s11748-011-0801-5.
27. Vadasz P, Kotsis L. Surgical aspects of 175 mediastinal goiters. Eur J Cardiothorac Surg. 1998;14(4):393-397. doi:10.1016/j.jsb.2003.11.020.
28. Sancho JJ, Kraimps JL, Sanchez-Blanco JM, et al. Increased mortality and morbidity associated with thyroidectomy for intrathoracic goiters reaching the carina tracheae. Arch Surg. 2006;141(1):82-85. doi:10.1001/archsurg.141.1.82.
29. Cohen JP. Substernal goiters and sternotomy. Laryngoscope. 2009;119(4):683-688. doi:10.1002/lary.20102.
30. Huins CT, Georgalas C, Mehrzad H, Tolley NS. A new classification system for retrosternal goitre based on a systematic review of its complications and management. Int J Surg. 2008;6(1):71-76. doi:10.1016/j.ijsu.2007.02.003.
31. Farmery A, Shlugman D, Anslow P. How high do the subclavian arteries ascend into the neck? A population study using magnetic resonance imaging. British Journal of Anaesthesia. 2003;90(4):452-456.
32. McHanwell S. Head and Neck.(2016) In: Standring S, Gleeson Meds. Gray's Anatomy: The Anatomical Basis of Clinical Practice. 41st ed.Elsevier :571-581.

How to cite this article: Subbiah S, Gopu G, Hussain SA, Singh RSB. Congenital Anomalies and Anatomical Aberrations in Head & Neck Oncology– A Single Institution Experience. Ann. Int. Med. Den. Res. 2018; 4(2):MC19-MC24.

Source of Support: Nil, **Conflict of Interest:** None declared