



## CASUISTIC PAPER

Abhishek Bhardwaj  (ABCDEF), Kartikesh Gupta  (ABDEFG), Manu Malhotra <sup>(BDEFG)</sup>,  
Madhu Priya <sup>(DE)</sup>, Mamta Verma <sup>(DE)</sup>

# Triple thyroglossal duct cysts in an adult: a rare case report and review of literature

Department of Otorhinolaryngology & Head-Neck Surgery, All India Institute of Medical Sciences, Rishikesh, India

## ABSTRACT

**Introduction.** Thyroglossal duct cyst (TDC) is the most common cause of congenital neck mass. These can present anywhere from foramen caecum to mediastinum. Usually presents as a solitary cyst, the double thyroglossal cyst is very rare and a triple thyroglossal cyst has never been reported.

**Aim.** Herein, we report an atypical case of triple thyroglossal cyst, at levels of hyoid, thyrohyoid membrane and thyroid isthmus managed surgically without any complication.

**Description of the case.** We are presenting case of a 48-year-old female who presented to us with the complaint of anterior neck swelling since birth. On work up it was diagnosed as a case of the thyroglossal duct cyst and was intraoperatively found to have 3 distinct cystic swellings connected to a common stalk lying beneath the hyoid. It was successfully treated by modified Sistrunk's procedure. There was no evidence of recurrence on follow up for 6 months. Considering atypical presentations, there are 9 cases reported with the double thyroglossal duct cyst, TDC within the thyroid gland and sublingual TDC. Such presentations make the diagnosis more challenging, leading to improper treatment.

**Conclusion.** We are presenting this case as there is no case reported in English literature with a triple thyroglossal duct cyst. An awareness that thyroglossal cyst can present as multiple cysts is important for clinician in order to perform correct surgical management and to avoid the most feared complication of recurrence.

**Keywords.** complication, diagnosis, duct, recurrence, thyroglossal cyst, treatment

## Introduction

Thyroglossal Duct Cyst (TDC) is the most common pathology seen in patients presenting with congenital median neck mass.<sup>1</sup> Majority of these occur infra-hyoid (25-65%) in location.<sup>2</sup> The prevalence of thyroglossal duct cyst is around 7% in general population.<sup>3</sup> The

most common presentation is anterior neck mass which moves with deglutition and protrusion of tongue, but may also have atypical presentations like lateral neck swelling or laryngeal communication. Mean age of presentation is 17.3 years.<sup>4</sup> Aetiology remains through stimulation of epithelial remains in the thyroglossal

**Corresponding author:** Abhishek Bhardwaj, e-mail: abhi04stanley@gmail.com

**Participation of co-authors:** A – Author of the concept and objectives of paper; B – collection of data; C – implementation of research; D – elaborate, analysis and interpretation of data; E – statistical analysis; F – preparation of a manuscript; G – working out the literature; H – obtaining funds

Received: 11.12.2019 | Accepted: 31.01.2020

Publication date: June 2020

duct, which were left in embryonic life, through upper respiratory tract infections.<sup>(5)</sup> Dermoid, lipoma, branchial cyst, lymphadenopathy and benign thyroid lesions are common differential diagnoses.<sup>1</sup> The Sistrunk's Operation is the procedure of choice for TDCs due to its low rate of recurrence.<sup>5,6</sup>

### Aim

Herein, we report an atypical case of triple thyroglossal cyst, at levels of hyoid, thyrohyoid membrane and thyroid isthmus managed surgically without any complication. We feel necessary to bring this rare anomaly which has not yet been reported to the attention of surgeons operating TDCs and hence reporting this case.

### Description of the case

A 48-year-old female presented to the outpatient department with swelling in midline of anterior neck since childhood. Swelling was insidious in onset, gradually progressive, not associated with - discharge, fever, pain and difficulty in swallowing. There was no history suggestive of hypothyroidism, hyperthyroidism or compressive symptoms. There were no other systemic comorbidities. On neck examination, there was 4 cm x 3 cm smooth, well-defined swelling in the anterior aspect of neck, situated between mentum and thyroid cartilage, at & below level of hyoid, moving with deglutition and pro-

trusion of tongue with overlying skin normal (Figure 1). No discharging sinus, ulcer or enlarged lymph nodes were seen. Laryngeal endoscopy was normal.



Fig. 1. Neck examination

Ultrasonography neck was performed which revealed a well-defined heteroechoic multiloculated cystic lesion measuring 40x38x25 mm in anterior neck with internal echo. FNAC was done which showed abundant macrophages with thick, mucinous background and ciliated columnar epithelium suggestive of benign cystic lesion. Contrast enhanced computed tomography neck was done which showed a well-defined multi-lobulated thick walled cystic lesion (HU-10) of size 37x29x35 mm in the infra-hyoid region of anterior neck with at-



Fig. 2. Contrast enhanced computed tomography of neck

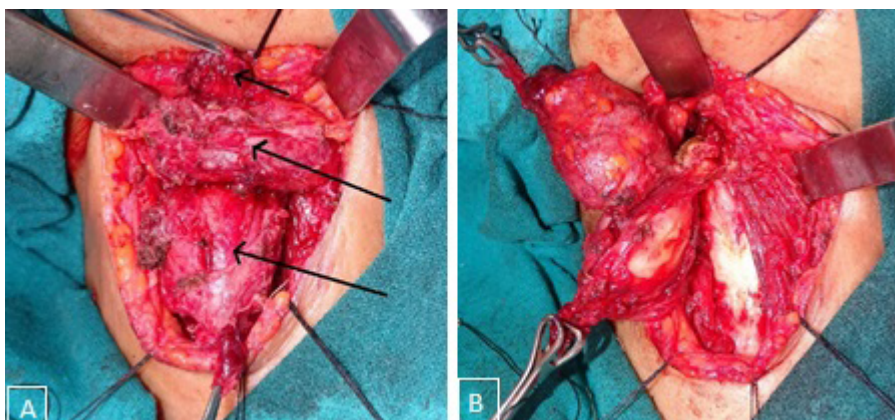


Fig. 3. Three separate cysts during operation

tachment to the isthmus of thyroid gland with few solid components (Figure 2.A, 2.B, 2.C, marked red arrow)

With these findings, the provisional diagnosis of thyroglossal duct cyst was made. Patient was taken up for surgery under general anaesthesia (GA).

Surgery: Sistrunk's operation was done. A transverse incision along the neck crease was given below the inferior border of swelling. Sub-platysmal flap was elevated superiorly till 1 cm above hyoid bone. Strap muscles identified and retracted laterally. Three separate cysts were seen originating from the same tract (Figure 3.A, 3.B)

Inferior most cyst had its attachment to thyroid isthmus. Extracapsular cyst's dissection was done from inferior to superior. A cuff of tissue from thyroid isthmus at its attachment with the inferior most cyst was excised. At the level of hyoid was the superior cyst, the smallest out of three and was excised along with the body of hyoid. Persistent thyroglossal tract was traced superiorly till base of tongue and tied. This was followed by excision of the tract along with all three cysts in Toto. Wound closure was done in two layers. Postoperative period was uneventful. Specimen (figure 4) was sent for histopathology.



Fig. 4. Specimen obtained during surgery

The postop histopathology report showed oval surface with clear mucinous fluid, outer wall lined by pseudostratified epithelium with follicular cells containing colloid and lined by cuboidal epithelium with intense lymphocytic and macrophage infiltration, corresponding with the diagnosis of thyroglossal duct cyst (Figure 5A, 5B, 5C). Patient was followed up for a period of 6 months and was symptom free with no evidence of recurrence. Figure 6A and 6B shows follow up photo of neck wound at postoperative day 10 and at 6 months.

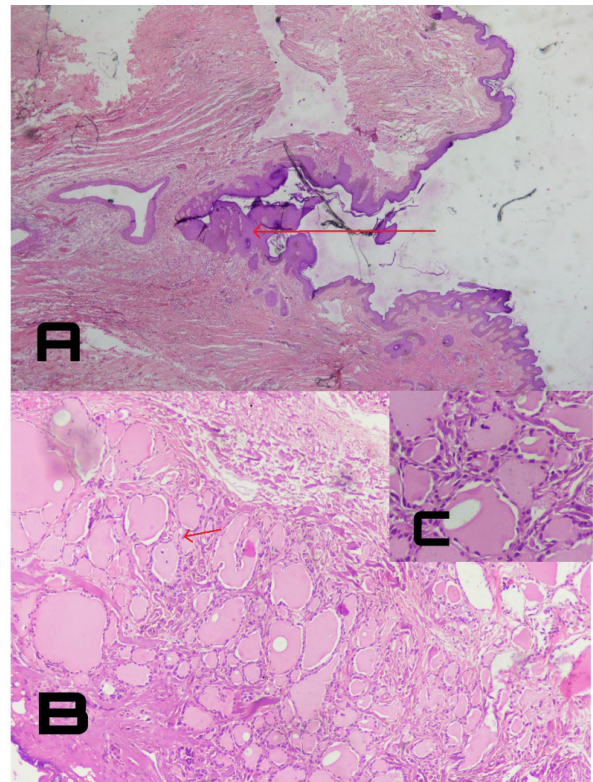


Fig. 5. Thyroglossal duct cyst (H&E stain)



Fig. 6. Photo of neck wound at postoperative day 10 (A) and at 6 months (B)

## Discussion

Neck swelling consists of several entities varying from benign (cysts to cyst-like-masses) to malignant. Cystic masses in neck can be either congenital or acquired. TDCs comprise almost 70% of congenital neck cysts.<sup>1</sup> During development, the anlage of thyroid/parathyroid descends in lower neck from the foramen caecum and passes anterior to hyoid, leaving the epithelial tract behind.<sup>1,7</sup> Thyroglossal duct is a transient connection between descending thyroid and pharyngeal endoderm. Foxe 1 gene is responsible for migration of the thyroid bud from tongue base to its normal location.<sup>7</sup> Generally, this tract degenerates in foetal life around 8th week, if persists leads to development of cysts (most commonly), fistulae or solid nodules. These mostly present in paediatric age groups.<sup>3,8</sup> Clinical features vary from asymptomatic to painless cystic swelling in the anteri-



or aspect of neck (most common).<sup>3</sup> Up to one quarter, these cysts present as the draining sinuses. Atypical presentations like swelling not moving with protrusion of tongue, lateral neck swelling, swelling in the floor of mouth, aberrant pharyngeal communication, tuberculous cyst, dysphagia, severe respiratory distress and sudden infant death syndrome may also be seen rarely, making the diagnosis difficult.<sup>3,4,8</sup> Airway obstruction in intra-laryngeal or para-glottic TDC, though a very rare presentation, caused due to mass effect over hypopharynx and posterior displacement of epiglottis, also described as the ball valve effect between laryngeal inlet and cyst.<sup>4,9</sup> It may also mimic a laryngocele.<sup>10</sup> They can be located from tongue to mediastinum anywhere (majority being infra-hyoid in location, 70%) and even within thyroid, very rarely.<sup>8,11</sup>

Diagnosis is mainly clinical, supported by radiology and confirmed on cytology. Ultrasonography (USG) is done to ensure if a normal thyroid gland is present or not. USG shows an anechoic/hypoechoic/hyperechoic midline mass with internal echoes. In USG, there is no significant vascularity with the majority being unilocular and with pseudo solid appearance (Presence of a solid component suggests the possibility of TDC carcinoma or infection).<sup>20,21</sup> Specific features in USG or suspicion of some other pathology warrants use of CT in a case of TDC. Computed tomography characteristics are of low density mass lesion predominantly, with or without septations, and with peripheral rim enhancement rarely (High density lesion correspond to high protein content either due to a malignancy within or infection).<sup>22</sup> The fascial planes of thyroglossal duct cysts are generally well preserved (abnormal fascial planes demonstrate post-inflammatory changes). As in this case, predominantly suspicion of triple TDC was made on CT only. It becomes especially important because as in our case, even multiple thyroglossal cyst may appear as a solitary lesion clinically. Thyroid scintigraphy is done only if ectopic thyroid is suspected. Next step in management comes for FNAC. Cytology will help in characterization of swelling. Pathologic mimics like dermoid, epidermoid cysts, branchial cleft cyst, laryngocele, thymic cysts, lymphatic malformation, ectopic thyroid and necrotic lymph nodes, can be mistaken as TDC.<sup>23</sup> Pathologic features include a respiratory or squamous epithelium lining in the cyst wall, muco-serous glands with inflammatory changes. Primary treatment is surgical excision. Sistrunk's procedure is treatment of choice. From our search, alternate techniques for TDC treatment includes simple cystectomy, Schlange procedure and sclerotherapy (using OK-432 or ethanol). Over a period of time, Sistrunk's operation has been modified than its conventional procedure. Sistrunk's 1920 description of TDC excision defines excision of the ¼ inch of central portion of hyoid

along with the cyst and a cuff of soft tissue of same diameter up to and including foramen caecum, followed by repairing the opening in the oral cavity and approximating strap muscles and hyoid. In 1928 description, he no longer advocates dissection in the oral cavity with a success rate of 96%.<sup>6</sup> With time, there have been modifications like (1) coring out the central portion of hyoid using 4.5 mm skin punches and then resecting out under magnification of a 3.5x loupe (2) hyoid cartilage division method.<sup>5,24</sup> Results are seen better including amount of drainage and operation time, in cases where hyoid was divided at points of non-fusion of the cartilaginous portion of hyoid compared to conventional hyoid bone cutting.<sup>5,6,24</sup> Extended Sistrunk's operation is being advocated for cases where there is a recurrence following Sistrunk's procedure. Regarding TDC excision, there are two surgical approaches (1) cervical approach - Sistrunk's procedure, most commonly used (2) trans-oral endoscopic approach used for lingual or sublingual TDCs. However, there is no data regarding the efficacy of trans-oral approach. Complication of Sistrunk's procedure included local wound infection (most common), seroma formation, hematoma, salivary fistulae, airway stenosis and rarely hypothyroidism.<sup>3</sup> Our literature search shows that Sistrunk's procedure has significantly brought down the recurrence rate. Schlange procedure which is rapid and easier to perform is simple cystectomy with removal of the body of hyoid is associated with around 30 % of recurrence. Sclerotherapy acts by the reduction of lymphatic fluid and shrinkage through the fibrosis but not advocated much due to the presence of another very effective procedure such as Sistrunk's operation and less research regarding its efficacy and outcome. Infection is a common complication seen in patients with TDC due to its anatomical proximity with oral cavity, leading frequent visits to the hospital. Common pathogens include *Haemophilus* and *Staphylococcus*. A major problem for management of TDCs is recurrence even in hands of a skilled surgeon, which makes it a challenge for treatment. Maximum recurrence was seen with simple cystectomy and was more common in younger age groups and patient who had a previous history of recurrent infection in TDC, rupture of cyst intraoperatively and skin involvement.<sup>3</sup> Efficacy of postoperative use of drain placement and prophylactic antibiotics is insignificant.<sup>25</sup> Malignant changes in TDCs are rare (less than 1%) and diagnosed either as incidental finding in postop histopathology (73%) or in preoperative FNAC.<sup>26</sup> Most common of TDCs malignancy were papillary carcinoma followed by squamous variety. Management of TDCs malignancy remains same as original procedure for TDCs (Sistrunk's procedure).

Double TDCs may present as cystic swellings nearby or far away from each other. On literature search,

(using PubMed, Embase, Google scholar, Cochrane review) there are 9 cases reported of double TDCs but none for triple TDC.<sup>11-19</sup> Sarmiento et al described a patient with double TDC in floor of mouth on geniohyoid muscles and sublingual region.<sup>16</sup> Yorancilar et al mentioned a double TDC in hyoid and base of tongue.<sup>15</sup> Pueyo et al described a double TDC with one part intrathyroidal and inferred that failure of involution of TDC remains is responsible for cyst development. Yildiz et al in a review mentioned that double TDCs are rare and thyroid scintigraphy is must in such cases to differentiate it from thyroid pathologies. However, a classical Sistrunk operation suffice as treatment for double TDC. Valentino et al mentioned that ultrasonography is must in cases where suspicion of double TDC is suspected. However, it is not sensitive enough to detect all.

Despite a lot of literature being available on thyroglossal duct cyst, there is limited data on the outcome of atypical presentations of thyroglossal duct cyst. Further research needs to be done regarding the cause of such atypical presentations and their effect on management of disease.

## Conclusion

Thyroglossal duct cyst, though a very common entity, may have atypical presentations as double or triple cyst. Failure to identify such variation may lead to inadequate surgery and leaving behind residual disease. Hence, pre-operative radiological as well as intra-operative assessment is important in terms of complete excision and prevention of recurrence.

## Compliance with ethical standards

The study has not received funding from any organization or institution and does not involve any potential conflict of interest (financial and non-financial). Procedures performed in the study was in accordance with the ethical standards of the institution and with the 1964 Helsinki declaration and its later amendments.

## References

- Imhof H, Czerny C, Hormann M, Krestan C. Tumors and tumor-like lesions of the neck: from childhood to adult. *Eur Radiol Suppl.* 2004;14(4):L155-L165.
- Mittal M, Malik A, Sureka B, Thukral B. Cystic masses of neck: A pictorial review. *Indian J Radiol Imaging.* 2012;22(4):334.
- Gioacchini FM, Alicandri-Ciufelli M, Kaleci S, Magliulo G, Presutti L, Re M. Clinical presentation and treatment outcomes of thyroglossal duct cysts: a systematic review. *Int J Oral Maxillofac Surg.* 2015;44(1):119-126.
- Thabet H, Gaafar A, Nour Y. Thyroglossal duct cyst: Variable presentations. *Egypt J Ear Nose Throat Allied Sci.* 2011;12(1):13-20.
- Naik SM, Kumar BY, Ravishankara S, Shashikumar T, Deekshith R. Modified Sistrunk Procedure: A Novel Method of Hyoid Resection using Skin Punches in Subhyoid Thyroglossal Cysts. *Int J Otorhinolaryngol Clin.* 2016;8(3):97-100.
- Balfour AM, Al-Reefy HM, Dilkes MG. Sistrunk's 1920 description of thyroglossal cyst excision. *Grand Rounds.* 2006;6:L1-L2.
- Nilsson M, Fagman H. Development of the thyroid gland. *Development.* 2017;144(12):2123-2140.
- Narayana Moorthy S, Arcot R. Thyroglossal Duct Cyst—More Than Just an Embryological Remnant. *Indian J Surg.* 2011;73(1):28-31.
- Ng ACW, Yuen HW, Huang XY. Atypical thyroglossal duct cyst with intra-laryngeal and para-glottic extension. *Am J Otolaryngol.* 2019;40(4):601-604.
- Booth R, Tilak AM, Mukherjee S, Daniero J. Thyroglossal duct cyst masquerading as a laryngocele. *BMJ Case Rep.* 2019;12(3):e228319.
- Khadivi E, Ardekani HP. Double Thyroglossal Duct Cyst Derived from a Single Tract: A Rare Presentation. *Iran J Otorhinolaryngol.* 2010;22(60):103-106.
- Yıldız T, İlçe HT, Küçük A, İlçe Z. A Rare Form of Thyroglossal Duct Cyst: Double Thyroglossal Cyst and a Review of the Literature. *J Turgut Ozal Med Cent.* 2014;21(2):148-150.
- Lee DH, Yoon TM, Lee JK, Lim SC. Double Thyroglossal Duct Cysts in an Adult: *J Craniofac Surg.* 2017;28(1):e90-e91.
- Pueyo C, Royo Y, Maldonado J, et al. Double cervical cyst derived from a single thyroglossal duct tract. *J Pediatr Surg.* 2008;43(4):748-750.
- Yorgancilar E. Double thyroglossal duct cyst located in the hyoid region and the tongue base: an unusual coexistence. *Turk J Ear Nose Throat.* 2011;21(2):106-109.
- Sarmiento DJ de S, Araújo PPT, da Silveira ÉJD, Germano AR. Double Thyroglossal Duct Cyst Involving the Floor of the Mouth and Sublingual Gland Region: *J Craniofac Surg.* 2013;24(2):e116-e119.
- Bozan N, Sakin YF, Bozkus F, Kanmaz A. A Thyroglossal Cyst With Double Duct Seen in an Adult Patient: *J Craniofac Surg.* 2016;27(4):e414-e415.
- Banchini G, Ghinelli C, De Angelis G, Butturini A, Balestrazzi P. Double cysts of the thyroglossal duct. Description of a case. *Acta Biomed Ateneo Parm.* 1981;52(1):71-74.
- Fu Y, She C, Zhang Q. Thyroglossal cyst of double cyst: one case report. *J Clin Otorhinolaryngol Head Neck Surg.* 2014;28(5):343-344.
- Ahuja AT, King AD, King W, Metreweli C. Thyroglossal Duct Cysts: Sonographic Appearances in Adults. *Am J Neuroradiol.* 1999;20(4):579-582.
- LaPlante JK, Pierson NS, Hedlund GL. Common Pediatric Head and Neck Congenital/Developmental Anomalies. *Radiol Clin North Am.* 2015;53(1):181-196.

22. Reede DL, Bergeron RT, Som PM. CT of thyroglossal duct cysts. *Radiology*. 1985;157(1):121-125.
23. Patel S, Bhatt AA. Thyroglossal duct pathology and mimics. *Insights Imaging*. 2019;10(1):12.
24. Ryu Y-J, Kim DW, Jeon HW, Chang H, Sung MW, Hah JH. Modified Sistrunk operation: New concept for management of thyroglossal duct cyst. *Int J Pediatr Otorhinolaryngol*. 2015;79(6):812-816.
25. Danau T, Verfaillie G, Gordts F, Rose T, De Backer A. Thyroglossal duct cysts in children: a 30-year survey with emphasis on clinical presentation, surgical treatment, and outcome. *Acta Chir Belg*. 2018;119(6):357-362.
26. Rayess HM, Monk I, Svider PE, Gupta A, Raza SN, Lin H-S. Thyroglossal Duct Cyst Carcinoma: A Systematic Review of Clinical Features and Outcomes. *Otolaryngol-Head Neck Surg*. 2017;156(5):794-802.