Acta Scientific Otolaryngology (ISSN: 2582-5550)

Volume 6 Issue 8 August 2024

Case Report

Discover the Uncommon: A Case Report and Systematic Review of a Neck and Mediastinum Tumor in the Elderly

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Received: April 27, 2024

Published: July 31, 2024

Abstract

Branchial cleft cysts are growths that may develop abnormally from the branchial apparatus, a structure that forms the embryonic precursors for the neck tissue. Although these growths are commonly observed in children, they can also affect adults. However, there is limited information available on the adult population that undergoes branchial cleft excision. The cervical area, located anterior to the sternomastoid muscle in the upper or middle portion of the neck, is the most common anatomical site for the appearance of branchial cleft cysts. It is worth noting that mediastinal branchial cleft cysts are extremely rare and only a few cases are reported. Misdiagnosis of the fourth branchial cleft anomaly is widespread and often leads to inappropriate treatment.

We present a case of a 67-year-old woman who had a cystic mass situated below her left thyroid lobe and posterior to the sternum. The patient had painless swelling for the past seven months. Imaging tests revealed a cystic between the thyroid and the aortic arch. The patient underwent complete excision of the lesion through a neck dissection and a sternotomy, and the postoperative histopathological examination confirmed the diagnosis of the branchial cleft cyst. The patient had a successful recovery without any recurrence at the six-month follow-up.

The course of the sinus track indicates that this was a fourth branchial cleft cyst. These are the rarest among the branchial anomalies, and the extension below the peri-thyroid region to the mediastinum is infrequently described. To prevent misdiagnosis and ensure proper treatment, a systematic literature review (PubMed, Cochrane, ResearchGate) was conducted in English and Spanish to summarize the clinical features of the fourth branchial cleft cyst and identify the best options for diagnosis and treatment.

Keywords: Fourth Branchial Cleft Cyst; Branchial Anomalies; Mediastinal Cyst

Introduction

Cystic masses found in the neck can be caused by a variety of different conditions. Among these, branchial cleft anomalies are typically found in children and account for about 20% of head and neck lesions in this age group [1,2]. These anomalies occur from the first to the third brachial arch during embryonic development and can result in tissue developing incorrectly on one or both sides of the neck [3]. While little is known about the demographics or perioperative outcomes in the adult population, these masses are usually cancerous, either primary or metastatic [4]. Lateral cystic neck masses are often attributed to branchial cleft anomalies, but other cystic, infectious, or reactive processes can also cause them [2,5,6]. While biopsy can help distinguish between these causes, complete removal is often recommended to rule out the possibility of cancer [7].

Fourth branchial cleft cysts are a rare form of branchial cleft anomalies, first described in 1973 [8,9], with a prevalence of 1 to

4% of cases [10,11]. They are typically situated in the thyroid gland and mediastinum and are present in early childhood, often following a recurrent abscess or preceding thyroiditis.

The sinus tract of these cysts runs from the apex of the piriformis sinus medial and inferior to the recurrent laryngeal nerve, forms a loop superior to the hypoglossal nerve, parallels the course of the recurrent laryngeal nerve within the trachea-esophageal groove, and envelops the aorta. arch on the left or subclavian artery on the right. It then runs superiorly, dorsal to the common carotid artery, surrounds the hypoglossal nerve and extends medially to the sternocleidomastoid muscle [11,12].

Anomalies in the fourth branchial area are usually seen on the left side of the neck [13-15]. The reason for this is not yet clear, but it may be related to differences in the development of blood vessels on the left and right fourth arch [13,14]. During normal embryological development, the fourth arch artery on the left side becomes part of the aortic arch, while the fourth arch artery on the right side becomes the proximal part of the right subclavian artery [16,17]. However, the fact that ultimobranchial bodies generally develop more on the left side in most mammalian species may also be a contributing factor, although the reason for this remains unknown [17,18].

Due to their rarity, there are no established procedures for managing fourth branchial cleft cysts. Surgical excision (open neck surgery) is the definitive treatment because this ensures the best result [19,20].

Case

67-year-old women presented to clinic with a left-sided neck swelling that had been present for six months, she had gradually development dysphagia, odynophagia, and throat pain. She did not have a fever, weight loss or breathing difficulties. Her medical history included hypertension and diabetes. Physical examination revealed a single cystic swelling on the left side of her neck that was not warm but painful to the touch. The swelling had a smooth surface, was pulsating and moved with swallowing but not with the protrusion of the tongue.

Doctors explored her oral cavity, oropharynx and larynx but found no lesions. Nasal endoscopy and video laryngoscope revealed no abnormalities, and there was no lymphadenopathy in the neck. The neck swelling was palpated as fluctuating and pulsatile, so fine needle aspiration was not performed. A subsequent computed tomography (CT) scan reported a 3.5 cm × 2.2 cm × 4.6 cm left border-enhancing thyroid mass (Figure 1A,B,C).

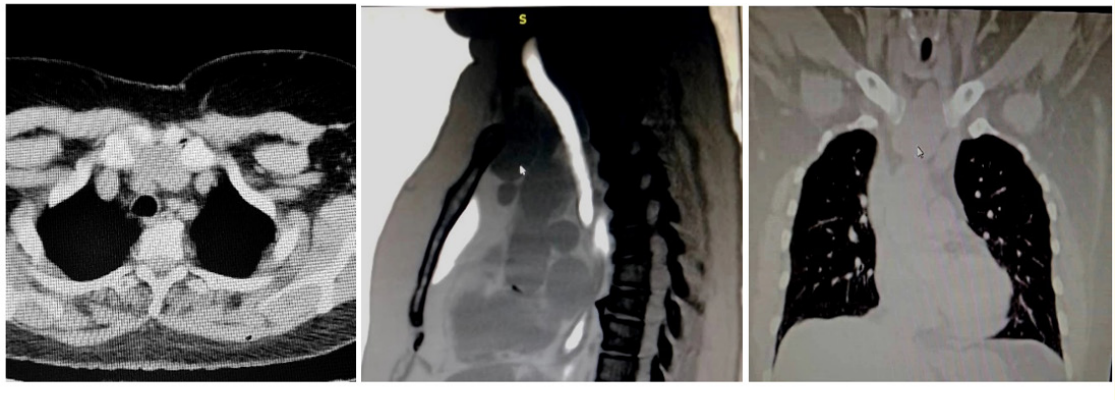

Figure 1: A. reveal an oval tumor in axial view on the left side upper mediastinum, B. sagittal and C. coronal view. The tumor has soft tissue density and extends from the thoracic opening above the clavicles to the midline prevascular space. It measures 55 by 38 millimeters and shows a plane of fat division with respect to the vascular and thyroid structures.

An MRI study was done, and imagens show suggestive changes of fourth brachial cyst anomaly (Figure 2A, B.).

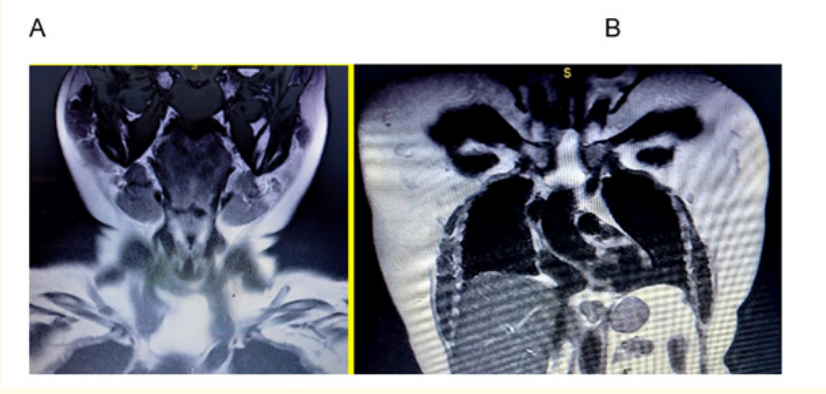

Figure 2: A-B- MRI of the neck and chest showing in coronal view, a cystic tumor located at the anterior cervicothoracic region extending from the lower edge of the left thyroid gland to the anterior mediastinum up to the aortic arch. There is no infiltration into vascular structures or adjacent organs.

During the surgical procedure (open neck dissection and a medial sternotomy) under general anesthesia, a tumor was discovered that was enclosed in a capsule. The tumor had originated in the lower part of the left thyroid lobe and had traveled behind the sternum before attaching to the carotid arch. The posterior plane of the capsule rested on the carotid arch. The surgical team carefully dissected the neck and performed a medial sternotomy to access and remove the tumor. The unicystic lesion had a smooth surface, fixed position, irregular edges, soft consistency, and a cystic appearance. It was dark brown in color with adhesions and soft areas and did not invade surrounding tissues. The tumor measured 5.7 x 1.7 x 0.9 cm, was completely removed, and contained 100 cc of mucoid, chocolate colored cyst fluid.

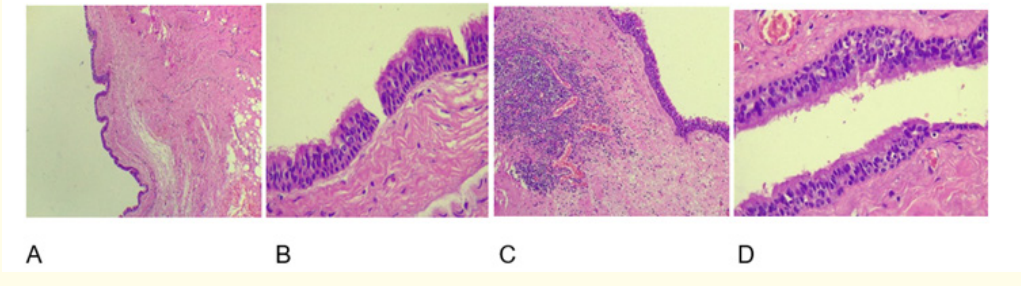

Figure 3

After pathological examination of the surgical specimen, the tumor was determined to be a brachial cyst, and lymphoid accumulations and fibrosis were found in the cyst wall. Additionally, vascular ectasia was observed, but there was no evidence of malignancy (Figure 3A, B, C, D).

The cystic wall shows fibrosis, ectasic vessels and lymphoid follicles, lined by ciliated stratified columnar epithelium. Other elements such as thyroid tissue, sebaceous or mucinous glands and/or cartilage are not observed (Hematoxylin-eosin stain, x40(A), x400(B, C, D).

The patient underwent a successful excision of the cyst and was discharged the day after surgery. At follow-up six months later showed no signs of recurrence (Figure 4).

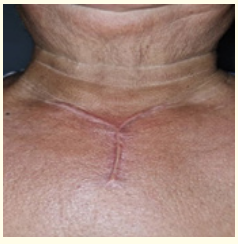

Figure 4: The photo shows the appearance of the scar after 6 months of follow-up.

Discussion

Fourth branchial cysts not only occur in newborns and young children but should also be considered in adults and the elderly. In fact, the condition may be asymptomatic for years before the typical presentation of recurrent neck infections and abscesses appears. In older adults with this presentation, it is important to exclude metastatic lymphadenopathy, lymphoma, or tuberculosis [21].

The cause of branchial cleft cysts is a topic of debate. There are four main theories about their origin. These theories include incomplete closure of the branchial mucosa, the persistence of remnants of the precervical sinus, the origin of the thymopharyngeal duct, and the origin of cystic lymph nodes [22,23]. The branchial apparatus starts to form in the second week of fetal life and is completed in the sixth or seventh week. Ascherson proposed in 1832 that the branchial cleft cyst develops from incomplete closure of the branchial cleft mucosa, which remains inactive until later in life, leading to cyst formation. Specifically, the lack of degeneration of the cervical sinus due to the growth of the second arch over the third and fourth arches is suggested as the cause.

The third and fourth arches, which are covered by the second arch, persist as small pockets with their ectodermal epithelium. These pouches usually fill during fetal development. However, if they fail to do so, cysts, sinuses, and fistulas may develop later. Another theory suggests that branchial fistulas are remnants of the cervical sinus rather than pharyngeal clefts or pouches [24]. In line with the branchial theory, the precervical sinus theory was extended to encompass lateral cervical cysts. Wenglowski proposed that cystic degeneration of the cervical lymph nodes led to the formation of lateral cervical cysts. He also suggested that incomplete closure of the thymo-pharyngeal duct resulted in a lateral cervical cyst [19]. Bhaskar and Bernier proposed that cyst disruption in cervical lymph nodes is caused by trapped epithelium. They identified three possible sources of these epithelial inclusions: brachial cleft, pharyngeal pouch, and parotid gland. Currently, immunohistochemistry is used to study the origin of these cysts at a molecular level, clarifying the nature of epithelial cells and their relationship with other types of epitheliums in the body [22,25].

Multiple diagnostic studies have been proposed as the gold standard for establishing the diagnosis of fourth branchial cysts. Fine needle aspiration cytology (FNAC), Ultrasound (US), computed tomography (CT), and magnetic resonance imaging (MRI) are useful diagnostic tools of choice and provide accurate information about the anomaly. They show the nature of the cyst and the anatomical relationship of the lesion.

However, CT was our radiological examination of choice. It was useful in identifying the tract and its relationship to surrounding structures, particularly the thyroid gland and the aortic arch. MRI was useful to recognize the characteristics of a fourth gill remnant.

The diagnosis of branchial cleft cysts is based on pathological examination. Fine needle aspiration cytology (FNAC) is useful for preoperative pathological diagnosis and can help exclude metastatic or inflammatory diseases, which will effectively avoid unnecessary surgeries. The cytological criteria for the diagnosis of branchial cleft cysts are the presence of nuclear keratinocytes and squamous epithelial cells of various degrees of maturity, as well as lymphoid aggregates in subepithelial lymphoid tissue structures [6].

Surgery is the main treatment for branchial cleft cysts and complete resection of the lesions may reduce the likelihood of postoperative recurrence. For branchial cleft cysts in the neck, a wide horizontally oriented incision is essential to fully expose the lesion and avoid intraoperative injury, and inappropriate surgical maneuvers are likely to lead to recurrence. For branchial cleft cysts in the mediastinum, sternotomy with direct visualization or video-assisted thoracoscopic surgery is recommended.

Conclusion

Branchial cleft cyst is a rare congenital neck disease caused by abnormal degeneration of the branchial organs, that can affect both children and adults. It can also occur in the anterior medias-

tinum. Radiological examinations such as CT and MRI, as well as ultrasound guided FNA, can aid in the preoperative diagnosis. Surgery is the most effective method for treating this disease.

We present a rare case of a fourth branchial cleft cyst in an elderly patient, emphasizing the importance of early diagnosis. Understanding the complex anatomy, embryology, and clinical presentation can help create a comprehensive list of possible conditions and initiate appropriate initial investigations, such as CT and MRI. These diagnostic tools simplify the diagnosis and facilitate surgical removal of the cyst for a definitive diagnosis.

Therefore, it is critical to consider branchial cleft abnormalities as a potential cause of cystic neck tumors in adults. Early intervention can prevent complications and ensure successful treatment.

Conflict of Interests

None.

Financial Support

The author denies public or private support.

Ethical Approval

The present case was carried out following the principles of the Declaration of Helsinki.

Author Contributions

All authors contributed to the article and approved the submitted version.

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Bibliography

1. Guldfred LA., *et al*. "Branchial cleft anomalies: Accuracy of preoperative diagnosis, clinical presentation, and management". *The Journal of Laryngology and Otology* 126 (2012): 598-604.

2. Goff CJ., *et al*. "Current management of congenital branchial cleft cysts, sinuses, and fistulae". *Current Opinion in Otolaryngology and Head and Neck Surgery* 20 (2012): 533-539.

3. Agaton-Bonilla FC., *et al*. "Diagnosis, and treatment of branchial cleft cysts and fistulae: a retrospective study of 183 patients". *International Journal of Oral and Maxillofacial Surgery* 25 (1996): 449-452.

4. Pynnonen MA., *et al*. "Clinical practice guideline: Evaluation of the neck mass in adults". *Otolaryngology–Head and Neck Surgery* 157 (2017): S1-S30.

5. Sira J and Makura ZGG. "Differential diagnosis of cystic neck lesions". *Annals of Otology, Rhinology and Laryngology* 120 (2001): 409-413.

6. Goldenberg D., *et al*. "Cystic metastasis from head and neck squamous cell cancer: A distinct disease variant?" *Head Neck* 28 (2006): 633- 638.

7. Robitschek J., *et al*. "Diagnostic efficacy of surgeon- performed ultrasound-guided fine needle aspiration: A randomized controlled trial". *Otolaryngology–Head and Neck Surgery* 142 (2010): 306-309.

8. Tucker NM., *et al*. "Fourth branchial cleft remnant". *Transactions - American Academy of Ophthalmology and Otolaryngology* 77 (1973): 368-71.

9. Garrel R., *et al*. "Fourth branchial pouch sinus: from diagnosis to treatment". *Otolaryngology–Head and Neck Surgery* 134 (2006): 157-163.

10. Nicollas R., *et al*. "Congenital cysts, and fistulas of the neck". *International Journal of Pediatric Otorhinolaryngology* 55 (2000): 117-124.

11. Franciosi JP., *et al*. "Pyriform sinus malformations: a cadaveric representation". *Journal of Pediatric Surgery* 37 (2002): 533-538.

12. Meng F., *et al*. "A unique location of branchial cleft cyst: case report and review of the literature". *International Journal of Oral and Maxillofacial Surgery* 48.6 (2019): 712-715.

13. Andrieu-Guitrancourt J., *et al*. "Recurrent lateral cervical suppuration Role of fistulae and cysts of fourth branchial pouch". *Annals Oto-Laryngology (Paris)* 105 (1988): 189-192.

14. Rosenfeld RM and Biller HF. "Fourth branchial pouch sinus: diagnosis and treatment". *Otolaryngology–Head and Neck Surgery* 105 (1991): 44-50.

15. Rea Hartley Bailey. "Third and fourth branchial pouch anomalies". *The Journal of Laryngology and Otology* 118 (2004): 19-24.

16. Wang HK., *et al*. "Imaging studies of pyriform sinus fistula". *Pediatric Radiology* 33 (2003): 328-33.

17. Chaudhary N., *et al*. "Fistula of the fourth branchial pouch". *American Journal of Otolaryngology* 24 (2003): 250-252.

18. Saxén L and Toivonen S. "The development of the ultimobranchial body in Xenopus Laevis Daudin and its relation to the thyroid gland and epithelial bodies". *Journals.biologist.com Development* 3 (1955): 376-384.

19. Nicoucar K., *et al*. "Management of congenital fourth branchial arch anomalies: a review and analysis of published cases". *Journal of Pediatric Surgery* 44 (2009): 1432-1439.

20. Annelien Boonen., *et al*. "Fourth Branchial Anomalies: Diagnosis, Treatment, and Long-Term Outcome". *Frontiers in Surgery* 8 (2021): 748351.

21. McClure MJ., *et al*. "Late presentation of brachial cyst". *Ulster Medical Journal* 67 (1998): 129-131.

22. Arndal H and Bonding P. "First branchial cleft anomaly". *Clinical Otolaryngology and Allied Sciences* 21 (1996): 203-207.

23. Glosser JW., *et al*. "Branchial cleft or cervical lymphoepithelial cysts: Etiology and management". *Journal of the American Dental Association* 134 (2003): 81-86.

24. Frazer JE. "The nomenclature of diseased states caused by Certain investigial structures in the neck". *British Journal of Surgery* 11 (1923): 131-136.

25. Little JW and Rickles NH. "The histogenesis of the brachial cyst". *American Journal of Pathology* 50 (1967): 533-547.