

A Case of Fourth Branchial Cleft Anomaly

F.El Mourabit*, G Hamza, M.Loudghiri, W.Bijou, Y.Oukessou, S.Rouadi, R.Abada, M.Roubal and M.Mahtar

Department of Otolaryngology Head and Neck Surgery, University hospital Ibn Rochd, Casablanca, Morocco

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***Corresponding author:** F. El Mourabit, Department of Otolaryngology Head and Neck Surgery, University hospital Ibn Rochd, Casablanca, Morocco.

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ABSTRACT

A rare source of lateral neck masses of congenital origin are branchial abnormalities, which arise from aberrant development during embryogenesis. The most frequent source of origin is the second branchial cleft; anomalies resulting from the first, third, and fourth clefts are less common. Even though branchial cleft-derived cysts are rare, it's crucial to take this condition into account when making a differential diagnosis for neck masses, especially those that are laterally situated. This article presents the rare case of a child of 6 years who presented the sudden appearance of a lateral collection in the neck fistulized to the skin with notion of recurrent neck infections at the same site. patient underwent extensive diagnostic examinations, including radiology, which were consistent with a left subcutaneous collection measuring 17.2*15 mm with irregular, heterogeneous and hypoechoic contours. A 5 centimeter sinus that descends to the thoracic orifice from the internal jugular vein and the common carotid artery was discovered during the cervicotomy. After resectioning it, each sinus extremity had 3.0 resorbable sutures placed. To guarantee complete closure of the sinus, a piece of sternocleidomastoid muscle was inserted. Nine months following the surgery, there were no signs of a neck infection or purulent episode. This clinical example highlights how critical it is to identify uncommon illnesses like branchial cleft cysts as soon as possible and treat them appropriately.

Keywords: Branchial apparatus; Branchial cyst; Branchial cleft anomaly

Introduction

During the fourth week of pregnancy, the gill apparatus, also known as the branchial arches, which are made up of endodermal pouches and ectodermal clefts, aid in the correct development of the head and neck. Incomplete obliteration causes congenital malformations of the ectodermal clefts of the branchial arches, which in most cases (75%) culminate in a cyst and in 25% in a sinus¹.

Roughly 17% of all pediatric neck masses are abnormalities related to branchial clefts². These are typical congenital lesions

that are typically identified in the early years of life in children³. A cyst, sinus, or fistula may occur as a result of a branchial apparatus failing to involute⁴. Less than 1% of branchial anomalies are fourth branchial arch anomalies, which primarily affect the left side and manifest as suppurative thyroiditis or recurrent neck infections⁵.

Cysts with a fourth branchial cleft parallel to the recurrent laryngeal nerve are extremely rare. Like third branchial cleft sinuses, they are most frequently found on the left side (80%), and they typically form a sinus that extends from the apex of

the piriform sinus. However, instead of passing superiorly to reach the anterior left upper thyroid lobe, they travel inferiorly. Cysts can occur anywhere in the neck, all the way down to the mediastinum, but they are typically found next to the thyroid gland. It is challenging to differentiate radiologically between anomalies involving the third and fourth branchial clefts due to their close closeness. The link between the sinus tract and the superior laryngeal nerve needs to be surgically identified in order to provide an appropriate diagnosis.

Case Presentation

We report the case of a 6-year-old girl, presenting for 2 years with a left cervical latero tumor fistulized at the skin, treated with antibiotics (amoxicilin and clavulanic acid) with a recurrent history of infections and abscesses of the neck, 2 biopsies were made which were not significant. a cervical ultrasound was requested revealing the presence of subcutaneous collection measuring 17.2*15 mm with irregular, heterogeneous and hypoechoic contours. There were also air bubbles appearing behind the left thyroid lobe, in front of the left thyroid cartilage when the patient was performing Valsalva's maneuver. Patient underwent hypopharyngoscopy revealing the presence of an orifice of the sinus was found in the left piriform fossa (**Figure 1**).



Figure 1: Orifice of the fistula in the left piriform fossa in endoscopic view.

The Cervicotomy revealed a sinus located between the common carotid artery and the internal jugular vein. The sinus had pus inside (**Figure 2**). It was resected and 3.0 absorbable sutures were placed at each end of the sinus. A fragment of sternocleidomastoid muscle was interposed to ensure complete closure of the sinus, before closing the planes over a drainage tube. The drainage tube was removed after 2 days. There were no postoperative events. No suppurative events or neck infections were noted 6 months after the operation.

Discussion

The branchial arches are the source of certain unique neck structures. The superior parathyroid glands, the laryngeal cartilages, the pharyngeal and laryngeal constrictor muscles, the superior laryngeal nerve, the left thoracic aorta, the proximal right subclavian artery, and the final branchial body that forms the thyroid's parafollicular cells are all descended from the fourth branchial arch. The neck structure is altered when the pharyngobranchial duct and fourth branchial cleft are not completely obliterated⁶.

Fourth branchial cleft sinuses are a rare condition with a left side predominance that were initially described by Sandborn and Shafer in 1972^{7,8}. In 105 individuals who underwent surgery to address branchial anomalies, Li et al. found that 3.7% of them had fourth branchial cleft anomalies⁹. In the pediatric population, they result in deep neck infections¹⁰. The 3-year-old child's left side had the suppurative mass in the instance that was being presented.

Thirty percent of congenital neck masses are branchial cleft anomalies, of which only two percent are fourth branchial cleft anomalies, an incredibly unusual occurrence. Anomalies resulting in a fourth branchial cleft typically show up as cysts in adults and as sinuses or fistulas in children. The tip of the piriform sinus is where these laryngotracheal sinus tracts start, and they extend inferiorly to leave the throat through the lateral cricothyroid membrane. After that, they go inferiorly and might be found encircling the right subclavian artery or left aortic arch¹¹. (**Figure 2**).

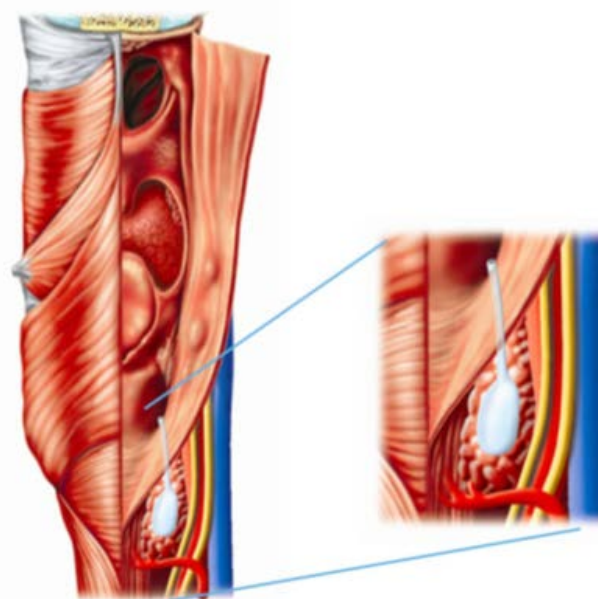


Figure 2: The predicted tract of a fourth branchial anomaly.

The clinical presentation of fourth branchial arch abnormalities varies with age. The most frequent presenting symptoms in neonates are dyspnea episodes¹². Cervical cutaneous fistulas appear in childhood, and these cysts later present classically with a recurrent history of infections and neck abscesses, typically on the left side in 93.6% of cases, the right side in 6%, and bilaterally in 0.5% of circumstances. Clinically, infectious episodes manifest as chronic neck edema. They can also affect the thyroid lobe on the ipsilateral side of the cyst, resulting in acute suppurative thyroiditis. When these signs are observed in the neck, subacute de Quervain's thyroiditis, Hashimoto's thyroiditis, or bleeding from a thyroid nodule should be considered as differential diagnoses⁶. 19% of cases of congenital abnormalities of the fourth branchial arch were found to be associated with bacterial superinfections as a result of the hypopharynx's retrograde transmission of flora, which includes anaerobic bacteria like Eikonella corrodens, Citrobacter, and Proteus as well as a variety of aerobic microorganisms like Streptococcus, Staphylococcus, Haemophilus, and Escherichia coli. The review of 526 cases revealed this information¹³.

Branchial cysts have been investigated using a range of diagnostic techniques, the most effective of which have been

direct laryngoscopy and barium esophagogram, which have the highest positive predictive values (between 88% and 90%, respectively). In contrast to laryngoscopy, barium studies might not reveal a sinuous tract if there is inflammation. Cystic lesions can also be diagnosed and assessed using other methods such as thyroid ultrasonography, computed tomography, and magnetic resonance imaging. Numerous anomalies are identified by chance discoveries¹³.

Treatment options for abscesses of the third and fourth branchial arches include drainage and incision, which are commonly used but have a high recurrence rate of up to 90%. Because of the possibility of a serious infection or primary cancer, surgical excision combined with a partial thyroidectomy is the suggested course of treatment for cysts, sinuses, and fistulas^{14,15}. Surgery carries a higher risk of complications or damage to cervical neurovascular systems, particularly in infants and neonates, but a decreased likelihood of recurrence¹².

After surgery, we discovered a 5 cm long and 1 cm wide sinus tract that was constrained by the thoracic orifice on his caudal side and the hypopharynx on his cranial side. Barberet¹⁶ described this specific surgical condition and noted that the sinus tract may terminate on the aortic cross. Before closing the fistulous orifice plan by plan, a 3.0 absorbable wire was used to push the fistula and insert a fragment of SCM, making the opening blind. On the mentioned orifice, there was no endoscopic intervention. To repair the fistula and lower the chance of recurrence, Givens et al. employed a flap of rotation of the sternohyoid muscle¹⁶. Numerous writers have referred to pharyngeal opening fistula closure as the gold standard of therapy to prevent recurrences^{8,9,17}. Peirera et al. also recommend this modality, associated or not with a loboisthmectomy depending on whether the thyroid lobe is affected or not¹⁸. In front of a thyroid gland that is macroscopically normal, we did not execute a loboisthmectomy. Several treatment approaches have been reported, most notably Watson's, who discovered positive outcomes in five patients undergoing endoscopic coagulation (thermal by electrocoagulation, chemical with silver nitrate/trichloroacetic acid, or CO2 laser) of the fistulous orifice^{7,18,20,21}. With 11, 10, and 19 infants treated with endoscopic cauterization, respectively, Kim, Verret, and Leboulanger have the biggest series of patients^{7,20,21}. In these series, the recurrence rate varies from 0% to 35%. Simple incisions drainage are the primary cause of recurrences, as seen in 94% of cases here¹⁷. Recurrence rates with endoscopic cauterization are 18%, while open surgery without touch on the thyroid has a 15% rate, according to Nicoucar et al. Considering these numbers¹⁷.

5% to 6% of patients have been documented to have surgical site infections, salivary fistulas, and vocal cord paralysis as consequences following surgical and cauterization operations. Due to inflammation and edema that may eventually compress these nerves during electrocautery, paralysis of the superior and recurrent laryngeal nerves may result. Despite the paucity of evidence, the majority of research come to the conclusion that cauterization which is less invasive, has a lower risk of complications, and can be done concurrently with other operations like incision and drainage in the event of an abscess should be the main course of treatment^{22,23}. Moreover, reports of sclerosing agent-treated cases that were successful have been published²⁴.

The outcomes of endoscopic surgery are comparable to those of open surgery, despite the fact that the less invasive endoscopic

approach appears to be favoured these days due to the decreased chance of laryngeal nerve injury. In their description of a novel endoscopic procedure, Huang et al. used the KTP laser in conjunction with fibrin glue on five children without reporting any complications or recurrences⁸.

Conclusion

To sum up, lateral neck tumors can have fourth branchial cleft anomalies as an uncommon but significant differential diagnosis. Age-related differences in clinical manifestations are common, and viral episodes can cause major side effects. A combination of imaging and direct laryngoscopy is usually used to make the diagnosis. Surgical excision and partial thyroidectomy are the suggested treatments to lower the risk of primary cancer and severe infection and to avoid recurrence. Because endoscopy with cauterization has a lower likelihood of problems, it is a minimally invasive method that may be recommended in some situations. All things considered, a complete assessment is required for any patient with inexplicable neck lumps, particularly on the left side, in order to guarantee a timely diagnosis and suitable therapy.

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