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Left Fourth Branchial Cleft Cyst - A Rare Clinical Entity

Case Report

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Abstract

Fourth branchial arch anomalies are extremely rare and are the result of abnormal development of the branchial apparatus during embryogenesis. They arise from an incomplete involution of the fourth gill slit during development. Failure to recognize appropriately of these anomalies may result in misdiagnosis. Here we present a unique presentation of female patient of 21 year old which presented an enlarging painless lateral neck mass. Diagnosis was obtained by ultrasound imaging.

Keywords: Branchial cleft anomalies; Neck mass; Fourth branchial cleft cyst, Complete surgical excision, Pyriform sinus, Branchial apparatus

Introduction

Branchial cleft anomalies originate from the errors during embryogenesis, resulting in incomplete involution of branchial clefts. Fourth branchial cleft cysts are particularly rare accounting for less than 30% of all branchial arch anomalies [1,2]. The branchial cleft cyst may originate from the 1st, 2nd and 3rd branchial arch remnants, however fourth branchial cleft cysts are rare [3,4].

The mean solar time between onset of symptoms and proper diagnosis seems to be 5 years. Treatment is to excise the cyst and often combined with partial thyroidectomy that may further decrease recurrence rates [5].

The incidence rates of types I, II, III and IV are 80%, 95%, 2% and 1-4% respectively with just 100 reported cases. Type IV is reported very rarely in previous studies. This disease is not linked with gender, anyhow as claimed by the evidence; it has been reported in females more than males, at the same time on the reverse some believe that 60% of this disease would happen in males [6].

Of all the fourth branchial cleft anomalies true cysts are particularly difficult to detect as a tract opening is often not identifiable on endoscopy or contrast enhanced radiography. Moreover while the number of reported fourth branchial cleft anomaly in the literature has tripled over the past 20 years [7-11].

Fourth branchial arch anomalies are extremely rare and almost occur on the left side [12,13]. These anomalies typically present as recurrent neck infections, abscess, or acute supparative thyroiditis [14,15]. Imaging is requisite in managing these lesions. For definitive diagnosis, the relationship of the anomaly to laryngeal nerves must be confirmed prior to operation [16].

Here we present a rare anomaly of fourth branchial cleft cyst in a young female.

Case Report

A 21-year-old female was referred to our diagnostic centre for imaging of swelling on the left side of neck. Patient complained of a solitary swelling on the left side of neck since one year of duration, which was incidental on onset and progressively increased since last four months. She mentioned no change in her voice or suffered from

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any breathing disorders or dysphasia. In addition, she reported that there was no history of such similar symptoms for any of her family members.

On examination patient was a febrile with all parameters within normal limits. The swelling was visible on the left side of neck measuring 5 x 3 cm extending about 5-6 cm below mandible. The mass was anterior and deep to sternocleidomastoid muscle but just above the clavicle. No abnormalities were noticed in nose, pharynx and larynx.

On palpation, the mass was soft, mobile and non-tender, no compressive signs were present. The swelling was anterior to sternocleidomastoid muscle almost palpable in the midline that moved with deglutition but was not moving with protrusion of tongue.

Ultrasound imaging of the neck was done which revealed a cystic lesion that was measuring 4.2 x 3 x 1.8 cm (Figure 1). It was located deep to carotid sheath almost in the laryngeal wall (Figure 2).

The lesion was well defined anechoic and perilaryngeal in position. The lesion was located on the left side near midline between thyroid cartilage and deep to infrahyoid muscle. The thyroid gland appeared to be normal and thyroid scan showed no functional thyroid tissue in clinically palpable swelling. The mass was centered in the left carotid space (Figure 3). The differential diagnosis was confirmed as fourth branchial cleft cyst.

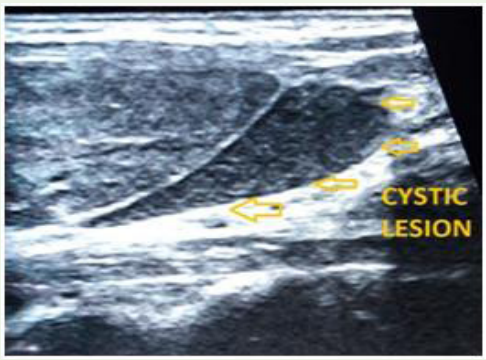

Figure 1: Showing the Cystic Lesion.

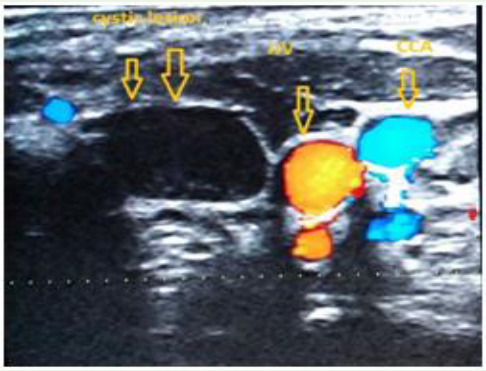

Figure 2: Showing the cystic lesion and the relation of carotid sheath.

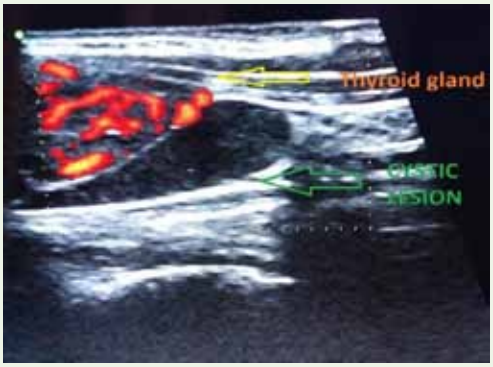

Figure 3: Showing the cystic lesion and normal thyroid tissue.

Discussion

Branchial arch anomalies are the second most common cause of congenital lesions of the head and neck representing 20% of the cervical masses [1,3,17,18].

Branchial cleft cyst is a common cause of soft tissue swelling in the neck of young adult, generally occurs unilaterally and typically seen in the lateral aspect of the neck. It is clinically apparent in the late childhood or early adulthood [19,20]. The branchial cleft cyst usually presents in the second through fourth decades of life [3,15]. However, in our case the patient was 21 years old.

Fourth branchial cleft anomalies are very rare among branchial cleft anomalies. Anomalies of the second branchial cleft commonly called lateral cervical cyst, which are most common and are about 90% [1,9,12,14,16,21]. The remaining 10% are comprised of first, third and fourth branchial clefts with fourth being the rarest [1,21], Representing only 1-4% of branchial apparatus anomalies [13,22]. Bilateral cases accounts for 2-3% of the cases [3,23].

The fourth brachial cleft anomaly was described by Sandborn and Shafer and not until 1981 that Liston theorized the fourth branchial arch anomaly [7,24]. This anomaly most commonly presents as sinus tract that course from the apex of pyriform fossa to the upper lobe of the left side of thyroid gland. This is because fourth branchial arch anomalies are remnants of the embryological tract, which arises from pyriform sinus [1,22,24,25]. The tract terminates in the neck posterior to the internal and common carotid arteries. A fourth branchial cleft cyst is differentiated from the other types of branchial cleft cysts through anatomical landmarks; it is bordered laterally by sternocleidomastoid muscle, medially by trachea, Anteriorly by common carotid and anteromedial by the strap muscles [24,25].

Four types of branchial cleft cysts are as follows -

- Type-I: is often superficial and located on the anterior surface of sternocleidomastoid muscle deep to platysma. However, it's not connected to the carotid sheath. The incidence rate is 8%.
- Type II: is most common and the incidence rate is 95% and located anterior to sternocleidomastoid muscle and posterior towards submandibular gland and lies lateral to carotid sheath.

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- Type III: it is located between the bifurcation of internal and external carotid arteries lateral to the laryngeal wall. The incidence rate is 2%.
- Type IV: It is located deep to carotid sheath almost in the laryngeal mucosal space and would open to the larynx. Such type of cyst emerges as neck mass, abscess and acute thyroiditis; therefore, some authors suggest that the possible existence of branchial archanomalies should be considered in all thyroiditis cyst [3,23]. The patient in this case suffered from type IV branchial cleft cyst.

Patients with fourth branchial cleft cyst usually presents with a painless lateral neck mass along the anterior border of the sternocleidomastoid muscle and most commonly located on the left side [1,6]. It was positively seen in our case.

A predilection for the left side is likely attributable to an embryologic tendency for vascular development in left hemi thorax and diminished growth of right ultimo branchial body [7]. Spread of infection usually occurs to the left thyroid lobe and causes supparative thyroiditis that reflects the tracts intimate relationship with the thyroid gland [7,27].

Third and fourth branchial cleft anomalies are differentiated anatomically by their relationship to the superior laryngeal nerve, with third cleft anomaly above and fourth cleft anomaly below [17]. Diagnosis of the fourth branchial cleft cyst anomaly can be made easily by imaging techniques such as CT, MRI or Ultrasonography being the modality most commonly used [1,28]. In our case, diagnosis was dome on ultrasonography imaging.

Although most of the branchial cleft anomalies appear as sinuses or fistulae, true cyst have been described as well [7,10,11,26]. In our case, it appeared to be true cyst.

The most common treatment for the fourth branchial cleft cyst is complete surgical excision due to risk of infection or obstruction [1,28,29].

In our case patient was referred to Department of Surgery where complete surgical excision of the cyst was done successfully. Post operatively patient was comfortable.

Conclusion

Although fourth branchial cleft cysts are rare, their existence has to be kept in mind when dealing with the patients presenting with neck mass. As it shows fascinating aberrations of foetal development, a proper preoperative evaluation should be done. Definitive management is achieved by complete surgical excision.

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