Fourth branchial pouch sinus – an unusually late initial presentation

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Abstract

Objective Here we present an unusual case of a male patient with 4th branchial pouch sinus with an initial presentation of pseudothyroiditis at an unusually late age of 20 years.

Method Case report and a review of the world literature concerning 4th branchial pouch sinus is presented.

Result The patient was diagnosed by radiology and endoscopy and treated surgically. Histopathology revealed chronic inflammatory changes, which pointed towards recurrent sub clinical low-grade inflammations over the years keeping the condition occult till 20 years of age.

Conclusion According to available literature it almost always presents in the first decade of life when it might be misdiagnosed. However our report shows it may also exist with sub clinical inflammations or infections and present at a much later stage in life, when the possibility of such a sinus shouldn't be ruled out on the basis of lack of history of recurrent neck inflammations.

Keywords 4th branchial pouch · pseudothyroiditis

Introduction

4th branchial pouch sinuses are rare entities. Almost 95% of congenital branchial apparatus anomalies are related to the 2nd branchial arch, pouch or cleft [1]. Out of the remaining 5% the majority are from the 1st and 3rd arches. It was first described by Tucker [1] in 1973. In fact in one study in 1992 only one case out of a total of 106 cases of branchial anomalies was a fourth arch sinus [2].

Fourth branchial arch anomalies usually manifest in the first decade of life and almost always have a long history of recurrent neck lesions refractory to treatment. On high index of suspicion further investigations lead to a definitive diagnosis.

We present a case of fourth branchial pouch sinus that presented at a relatively late age of 20 years, with a short history of left sided anterior neck pain of 15 days duration.

Case report

A 20-year-old male patient presented with pain in the throat, which aggravated with swallowing, for 15 days accompanied by fever for 2 days.

On examination pharynx was normal and indirect laryngoscopy revealed no abnormality. The upper part of left lobe of thyroid was tender. There was a barely perceptible swelling with ill-defined margins at the tender area. Clinically diagnosed as a case of acute thyroiditis the patient was put on steroids and antibiotics and FNAC was done. The pain and swelling reduced with improvement of odynophagia. FNAC report revealed neutrophilic infiltrate indicating acute inflammation. With improvement of symptoms the patient was discharged. However the patient returned after 7 days with recurrence of all symptoms except fever.

A Barium swallow with Valsalva revealed no abnormality. A CT scan showed a hypo dense area with ill-defined margins between the thyroid cartilage and upper part of the left lobe of thyroid gland (Fig. 1). Flexible fiberoptic

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hypopharyngo/laryngoscopy revealed a small opening at the apex of the left pyriform fossa. The patient was diagnosed as a case of fourth branchial arch sinus and prepared for surgery.

After general anesthesia the internal opening was confirmed by a microlaryngoscopy. The sinus was approached externally with a transverse neck incision at the lower end of the swelling. The strap muscles were retracted and the left upper lobe of the thyroid was exposed along with the superior thyroid vessels and nerve. Dissecting from medial to lateral the superior pole was separated from the thyroid cartilage revealing the dilated distal end of the sinus tract partially sandwiched between the two. Proximally the tract passed through the ala of the thyroid cartilage upto the floor of the pyriform sinus (Fig. 2). The tract was excised and the mucosal defect in the pyriform sinus closed with extramucosal sutures. Postoperative recovery was uneventful.

Fig. 1 CT scans showing hypo dense area overlying the left ala of thyroid cartilage with the sinus tract breaching it

Fig. 2 Operative view showing the sinus tract passing through the thyroid cartilage

Histopathology showed a sinus tract with fibrocollagenous tissue and numerous inflammatory cells composed of lymphocytes, histiocytes and neutrophils. No lining epithelium was noted. This was suggestive of an inflammatory sinus tract with fibrosis (Fig. 3).

Discussion

Though theoretically possible, a complete fourth branchial cleft / pouch fistula has never been demonstrated. What is seen is a sinus with a proximal opening in the apex of the pyriform fossa and the distal end extending to any point along the theoretical extent of the tract which starts from the pyriform sinus, passes between the thyroid (4th arch) and cricoid (5th arch) cartilages, then descends between the superior laryngeal nerve and cricothyroid muscle (4th arch) and thereafter between the trachea and recurrent laryngeal nerve. On the left side the tract would loop around the aortic arch and on the right side around the sub clavian artery, to rise in the cervical region posterior to the common carotid artery before looping over the hypoglossal nerve to finally descend and open on the skin of the lower neck along the anterior border of the sternocleidomastoid muscle [3]. In our case the sinus tract extended upto the perithyroid space, which is the most common among the reported cases.

Though communicated for the first time by Japanese authors in 1957 it was Tucker [1] who first published a case of 4th pouch sinus in English. Typically a 4th branchial pouch sinus presents in the 1st decade of life as recurrent neck abscesses often involving the perithyroid space, recurrent suppurative thyroiditis [4–7] which may even require a hemithyroidectomy [8], recurrent pseudothyroiditis [9], retro pharyngeal abscess with recurrent cellulitis [10] which may extend to the mediastinum to form an abscess there

Fig. 3 Histopathological slide showing sinus lumen, with multiple bits of fibrocollagenous tissue and numerous inflammatory cells. Note the lack of lining epithelium

[11], neonatal or pediatric respiratory distress with [12, 2] stridor or even as an asymptomatic mass [12] or as an incidental radiological finding [12]. This condition is often misdiagnosed initially until recurrence of symptoms leads to further investigations and a definitive diagnosis. The interval from onset of symptoms to correct diagnosis ranges from 3 yrs to 34 yrs with an average of 20 yrs [13].

The chief complaint of the patient was severe pain on swallowing. Inflammation of any part of the pyriform sinus will cause odynophagia as it is an important area participating in the passage of food. This location of the sinus opening makes it prone to inflammation and infections. The presentation of our case can also be described as pseudothyroiditis in view of the tenderness over the anatomical area of the superior pole of the left lobe of thyroid. What is unusual is the late age of presentation and diagnosis with no history of any of the above-mentioned symptoms in the past.

Diagnosis is clinched on the basis of an appropriate index of suspicion for its existence where neck swellings, abscesses and inflammations are either recurrent or refractory to treatment supported by radiological investigations, like a barium swallow with Valsalva (trumpet) maneuver, CT Scan or an MRI scan. However barium swallow is not helpful in many of these cases [14]. The hallmark of diagnosis is visualization of the sinus opening in the floor of the pyriform fossa on endoscopic examination.

After confirmation of diagnosis the mainstay of treatment is surgical excision of the tract after control of infection. Identification of the tract can be facilitated by cannulation and injection of methylene blue endoscopically with a cannula [2, 15] or by introduction of a Fogarty® embolectomy catheter. Other treatment modalities have also been described like laser coagulation by endoscopy [16], chemo-cautery with TCA [17] and endoscopic fibrin sealing [18].

The sinus tract is usually lined by pseudostratified ciliated columnar epithelium [15] or stratified squamous non-keratinised epithelium [19]. There was no evidence of any lining epithelium on histopathological examination of the excised specimen in our case, which was probably a result of chronic or recurrent sub clinical inflammation or infections over 20 years since the proximal opening of the sinus lay in the pyriform fossa that is a conduit for all things swallowed.

Conclusion

In conclusion it can be said that the 4th branchial pouch sinus is a rare entity whose diagnosis at first presentation needs an appropriate level of suspicion and awareness of the existence of this condition. Though it almost always presents in the first decade of life when it might be misdiagnosed, it may also exist with sub clinical inflammations or infections and present at a much later stage in life, when the possibility of such a sinus shouldn't be ruled out on the

basis of lack of history of recurrent neck inflammations or abscesses.

Summary

4th branchial pouch sinus is a rare entity. It almost always presents in the first decade of life. The case reported here had the initial presentation at 20 years age.

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