Pyriform sinus fistula as a cause of acute suppurative thyroiditis presenting as hyperthyroidism in a teenager with neurofibromatosis 1: a case report

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Background: A pyriform sinus fistula is a rare congenital anomaly that involves failure of obliteration of the third or fourth pharyngeal pouches during the embryological period. It is recognized as a cause of acute suppurative thyroiditis.

Case Description: We describe a case of a 14-year-old girl with Neurofibromatosis-1 that presented with malaise, sore throat, trouble sleeping, palpitations, dysphagia, and heat intolerance. Physical exam showed a tender thyroid nodule. Further evaluation revealed transient thyrotoxicosis. She had elevated free T4, low thyroid-stimulating hormone, and an elevated white blood cell count. A neck ultrasound and non-contrast computed tomography (CT) showed no evidence of a thyroid abscess. Radioactive iodine uptake revealed a cold nodule. Fine needle aspiration of the thyroid nodule diagnosed a thyroid abscess. Investigation of the anatomy during the direct laryngoscopy discovered a thyroid abscess caused by an infected pyriform sinus fistula. Antibiotics were given and the abscess was drained. After resolution of the infection, a hemi-thyroidectomy was performed to prevent re-infection.

Conclusions: Acute thyrotoxicosis of unknown origin should prompt investigation for a thyroid infection and a primary cause such as a pyriform sinus fistula. If discovery of a pyriform sinus fistula is made, the fistula should be surgically removed after resolution of the infection in order to prevent recurrence of infection.

Keywords: Pyriform sinus fistula; thyroid abscess; thyrotoxicosis; neurofibromatosis-1 (NF1); case report

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Introduction

A pyriform sinus fistula is a rare congenital anomaly involving failure of obliteration of the third or fourth pharyngeal pouches. Pyriform sinus fistulas are recognized as a cause of deep neck infections and acute suppurative thyroiditis. It is important to do a proper work-up when treating a thyroid infection or thyroiditis with unknown etiology and to diagnose pyriform sinus fistulas to prevent recurrence of infections. Here we present the case of a 14-year-old girl with a known diagnosis of neurofibromatosis-1 (NF1), who presented with symptoms of thyrotoxicosis with neck pressure symptoms that were due to a thyroid abscess caused by an infected pyriform sinus fistula. We present the following case in accordance with the CARE reporting checklist (available at https://pm.amegroups.com/article/view/10.21037/pm-21-25/rc).

Case presentation

A 14-year-old girl with NF1 presented with malaise, sore

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throat, trouble sleeping for one-week, intermittent chest pain with palpitations, difficulty swallowing, and heat intolerance for one day. A rapid strep test was negative while FT4 was high at 7.0 ng/dL and thyroid-stimulating hormone (TSH) was low at 0.04 µU/mL. On physical exam, the thyroid was enlarged with a 3cm mildly tender nodule in the right lobe. Thyroglobulin and thyroid peroxidase antibodies were negative. Ultrasound and non-contrast computed tomography (CT) neck showed an enlarged right thyroid lobe measuring 5.2 cm × 2.4 cm × 2.5 cm with a heterogeneous nodular region. There was no evidence of a thyroid abscess on the scans. The differential diagnosis at the time appeared to be focal thyroiditis, hemorrhage of the right thyroid gland, or a neoplasm. She was started on 5 mg TID methimazole for what was presumed to be subacute thyroiditis; atenolol was added as needed for tachycardia and a 5-day course of 10 mg BID prednisone to reduce inflammation. As a complete blood count (CBC) showed a left shift and the white blood cell (WBC) count was $17.2\times10^3/mm^3$, Augmentin® was added for 7 days. Radioactive iodine uptake (RAI^{123}) showed a cold nodule. Fine needle aspiration (FNA) was performed due to the size of the nodular region and the results of the RAI. The FNA was an outpatient procedure performed shortly after the results of the CT and RAI were received and revealed a thyroid abscess. A pyriform sinus fistula was discovered during an awake flexible fiberscope direct laryngoscopy to further investigate the anatomy. A small inflamed polyp was visualized in the pyriform sinus and purulent fluid was seen entering the sinus when external pressure was applied to the abscess. An 18-gauge needle was advanced into the neck, and a large amount of pus returned. A 15-blade scalpel was then used to incise the abscess to drain 40–50 mL of purulent fluid. She was started on a 20-day course of amoxicillin following this. Two months later, a CT neck showed marked improvement in inflammatory changes with some residual thyroid abscess. To prevent further recurrence, a right hemi-thyroidectomy was performed. Subsequently 50 µg of levothyroxine was started at follow-up for evolving partial hypothyroidism as indicated by a rising TSH of 7.32 µIU/mL. She continues to have follow-up visits with us and remains euthyroid on levothyroxine without recurrence of any of her prior symptoms related to the abscess. All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient.

Discussion

This case discusses a rare presentation of acute thyroiditis in the form of a thyroid abscess caused due to an underlying embryological anomaly involving the pharynx and the thyroid. Thyroid abscesses and suppurative thyroiditis are infrequent pathologies as the increased amount of iodine content, high vascularity, and thyroid gland encapsulation provides resistance to infection (1). However, there are pathways and predisposing factors that lead to infection of the thyroid gland including hematogenous spread, spread from an adjacent site, and pyriform sinus fistula (1).

Miyauchi *et al.* hypothesizes that pyriform sinus fistulas are a remnant of pharyngeal mucosa that was created with migration of C cells. The tract is formed from the migration pathway the ultimobranchial body takes caudally from the fourth pharyngeal pouch to join the developing thyroid and become C cells. (2). Pyriform sinus fistulas can allow bacteria to travel from the hypopharynx to the thyroid leading to abscess formation and/or recurrent infections.

Presentation of an infected thyroid from a pyriform sinus fistula can vary, but most patients are euthyroid. A retrospective study that evaluated 48 cases of pyriform sinus fistulas revealed 62.5% of the cases presented as a neck abscess while some presented with dyspnea, acute suppurative thyroiditis, or a thyroid nodule (3).

Acute suppurative thyroiditis can present as anterior neck swelling, pain, fever, and increased WBC count (4). Hyperthyroidism, as reported in a review by Lafontaine *et al.*, has been reported in up to 42% of case reports in patients with acute suppurative thyroiditis (5). Our patient presented with acute suppurative thyroiditis, thyroid nodule with pressure symptoms, along with hyperthyroid symptoms. Interestingly, the majority of pyriform sinus fistulas occur on the left; however, our patient's fistula was on the right.

Diagnosis of a pyriform sinus fistula is essential to prevent recurrent thyroid infection and abscess. Oral contrast CT can diagnose the fistula and should be performed in any suppurative inflammation of the neck with unknown etiology (6). A retrospective review of 48 cases described how barium esophagography showed the sinus tract in 100% of cases; oral non-contrast CT, intravenous (IV) contrast CT, and sonography were good options as

well (3). The fistula may also be discovered incidentally during direct laryngoscopy, such as in our case.

Treatment must include antibiotics to control the infection. Incision and drainage or needle aspiration are acceptable therapies for abscesses. Our patient was first prescribed methimazole and prednisone due to the inaccurate diagnosis of hyperthyroidism. Methimazole is contraindicated for a patient with destructive thyrotoxicosis due to potential aggravation of inflammation. After the infection is under control, the fistula should be removed or disconnected from the thyroid to prevent recurrent infection. Although several techniques have been successful including chemo-cauterization, fibrin glue, and electro-cauterization; the treatment of choice involves excising the fistula completely (7). Hemi-thyroidectomy is often performed to prevent recurrence of the sinus tract which was performed in our patient (3,8).

Thyroiditis can result in gland destruction and permanent hypothyroidism (9) and untreated thyroid abscesses can dissect and spread into the neck or rupture (10). According to the Lafontaine *et al.* review, patients with acute suppurative thyroiditis had 7.8% mortality (5). Because of these complications and increased morbidity from the symptomology of thyroiditis and thyroid abscesses, it is important that the clinician assess for an anatomical defect, so as to prevent recurrence.

Our patient also had NF1. Although numerous neuro-fibromas and freckles were seen in our patient, no skin ulceration was found. Literature review did not show any cases that shared this combination of NF1, pyriform sinus fistula leading to a thyroid abscess. Furthermore, there does not seem to be an increased association of NF1 with occurrence of a pyriform sinus fistula.

Acute thyrotoxicosis should prompt investigations into an infection of the thyroid gland caused by an anatomical defect such as a pyriform sinus, and aggressively treated medically and surgically to prevent spread and recurrence.

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Footnote

Reporting Checklist: The authors have completed the CARE reporting checklist. Available at https://pm.amegroups.com/article/view/10.21037/pm-21-25/rc

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Conflicts of Interest: Both authors have completed the ICMJE uniform disclosure form (available at https://pm.amegroups.com/article/view/10.21037/pm-21-25/coif). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

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