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Ten Years of Experience With Third and Fourth Branchial Remnants

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Background: Third and fourth branchial remnants may result in cysts and abscesses that are in close contact with the thyroid gland. These anomalies are rare and often present diagnostic and therapeutic challenges.

Methods: The charts of patients diagnosed with a branchial anomaly between July 1991 and July 2001 at the Montreal Children's Hospital were reviewed. All cases of third and fourth branchial remnants or pyriform sinus fistulae were identified. Clinical presentation, imaging, treatment, and outcome were recorded.

Results: Eight patients with a third or fourth branchial anomaly were identified and ranged in age from birth to 13 years. All anomalies were left sided. Presenting symptoms consisted of an asymptomatic cervical mass (n = 1), an infected mass (n = 5), neonatal respiratory distress (n = 1), and 1 incidental cyst found on magnetic resonance imaging. Ultrasonography was useful in suggesting the diagnosis in 7 cases. Barium swallow was performed in 3 patients with 2 positive results. Pharyngoscopy results showed the internal opening in 2 of 7 patients. A portion of the thyroid gland was resected in 6 patients. One patient has not yet undergone a definitive procedure. There was 1 recurrence in a patient whose pathology did not confirm a branchial remnant.

Conclusions: The diagnosis and management of pyriform sinus anomalies are challenging. Ultrasound scan, computed tomography scan, barium swallow, and pharyngoscopy are all useful. The portion of thyroid involved in the fistula must be excised en bloc with the inflammatory mass, and the tract should be ligated at the level of the pharynx to minimize recurrence.

INDEX WORDS: Pyriform sinus fistula, piriform fossa, branchial cleft anomalies, branchial cysts, neck mass, acute suppurative thyroiditis, surgical management.

FAILURE of the third or fourth branchial pouches to obliterate in utero results in cysts or sinus tracts that lie in close proximity to, or inside, the thyroid gland.[1-3] When present, the sinus tract originates in the pyriform (or piriform) fossa, also known as the pyriform sinus.[4,5] Hence, these anomalies have been called pyriform sinus "fistulae," even though an external opening to the skin rarely is present. The terms *pyriform fossa sinus*, *third or fourth branchial* (*or pharyngeal*) *pouch remnant*, or *third or fourth pharyngobranchial duct remnant* would seem more appropriate. These remnants almost always occur on the left side, although a small number of right-sided anomalies have been reported.[1,6] These anomalies are rare, and previous series typically included small numbers of patients, most of whom had undergone multiple procedures before a correct diagnosis was made.[1,2,7] We present 8 cases of third or fourth branchial pouch remnants to highlight recent trends in diagnosis and management.

MATERIALS AND METHODS

The charts for all cases coded as branchial anomalies at our pediatric hospital were reviewed manually for the period from July 1991 to July

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2001. All cases in which the diagnosis of third and fourth pouch anomalies was made by diagnostic imaging or at surgery were included. Presenting symptoms and signs, diagnostic imaging, medical and surgical treatments, as well as follow-up and recurrence were abstracted for each case. One of the patients was included in a previous publication.[7]

RESULTS

Over the 10 years, 8 patients were found with a diagnosis compatible with a lower branchial pouch anomaly. Ages ranged from newborn to 13 years (mean, 5 years). The male to female ratio was 3:5. All anomalies were on the left side (Table 1). Two patients had asymptomatic noncommunicating cysts that were partly intrathyroid (patients 2 and 7). One newborn presented with stridor and respiratory distress requiring intubation. The remaining patients presented with abscesses in or adjacent to the upper pole of the left thyroid gland with direct or indirect proof of a communicating sinus tract in 4 of the 5. All patients underwent ultrasound examination. This was useful in showing the close relationship of the mass with the thyroid gland in 7 cases and the presence of gas within the mass in 2 patients. In one instance, the ultrasound scan was suggestive of a malignant process (patient 3). Barium swallow confirmed a pyriform fossa sinus tract (Fig 1) in 2 patients, 1 on the third attempt only. Findings of this examination were normal in 1 patient. Rigid laryngopharyngoscopy allowed visualization of the pyriform fossa opening in 2 of 5 patients presenting with infected masses (Fig 2). In all patients in whom pathologic examination of the surgical specimen found evidence of a branchial origin, there was no recurrence, although follow-up is short. There was recurrence in 1 case in which no evidence of a tract was found. One patient had exploration of the left thyroid lobe and incision and drainage of a left thyroid abscess. Pharyngoscopy confirmed the presence of a sinus. The inflammatory process resolved, but ultrasound scan showed a persistent mass containing some gas. Parents refused definitive surgery (patient 3).

DISCUSSION

During embryologic development, the third pharyngeal pouch develops with dorsal and ventral components. The dorsal component develops into the inferior parathyroid, whereas the ventral component becomes the thymus. After this, the attachments of the thymus to the aortic arch cause the thymus and parathyroids to descend as the heart migrates caudally. This migration accounts for the more caudal location of the third pouch parathyroids (inferior parathyroids) in relation to the fourth pouch parathyroids (superior parathyroids). The fourth pouch, as well as the rudimentary fifth pouch, form the caudal pharyngeal complex, the dorsal expansion of which develops into the fourth pouch parathyroids, whereas the ventral portion (ultimo-branchial body) becomes incorporated into the thyroid gland to form the parafollicular C cells.[5,8]

Both the third and fourth pouches are connected to the pharynx by the pharyngobranchial duct, which degenerates during the seventh week of development. Persistence of this duct results in a sinus tract that communicates with the pyriform fossa. Third pouch remnants are described as passing superior to the superior laryngeal nerve and posterior to the common carotid artery. In theory, the tract should emerge above the thyroid cartilage (fourth arch origin). Fourth pouch remnants should emerge caudal to the thyroid cartilage and cricothyroid muscle and pass between the superior and recurrent laryngeal nerves.[5,7,9,10] Other criteria that have been used to differentiate third from fourth pouch remnants are the location of the internal opening into the pyriform fossa (third, cephalad fourth, at apex, ie, caudal part of the fossa, or even in the proximal esophagus) and the presence of thymic tissue (third pouch) or thyroid tissue (into which fourth pouch derivatives incorporate). Although the theoretical origin of the tract is of interest, overlapping features can be found,[1,5] and postinfectious fibrosis at the time of surgery often precludes a precise identification of the anatomic relationships. The external opening of a complete third or fourth branchial fistula should be at the same level as a second fistula, with a fairly straightforward course for the former and a fascinating course above the carotid bifurcation and below the aortic arch before ascending to the pyriform fossa for the latter.[5,9] However, it appears that complete congenital third and fourth branchial fistulae are rare, most being secondary to recurrent infection and repeated surgery.[1,7] As with other branchial remnants, the sinus tracts are lined by stratified squamous epithelium, which may be replaced in areas with respiratory epithelium.[5,10-12] The predominance of left-sided lesions has been noted previously in all reports and is likely related to the embryology of the branchial apparatus.

Third and fourth branchial remnants have been reported to present at any age, from diagnosis in utero to adulthood.[1,2,10,13-15] In the neonate, these anomalies can be dangerous because of rapid enlargement as the infant swallows saliva, formula, or milk, leading to tracheal compression and respiratory distress.[10,13,16]

Noncommunicating or noninfected communicating cysts may present as cold thyroid nodules.[17] As we have noted in a previous publication and seen again in the current series, the cysts may be partly or completely intrathyroid and can be confused with thyroglossal duct cysts.[17]

When infected, diagnosis and successful excision of a pyriform fossa sinus is very challenging and requires a meticulous approach. A history of recurrent upper respi-

Table 1. Summary of Patients

Patient No.	Age	Sex	Presentation	Year	Imaging	Pharyngoscopy	Operation	Pathology
1	4 yr	F	Infected TDC	1992	*Plain films*		Incision and drainage	
	6 yr		Infected TDC	1994	*US*: midline phlegmon		Antibiotics only	
	7 yr		Fever, painful L neck mass	1995	*US*: heterogeneous mass with gas in area of L thyroid *CT*: mass with gas *Ba swallow*: tract from L piriform fossa (Fig 1) *Tc99 scan*: lack of uptake upper portion L thyroid	Negative	L upper pole of thyroid taken en-bloc with mass	No sinus tract seen
	12 yr		Torticollis, fever, neck pain	2000	*CT*: abscess with gas anterior to vessels, extending down to first rib		Incision and drainage	
2	13 yr	M	Incidental thyroid nodule on MRI	1996	*MRI*: cystic nodule in lower pole of L thyroid *US*: Mixed solid/cystic nodule lower pole L thyroid *Tc99 Thyroid scan*: cold area intra *v* lateral to L thyroid		L thyroidectomy	Branchial cleft origin
3	4 yr	F	Painless midline neck mass, cough, fever, URI	2000	*US*: mass involving L lobe of thyroid, multiple nodes, ? Ca *US* 2 months postoperative: mass containing gas	Positive	Exploration L thyroid lobe, I+D abscess	Thyroid tissue with inflammatory cells
4	1.5 yr	F	Painful L neck mass, dysphagia	2000	*US* + *CT*: mass adjacent to superior aspect of L thyroid lobe *Barium swallow*: normal × 2	Negative (flexible)		
	3 yr		Planned follow-up	2001	Ba swallow: tract from L pyriform fossa	Negative	L superior pole of thyroid taken en bloc with lesion	No epithelium lined tract seen
5	6 yr	F	Tender L neck mass, dysphagia, hoarseness	2000	*US*: L neck mass *CT*: Abscess between pyriform fossa and thyroid gland (Fig 3)	Negative	L upper pole of thyroid taken en bloc with mass, tract excised	Branchial cleft origin
6	8 yr	F	Tender L neck mass	2000	*US*: Inflammatory mass adjacent to superior pole of left thyroid gland *Barium swallow*: normal	Positive (Fig 2)	L upper pole of thyroid taken en bloc with mass. Tract dissected out, clipped and removed	Branchial cleft origin
7	4 yr	M	Asymptomatic L neck mass	2000	*US*: 2 cysts adjacent to L thyroid	Negative	L upper pole of thyroid taken en bloc with cysts	Branchial cleft origin
8	Birth	M	Stridor, respiratory distress	2001	*US* + *CT*: Cystic mass medial to upper pole of L thyroid gland extending superiorly	Negative (direct laryngoscopy without telescope)	Cyst between upper pole of thyroid gland and thyroid cartilage excised, tract penetrating cricothyroid membrane ligated	Compatible with bronchogenic cyst

Abbreviations: TDC, thyroglossal duct cyst; Ba, barium; L, left; URI, upper respiratory infection.

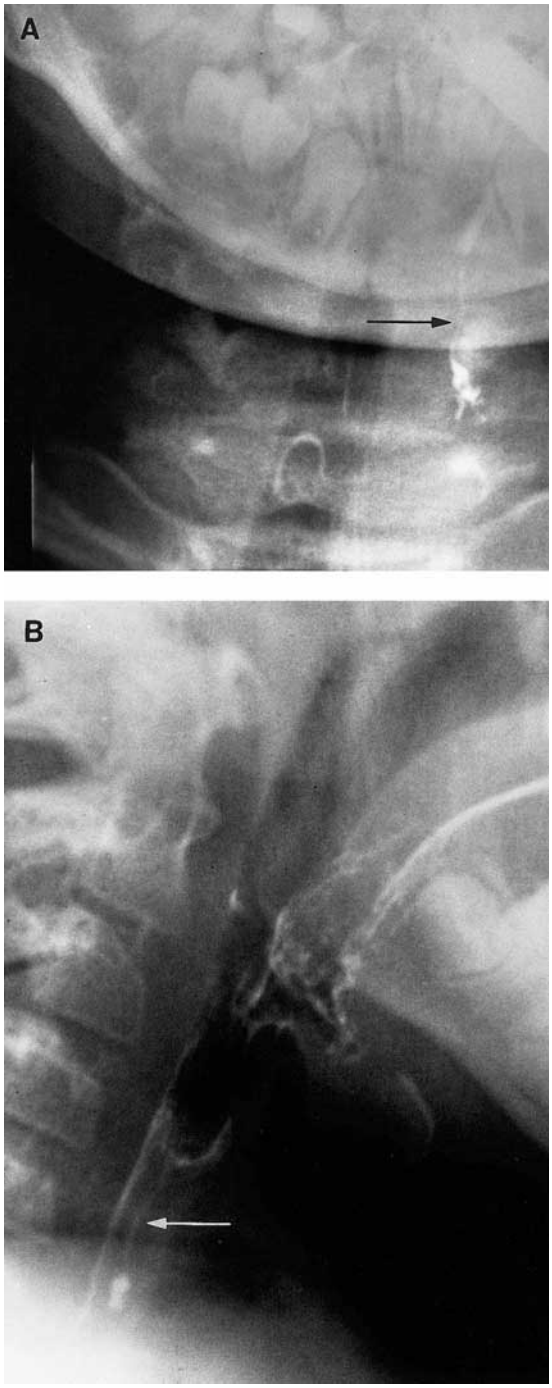

Fig 1. Barium pharyngoesophagram of patient 1 shows a tract (arrow) originating from the left pyriform fossa. (A) Anteroposterior view; (B) lateral view.

ratory tract infection, neck or thyroid pain and tenderness, as well as neck mass is common.[18] Other presentations include cellulitis, hoarseness, odynophagia, thyroiditis, abscess, and stridor. Since the initial report linking acute suppurative thyroiditis and pharyngeal pouch remnants in 1979,[2] it has become accepted that thyroid abscesses in children often indicate an underlying branchial remnant, especially when cultures reveal a mixed flora.[1,3,7,9,19,20] A combination of ultrasound scan, computed tomography (CT) with or without oral contrast, barium swallow, thyroid scan, or magnetic resonance imaging (MRI) may aid in diagnosis. A high index of suspicion is important for the radiologist performing neck imaging of this rare anomaly.[3,11,16,19-27] Multiple modalities should be used to secure the proper diagnosis because we have seen false diagnoses made with single studies. Ultrasound scan often is used as a screening tool in children with neck masses. In recent cases in our series, this modality often offered the first clue to diagnosis by showing an inflammatory process or a cyst closely associated with the left upper pole of the thyroid gland. Ultrasound scan (US) also can show gas within the area, a sign that should be considered pathognomonic of a pyriform fossa sinus.[7,10] When US cannot establish a clear diagnosis, we have found CT scan of the neck with intravenous contrast to be very useful (Fig 3). On CT, the involved thyroid lobe shows low attenuation, and the inflammatory process is seen to extend toward the pyriform fossa; gas also can be seen within the mass. The sensitivity of barium swallow has been reported to be 80%. Diagnostic accuracy is improved by using thin contrast material and by performing the test in the noninfected state, because edema may prevent contrast from entering the tract.[19,28] This was seen in patient 4, in

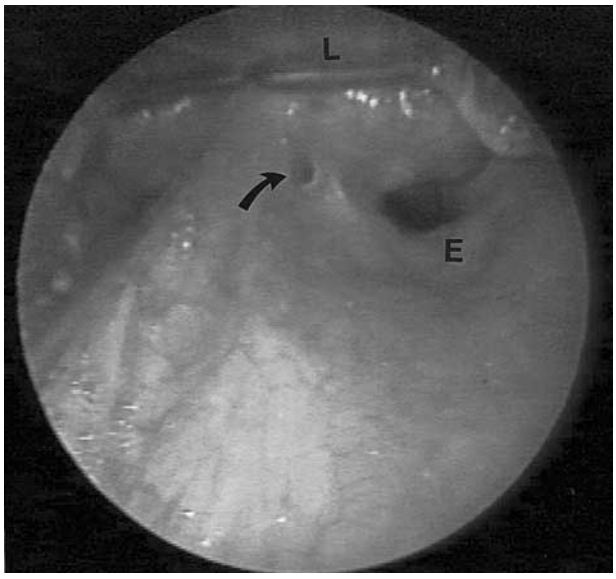

Fig 2. Tract opening (arrow) at apex of pyriform fossa just to the left of the esophageal lumen (E), as seen at rigid pharyngoscopy in patient 6. The laryngoscope blade (L) is retracting the larynx anteriorly. The tract was too small to allow passage of a Fogarty catheter. At operation, identification of the filiform tract within the fibrous tissue could be made only after intraoperative pharyngoscopy and injection of methylene blue via a small suction catheter wedged against the opening.

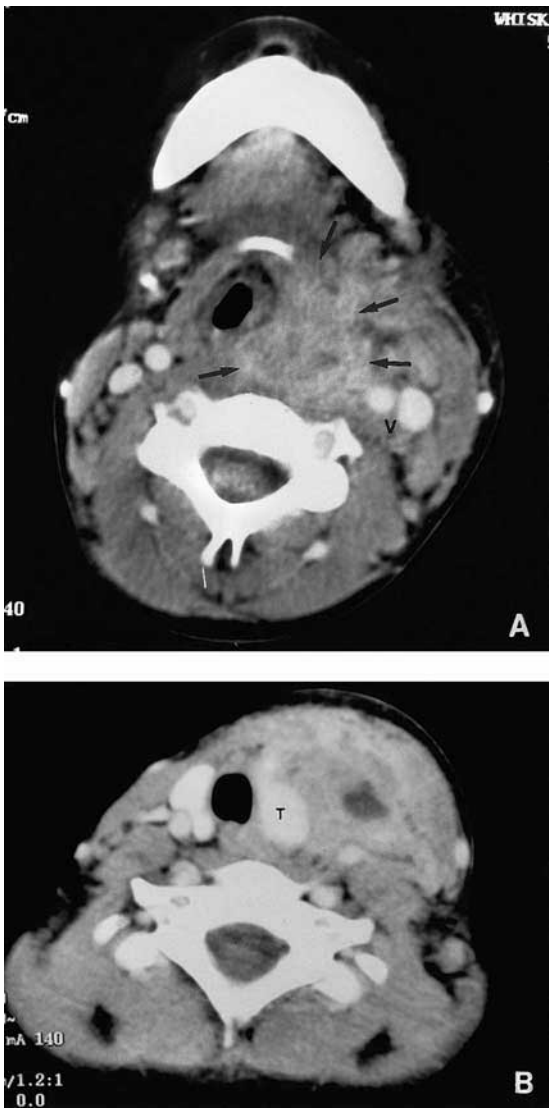

Fig 3. CT scan of the neck with intravenous contrast in patient 5. (A) Arrows indicate a 4- × 4-cm ill-defined mass in the left neck below the larynx. The trachea is being pushed to the right and the neck vessels (V) are displaced laterally and posteriorly. (B) A lower cut shows decreased uptake in the left lobe of the thyroid gland (T) adjacent to the inflammatory mass.

whom the findings were normal at initial presentation and at 6 weeks, but positive several months later. Others have suggested using carbonated beverages to demonstrate air in the tract as an alternative to the barium swallow.[29]

When the child presents with an acute infection, aggressive antibiotic treatment usually is effective, and elective resection of the branchial abnormality should be performed several weeks after the infection has resolved. In the operating room, rigid pharyngoscopy using a Hopkins rod-lens telescope should be performed. If an opening is seen in the pyriform fossa, an attempt should be made to intubate or inject methylene blue through the aperture to make localization of the tract more obvious during neck dissection (Fig 2). Others have used a fine guide wire passed through a flexible fiberoptic gastroscope for this purpose.[30] Another technique is to shine a bright light into the pyriform fossa during direct laryngoscopy and look for transillumination in the operative field.[31]

Meticulous dissection of the neck should be undertaken to visualize the recurrent and superior laryngeal nerves. During neck dissection, the mass or fibrotic tissue should be mobilized and taken en bloc with the adjacent thyroid tissue (excision of superior pole versus hemithyroidectomy will depend on the amount of scarring and ability to visualize the tract or cyst).[7] If a tract is identified, it should be dissected up as high as possible toward the pyriform fossa, ligated, and excised. As mentioned previously, the proximal tract often is embedded in scar tissue or adherent to the thyroid cartilage. Complete excision is facilitated by intraoperative endoscopy, incision of the fibers of the inferior pharyngeal constrictor, and exposure of the inferior cornu of the thyroid cartilage.[32] Pharyngotomy has been advocated to identify the internal opening of the tract but is unnecessary when intraoperative rigid pharyngoscopy is used. In our experience, as well as in published reports, recurrence occurs when thyroid tissue is not removed with the tract as it enters the thyroid or when no tract is identified.[7,30] There were no recurrences when the tract and involved thyroid tissue were resected en bloc, and pathology was consistent with a branchial cleft remnant, although follow-up is short. Although it is recognized generally that the epithelium lining the tract may be destroyed by recurrent infection, the absence of such epithelium in the resected specimen certainly increases the likelihood that the tract was missed and that recurrence will follow.

Recently, treatment of pyriform fossa sinuses by chemocauterization of the internal opening has been reported by a group from Korea with encouraging results.[22,33] Until the long-term efficacy of this method is proven, it should be reserved for patients in whom surgery is refused or considered to be high risk (ie, multiple recurrences).

The diagnosis and management of third and fourth branchial pouch anomalies are challenging. Diagnosis requires a high index of suspicion, both from the clinician and the radiologist. US is first performed, complemented by CT scan if the diagnosis is not clear. Barium swallow is useful to confirm the presence of a sinus tract once the initial episode of infection has resolved. Using various combinations of these imaging modalities, we were able to perform a single definitive operation in 6 patients in the current series. By avoiding multiple epi-

sodes of infection, definitive surgery is facilitated. We stress the importance of excising the portion of involved thyroid gland en bloc with the cyst or inflammatory mass. High ligation of the tract at the level of the pharynx also is important in preventing recurrence, and this is facilitated by intraoperative laryngopharyngoscopy.

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