

Case report

Third branchial cleft anomaly presenting as a retropharyngeal abscess[☆]

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Abstract

Branchial cleft anomalies are congenital developmental defects that typically present as a soft fluctuant mass or fistulous tract along the anterior border of the sternocleidomastoid muscle. However, branchial anomalies can manifest atypically, presenting diagnostic and therapeutic challenges. Error or delay in diagnosis can lead to complications, recurrences, and even life-threatening emergencies. We describe a case of an infected branchial cleft cyst that progressed to a retropharyngeal abscess in a 5-week-old female patient. The clinical, radiographic, and histologic findings of this rare presentation of branchial cleft cyst are discussed. © 2000 Elsevier Science Ireland Ltd. All rights reserved.

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1. Introduction

Branchial cleft anomalies are congenital developmental defects which arise from the primitive branchial apparatus. There are three general types, sinus, fistula, and cyst. These lesions most

often are present as a soft fluctuant mass or draining tract located along the anterior border of the sternocleidomastoid muscle [1–3]. Since most patients present with signs and symptoms of a cystic neck mass or draining fistula, the diagnosis is usually straightforward. Complete surgical excision of the cyst and cyst tract is the treatment of choice for branchial remnants.

Branchial cleft anomalies may also have atypical presentations. This can result in a misdiagnosis of thyroglossal duct cyst, dermal inclusion

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cyst, lymphatic malformation, or malignant neoplasm [2]. In addition, a branchial cleft anomaly can become infected and develop into an abscess. Patients present with tender erythematous infected neck masses and are diagnosed with simple neck abscesses. Such errors in diagnosis can lead to complications and recurrences.

Abscesses due to third branchial cleft anomalies are uncommon. The progression of the infection into the retropharyngeal space is even more un-

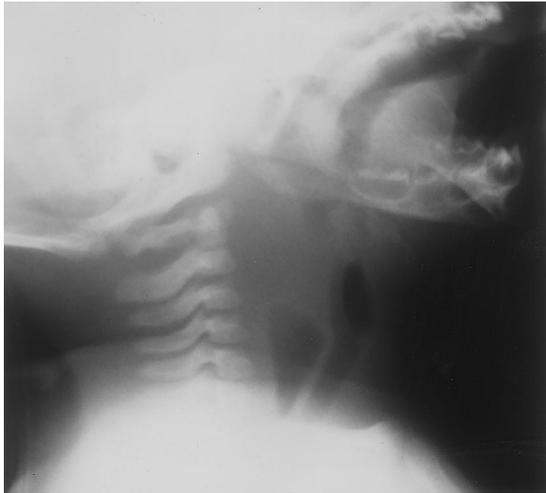


Fig. 1. Lateral neck radiograph demonstrating a prevertebral soft tissue density from C2 to C3 with ventral displacement of the hypopharynx.



Fig. 2. Contrast enhanced CT scan illustrating a large retropharyngeal abscess.

usual. We describe a case of a third branchial remnant presenting as a retropharyngeal abscess in a 5-week-old female patient. Few such cases have been described in the literature [4,5]. The clinical, radiographic, and histologic findings of this rare presentation of branchial cleft cyst will be discussed. This case illustrates the potential difficulties in the diagnosis of branchial cleft anomalies. Awareness of atypical presentations enables surgeons to provide the optimal treatment and timely surgical excision.

2. Case report

A 5-week-old female presented to a community hospital with an enlarging left neck mass, fever, and dysphagia of 1-week duration. She had been given oral cefixime and intramuscular ceftriaxone for several days without clinical improvement. On admission, she had a temperature of 37.9°C. A left sided 4 × 4 cm erythematous neck mass was palpable at the mid-anterior cervical region. Laboratory examination revealed her white blood cell (WBC) count to be 21,400 cells/mm⁻³. A lateral neck radiograph demonstrated a prevertebral soft tissue density from C2 to C4 with ventral displacement of the hypopharynx (Fig. 1). An initial diagnosis of retropharyngeal abscess was made.

Intravenous penicillin and cefuroxime were given. Incision and drainage was performed by the otolaryngology service through a left lateral neck approach. Culture grew multiple organisms, including *α-hemolytic streptococcus*, *Group A streptococcus*, *Group D streptococcus*, *Klebsiella* species, and *Veillonella* species. Her antibiotic regimen was changed to ampicillin and gentamicin.

The patient developed recurrence of the left neck swelling 4 days later. A repeat incision and drainage was performed, again through a left lateral neck approach. Despite the surgery and appropriate antibiotic therapy, the patient continued to have erythema, swelling and induration of wound as well as recurrent fever and dysphagia. Contrast enhanced computed tomography (CT) of the neck on the 10th hospital day demonstrated a large, persistent retropharyngeal abscess (Fig. 2). The patient was then transferred to the Children's Hospital of Los Angeles for further management.

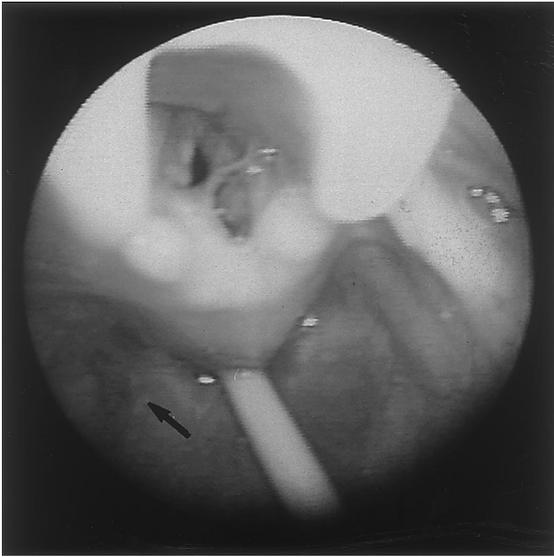


Fig. 3. Examination under direct laryngoscopy showing an opening into the left piriform sinus (arrow).

The patient underwent direct laryngoscopy, which revealed an enlarged erythematous left posterior pharyngeal wall. In addition, a pit was seen in the left piriform sinus (Fig. 3). The diagnosis of an infected branchial cleft anomaly originating

from the piriform sinus was suspected. A decision was made to delay a definitive excision at that setting. She underwent transoral incision and drainage of the retropharyngeal abscess. The patient was continued on intravenous antibiotics post-operatively to allow for the acute infection to resolve.

One week later, she underwent surgical re-exploration with direct laryngoscopy, rigid bronchoscopy, and neck exploration. During the direct laryngoscopy, a probe was placed through the opening at the left piriform sinus, identifying a tract that coursed through the anterior left neck. The neck exploration revealed a sinus tract originating at the left piriform sinus, crossing lateral to the internal and external carotid arteries, and ending in a cyst in the left anterior neck region, adjacent to the anterior border of the mid-sternocleidomastoid muscle (Fig. 4). The cyst and the sinus tract were excised, and the piriform sinus opening was oversewn. Histopathologic analysis revealed a thick-walled cyst lined by stratified squamous epithelium, consistent with branchial remnants. Post-operatively, she was maintained on nasogastric tube feeding and ten days of intravenous antibiotics. Her diet was slowly advanced,



Fig. 4. Neck exploration revealing the branchial cleft sinus tract. The tract courses lateral to the internal and external carotid arteries and ends as a cyst in the left anterior neck.

and she was discharged to home on the tenth post-operative day. She has had no further infection or evidence of recurrence at a 1-year follow-up period.

3. Discussion

Branchial remnants are common congenital cervical anomalies. They account for 17% of all the pediatric cervical masses [2] and remain the most common congenital cervical anomalies in adults [3]. There are three general types of branchial cleft anomalies, cysts, sinuses, and fistulae. Branchial cleft cysts are epithelium lined cavities with no external or visceral opening. [6] They usually present in adulthood [2,3,5,7]. Branchial cleft sinuses are tracts with or without a cyst communicating to either skin or gut whereas branchial cleft fistulae are tracts connecting the gut to the skin. [6] Most sinuses and fistulae present in the first decade, presumably because the external opening of a fistula or sinus allows for early detection [2,5,7].

The etiology of the branchial anomalies remains unproven. The most popular theory proposes that branchial cleft anomalies represent a persistence of the primitive branchial apparatus due to the incomplete closure of clefts and pouches or the failure of obliteration of the cervical sinus of His [3,7]. Sinuses may represent vestigial branchial pouches or clefts while fistulae result from the union of persistent clefts and pouches following rupture of the interposed branchial plate [6]. Cysts may represent the entrapped remnants of either branchial clefts or pouches without formation of fistulas or sinuses [6]. Finally, some authors suggest that cysts arise from epithelial inclusions within lymph nodes [9,10].

First branchial cleft anomalies account for only 5–10% of branchial remnants. The vast majority of branchial anomalies are felt to arise from the second branchial arch [2,7,9,11,12] and comprise 67–95% of branchial anomalies in some series [2,7]. Third arch anomalies account for only 3% of all branchial anomalies [2] and are usually located along the anterior border of the sterno-

cleidomastoid muscle at the junction of its middle and lower third [4,7]. The fistulous tract ascends along the carotid sheath posterior to the internal carotid artery, superior to the hypoglossal nerve, inferior to the glossopharyngeal nerve, superficial to the vagus nerve, and opens into the piriform sinus [7].

The typical presentation of branchial cleft anomalies is well described. These lesions present most often as a soft, round mass or fistula tract located along the anterior border of the sternocleidomastoid muscle [1–3]. They can increase in size during upper respiratory infection [7].

In uncomplicated cases of branchial anomaly, the definitive treatment is complete surgical excision of the cyst and tract. Other modalities such as aspiration and incision and drainage have been shown to be associated with increased risk of recurrence and complications such as hemorrhage, wound infection, cranial nerve injury, and airway obstruction [2,12–14]. In the pediatric patient, surgical management of an uncomplicated, asymptomatic anomaly can generally be delayed until the child reaches 3 or 4 years of age to ensure greater operative ease and safety [9].

A retropharyngeal abscess may complicate an upper respiratory tract infection in young children. Acute lymphadenitis of the retropharyngeal lymph nodes (Rouvier's nodes) progresses to liquefaction and abscess formation. Patients present with erythema and fluctuance of the posterior pharyngeal wall, muffled speech, dysphagia, cervical rigidity, and fever. Lateral neck radiographs may reveal prevertebral soft tissue widening. CT may demonstrate edema or fluid collection in the retropharyngeal space. They can occasionally develop airway compromise [15].

Presentation of a retropharyngeal abscess in a neonate is distinctly unusual [15]. However, this patient presented with typical manifestations of a retropharyngeal abscess, including fever and dysphagia. The lateral neck radiograph demonstrated a prevertebral soft tissue thickening. The presumed diagnosis of retropharyngeal abscess seemed straightforward. However, differentiating primary retropharyngeal abscess from abscess secondary to a congenital cystic lesion posed some clinical challenges.

There were several diagnostic difficulties in this patient's presentation. Lateral neck radiograph did not demonstrate an air fluid level, which is often seen in branchial cleft anomalies [14]. Branchial remnants have a characteristic location along the anterior-medial margin of the sternocleidomastoid muscle [3]. This was not seen in our case. Barium swallow study may reveal a tract, leading to diagnosis of branchial cleft anomaly. While barium swallow has been used in the past to identify a piriform sinus fistula, the fistula may not be seen during episodes of acute inflammation [16,17]. Flexible fiberoptic laryngoscopy may not identify the opening by the piriform sinus because of the redundant mucosa [17]. Therefore, the underlying diagnosis of a congenital cystic lesion was not made until direct laryngoscopy revealed the origin of the cyst tract at the piriform sinus. The opening into the piriform sinus suggested the diagnosis of a third or fourth branchial cleft anomaly. The anatomic pathway of the sinus tract seen during the neck exploration is consistent with a third branchial cleft anomaly. Histopathologic evaluation confirmed the diagnosis of branchial cleft anomaly.

The delay in diagnosis of infected branchial remnants is not unusual. Most cases of infected second and third branchial anomalies reported in the literature are not diagnosed pre-operatively [2,7,8,17,18]. Similarly, these infections were initially treated with incision and drainage. The appropriate diagnosis was recognized only after the development of recurrence or complications [2,7,17,18]. In addition, presentation of branchial anomalies in neonates, such as the patient in this study, is distinctly unusual and can also add to a delay in diagnosis [2–4,11].

Third branchial cleft anomalies often present in an atypical manner [7,11,18]. They have been associated with recurrent lateral neck swelling and abscesses [7,11] as well as with suppurative thyroiditis [18]. However, a branchial cleft anomaly presenting as a retropharyngeal abscess is extremely rare. Only two cases of retropharyngeal abscesses associated with a third branchial remnant have been previously described [4,5]. One case occurred in a 44 year-old female in whom an infected third branchial cleft cyst was initially

treated as an abscess and was drained surgically [4]. The second case involved a 3-day-old child who rapidly developed a retropharyngeal abscess and airway obstruction secondary to an infected third branchial cleft anomaly [5].

Developmentally, the third branchial pouch contributes to the thymus gland and to the inferior parathyroid gland. In this patient, the third branchial pouch persisted as a cul-de-sac originating from the piriform sinus. The opening into the aerodigestive tract allowed for enteric organisms and oral flora to contaminate the cyst. The sac became infected, and the infection then progressed to a deep neck infection. As the infection progressed, it extended posteriorly to the retropharyngeal space.

Management of an infected branchial anomaly is not always straightforward. Some authors recommend antibiotic therapy in the acute phase, followed by definitive surgical excision after the inflammation subsides [7]. Others suggest concomitant antibiotic therapy with aspiration of cyst contents, followed by a delayed definitive surgical excision [5]. Incision and drainage are discouraged, because of the potential for distorting the anatomic planes and thereby complicating subsequent complete excision. Most authors agree that after the inflammation subsides definitive excision is necessary [6].

In this case, incision and drainage alone was not adequate because the underlying source of the infection was not addressed. The delay in diagnosis resulted in recurrent infections over a brief time period. Ultimately, the appropriate treatment consisted of drainage, intravenous antibiotic therapy, and a delayed definitive excision with identification and complete excision of the cyst and tract to its origin. Surgery should be delayed to ensure complete excision and greater operative safety.

Branchial cleft anomalies must be part of the differential diagnosis in the management of pediatric patients with retropharyngeal abscesses. It is important to examine the piriform sinuses for evidence of fistulae in patients with recurrent retropharyngeal abscess. Awareness of atypical presentations enables surgeons to provide the optimal treatment and timely surgical excision.

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