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Nonsyndromic bilateral second branchial cleft fistulae: A case report

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A B S T R A C T

Branchial cleft anomalies are rare congenital malformations that result from the abnormal persistence of branchial clefts during embryogenesis and manifest clinically as cysts, sinuses, or fistulae. In greater than 95% of cases, branchial cleft anomalies originate from remnants of the second branchial cleft. Identification of branchial cleft anomalies, particularly branchial cleft fistulae, are clinically important as these findings may be part of a larger syndromic clinical presentation such as the branchiootorenal syndrome, which necessitates further workup. Branchial cleft anomalies are bilateral in approximately one percent of cases; however, bilateral second branchial cleft fistulae are, for unknown reasons, much rarer. To the best of our knowledge, there have been less than ten cases of nonsyndromic, bilateral second branchial cleft fistulae recorded in the literature. In this report, we present the CASE of a 50-year-old woman with recent left-sided pain, drainage, and swelling in the lower one-third of her neck. The patient reported a history of bilateral "cysts" in the lower one-third of her neck for most of her adult life, which frequently become infected. She denied a personal or family history of renal anomalies or hearing loss. Computed tomography scan with intravenous contrast of the soft tissues of the neck revealed bilateral soft tissue tracts beginning in the region of the tonsillar fossae and extending bilaterally along the anterior borders of the sternocleidomastoid muscle (SCM) down to the skin surface near the level of the thyroid gland, consistent with bilateral second branchial cleft fistulae.

Introduction

Branchial cleft anomalies are rare, congenital malformations that result from the abnormal persistence of branchial clefts during embryogenesis and manifest clinically as cysts, sinuses, or fistulae [1–4]. Second branchial cleft anomalies are overwhelmingly the most common [5]. Bilateral anomalies account for approximately one percent of all cases; however, for unknown reasons, bilateral second branchial cleft fistulae are much rarer [6]. Identification of branchial cleft anomalies, particularly branchial cleft fistulae, is clinically important as these findings may be part of a larger syndromic clinical presentation such as the branchiootorenal syndrome, which necessitates further workup. To the best of our knowledge, there have been less than ten cases of non-syndromic, bilateral second branchial cleft fistulae reported in the literature [1–3,7–11]. Here we present the CASE of a 50-year old woman with bilateral second branchial cleft fistulae presenting to our clinic with left-sided pain, swelling, and drainage in the lower one-third of her neck.

Case report

A 50-year old woman presented to our outpatient otolaryngology clinic with complaints of recent left-sided pain, swelling, and drainage in the lower one-third of her neck. She reported that she had a history of bilateral "cysts" in the lower one-third of her neck for most of her adult life, which frequently become infected. When infected, she stated that these areas feel like palpable, swollen knots which are painful to the touch and are associated with tan-colored drainage. She denied any increased drainage from the area after eating or drinking. She reported that the cyst on the right side of her neck has not been infected for the last few years and has otherwise been asymptomatic. In addition, she reports a similar history of intermittent swelling and drainage associated with an area in front of her right ear. She notes that this area has been present for as long as she can remember. The patient is a lifetime non-smoker and has no personal or family history of renal anomalies or hearing loss. She has no past surgical history. Review of symptoms was contributory only for lack of energy and hoarseness.

On exam, the patient's vital signs were within normal limits. Physical exam was notable for a right sided pre-auricular pit without erythema, swelling, or drainage (Fig. 1). There was a left-sided branchial cleft fistula vs sinus orifice anterior to the lower third of the sternocleidomastoid (SCM) muscle (Fig. 2). There was an identical spot on the anterior, lower third of the SCM muscle on the right which was scarred shut. Neither area was associated with drainage on palpation. Fistula tracts were palpable bilaterally but non-tender, and there were no associated lesions on the skin superficial to the tracts.

A computed tomography (CT) scan with intravenous contrast of the soft tissues of the neck was performed and showed a small, linear, 3–4 mm wide soft tissue tract beginning in the region surrounding the left

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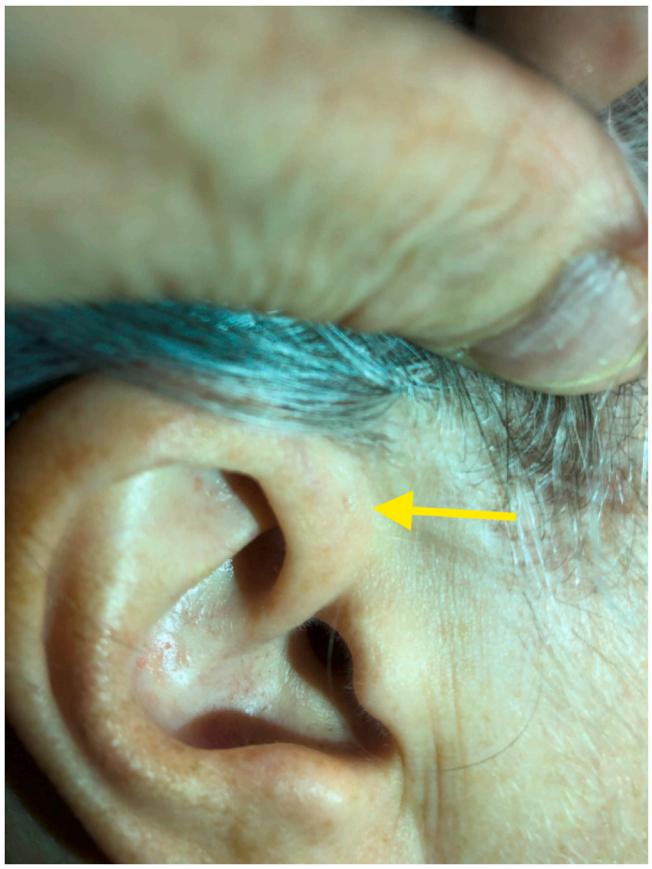

Fig. 1. Right sided preauricular pit.

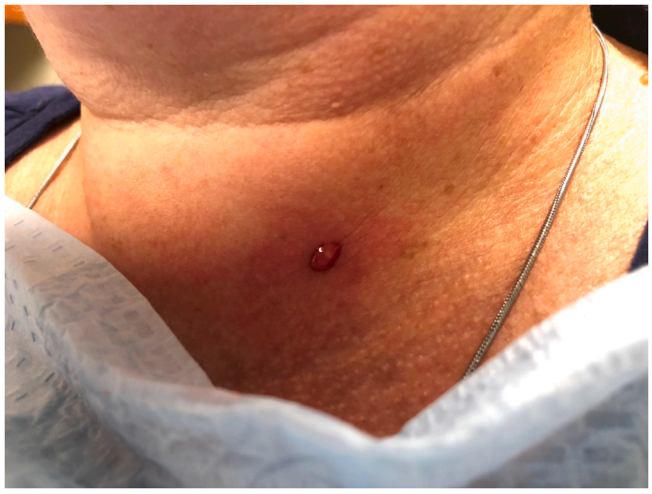

Fig. 2. Left-sided branchial cleft fistula vs sinus orifice anterior to the lower third of the sternocleidomastoid.

tonsillar fossa and extending along the anterior carotid sheath and anterior border of the SCM down to the skin surface near the level of the thyroid (Fig. 3). In addition, there appeared to be some mild inflammatory changes surrounding the distal aspect of the tract. A similar symmetric, but less conspicuous tract was suggested on the right without the associated inflammatory changes appreciated on the left side (Fig. 3). From these findings, a diagnosis of bilateral second branchial cleft fistulae was considered most likely.

The patient was subsequently lost to follow-up, but returned to clinic after a period of 10 months since her initial presentation. At this visit,

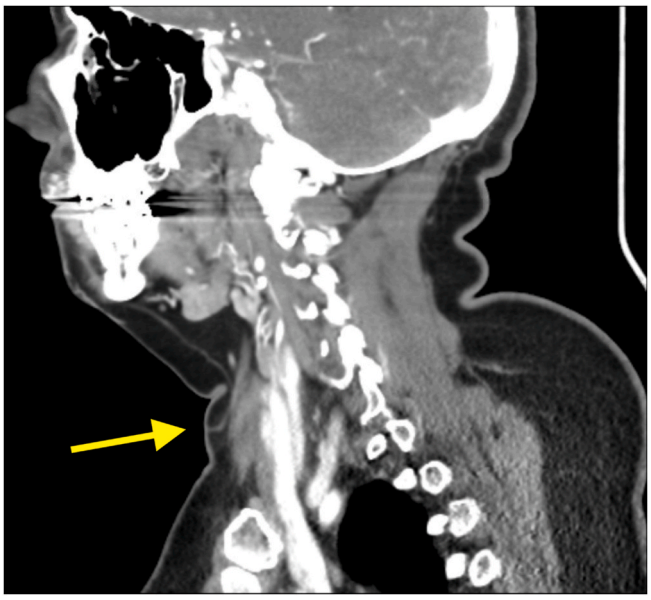

Fig. 3. Sagittal computed tomography scan showing a right-sided 3–4 mm wide soft tissue tract extending along the anterior carotid sheath and anterior border of the SCM down to the skin surface near the level of the thyroid.

she reported resolution of her symptoms. On exam, the left-sided branchial cleft fistula orifice was visible without drainage, and the right orifice was remained scarred shut. There was no swelling or erythema of the skin overlying the fistula tracks. Fistula tracts were palpable bilaterally but non-tender. Our patient opted to refrain from surgical intervention at this time due to symptom resolution.

Discussion

The branchial apparatus consists of six pairs of mesodermal-derived arches, which are lined internally by endodermal-derived pouches and externally by ectodermal-derived clefts [2]. Each branchial cleft, arch, and pouch complex will eventually go on to develop specific structures in the head and neck region [3]. While there are many hypotheses surrounding the development of branchial cleft anomalies [6], the most widely accepted theory is that they result from the abnormal persistence of branchial clefts and pouches during embryogenesis [4]. In greater than 95% of cases, branchial cleft anomalies originate from remnants of the second branchial cleft [12]. First cleft anomalies account for 1–4% of total branchial cleft anomalies, while third and fourth cleft anomalies are significantly rarer [13]. Branchial cleft anomalies are bilateral in approximately one percent of cases; however, bilateral second branchial cleft fistulae are, for unknown reasons, much rarer [14].

Occasionally, branchial cleft anomalies, particularly bilateral branchial cleft fistulae, may be a part of a larger syndromic clinical presentation. The branchiootorenal syndrome, for example, is characterized by branchial anomalies (e.g. branchial cleft cysts, sinuses, fistulae), auricular malformations (e.g. pre-auricular pits), hearing impairment (e.g. conductive or sensorineural hearing loss), and renal malformations (e.g. renal hypoplasia or agenesis) [15]. While our patient presented with bilateral second branchial cleft fistulae and a right sided preauricular pit, she did not possess any of the other characteristic findings present in the branchiootorenal syndrome. Similarly, as the branchiootorenal syndrome is transmitted in an autosomal dominant manner, a family history of similar findings would be expected, which was not evident with this patient [15].

The diagnosis of a branchial cleft fistula can be established by a detailed history and physical exam based on clinical signs that have been described in-depth previously [1–4,6,12]. However, various imaging

methodologies such as fistulography, CT, or MRI can assist with confirming the diagnosis as well as defining the extent of the lesion [2,5]. The definitive management for all branchial anomalies, including fistulae, is surgical excision as these lesions do not spontaneously regress. Various surgical techniques have been described in the literature; however, the most common approach is via an elliptical incision at the level of the external opening of the tract [3,7,16]. For patients with bilateral fistulae, an important question in surgical planning is whether to offer a unilateral or bilateral fistula tract excision. Additionally, if bilateral fistula tract removal is desired, whether the procedure should be staged. As our patient's right-sided fistula tract was asymptomatic for many years and she was experiencing predominately left-sided symptoms, we decided to offer a left-sided fistula tract excision if her symptoms were becoming sufficiently bothersome. Although our patient opted to refrain from surgical intervention at the time of her follow-up visit due to symptom resolution, she agreed to have continued outpatient follow-up for a possible future intervention if her symptoms worsened.

Conclusion

Identification of branchial cleft anomalies, particularly bilateral branchial cleft fistulae, is clinically important as these findings may be part of a larger syndromic clinical presentation such as the branchiootorenal syndrome, which necessitates further workup. To the best of our knowledge, there have been less than ten cases of nonsyndromic, bilateral second branchial cleft fistulae reported in the literature. Diagnosis of branchial cleft fistulae is often based solely on a detailed history and physical exam; however, various imaging methodologies can assist with confirming the diagnosis as well as defining the extent of the lesion. The definitive management for all branchial anomalies is surgical excision, as these lesions do not spontaneously regress.

References

[1] Maddalozzo J, Rastatter JC, Dreyfuss HF, et al. The second branchial cleft fistula. Int J Pediatr Otorhinolaryngol 2012;76(7):1042–5.
[2] Zaifullah S, Yunus MRM, See GB. Diagnosis and treatment of branchial cleft anomalies in UKMMC: a 10-year retrospective study. Eur Arch Oto-Rhino-Laryngol 2013;270(4):1501–6.
[3] Bajaj Y, Ifeacho S, Tweedie D, et al. Branchial anomalies in children. Int J Pediatr Otorhinolaryngol 2011;75(8):1020–3.
[4] Waldhausen JH. Branchial cleft and arch anomalies in children. Semin Pediatr Surg 2006;15(2):64–9.
[5] Rattan KN, Rattan S, Parihar D, et al. Second branchial cleft fistula: is fistulogram necessary for complete excision. Int J Pediatr Otorhinolaryngol 2006;70:1027–30.
[6] Chandler JR, Mitchell B. Branchial cleft cysts, sinuses, and fistulas. Otolaryngol Clin 1981;14:175–86.
[7] Genc S, Demir A, Selcuk A. Bilateral second branchial cleft fistula opening to skin with multiple orifices. J Craniofac Surg 2013;24(4):1496–8.
[8] Süslü N, Süslü AE, Akyol U, et al. Minimally invasive endoscope-assisted surgery for bilateral branchial cleft fistula. Laryngoscope 2013;123:2296–9.
[9] Cheng J, Elden L. Management of pediatric second branchial fistulae: is tonsillectomy necessary? Int J Pediatr Otorhinolaryngol 2012;76(11):1601–3.
[10] Guarisco JL, Fatakia A. Intraoperative fistulograms in the management of branchial apparatus abnormalities in children. Int J Pediatr Otorhinolaryngol 2008;72(12):1777–82.
[11] De Korte FI, van Rijn RR. Bilateral branchial cleft fistula. J Belg Radiol 2013;96(4):218–9.
[12] Agaton-Borila FC, Gey-Escudo C. Diagnosis and treatment of branchial cleft cyst and fistula. A retrospective study of 183 patients. Int J Oral Maxillofac Surg 1996;25:449–52.
[13] Mounsey RA, Forte V, Friedberg J. First brachial cleft sinuses: an analysis of current management strategies and treatment outcomes. J Otolaryngol 1993;22(6):457–61.
[14] Doshi J, Anari S. Branchial cyst side predilection: fact or fiction? Ann Otol Rhinol Laryngol 2007;116(2):112–4.
[15] Can IH, Doğan S, Dönmez M, et al. A different type of branchial fistula as part of a branchiootorenal syndrome. J Pediatr Surg 2012;47:404–7.
[16] Keogh IJ, Khoo SG, Waheed K, et al. Complete branchial cleft fistula: diagnosis and surgical management. Rev Laryngol Otol Rhinol 2007;128(1–2):73–6.