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Case Report
Craniofacial Anomalies

A unique location of branchial cleft cyst: case report and review of the literature

F. Meng[1], Z. Zhu[1], R. A. Ord[2], T. Zhang[1]
[1]Department of Stomatology, Peking Union Medical College Hospital, Beijing, China; [2]Department of Oral and Maxillofacial Surgery, University of Maryland Medical Center, Baltimore, Maryland, USA

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Abstract. Branchial cleft cysts (BCC) are benign lesions caused by anomalous development of the branchial apparatus. This case report describes a 63-year-old woman with a 12 cm × 12 cm sized cystic mass located anterior to the manubrium sternum and sternum. MRI revealed a cystic lesion with a sinus tracking to the piriform sinus. Postoperative histopathological examination confirmed the diagnosis of branchial cleft cyst. Because of the course of the sinus track, it is believed that this was a fourth branchial cleft cyst. These are the rarest of the branchial anomalies, and extension below the peri-thyroid region is very infrequently described. When this extension occurs, it is always post-sternal into the mediastinum, and the pre-sternal presentation here appears to be unique. A review of the relevant literature was performed to summarize the clinical features of fourth branchial cleft cyst and to identify the best options for diagnosis and treatment.

Key words: branchial cleft cyst; unique location; pre-sternal region; fourth branchial cleft cyst; congenital.

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The branchial cleft cyst (BCC) is a congenital cyst derived from the remnants of the branchial apparatus. The term branchial cyst was first used by Ascherson in 1832[1]. The second branchial cleft cyst is the most common lesion of the malformations of the branchial arches and accounts for 95% of branchial anomalies. First cleft anomalies account for 1–4% of these lesions and third and fourth pouch anomalies are rare. Sandborn and Shafer reported the first case of a fourth branchial anomaly in 1972[2]. A consistent feature of these fourth branchial pouch anomalies is that they are primarily left-sided (95–97%)[3], and a clear reason for this is still not understood. A unique case of a right-side fourth branchial cleft cyst anterior to the manubrium sternum and sternum is presented herein. A review of the literature was performed, but there is no previous report of this location. Thus, the case presented here appears to be the first report of this phenomenon.

Case report

The patient was a 63-year-old woman with a 12 cm × 12 cm mass in front of her sternal region (Fig. 1). Her parents had noticed a fistula on her right neck when she was born. There was recurrent discharge of pus from the fistula. About 10 years ago, the fistula had healed by itself. Since then, the mass had grown progressively, with frequent bouts of swelling. When manual pressure was applied over the sternal mass, whitish and smelly pus would flow out from the base of the patient's tongue. Afterwards the mass would partially regress. The patient referred to similar episodes occurring every 2 or 3 months bimonthly or trimonthly for the last 10 years. On palpation, the swelling was non-pulsatile and painless.

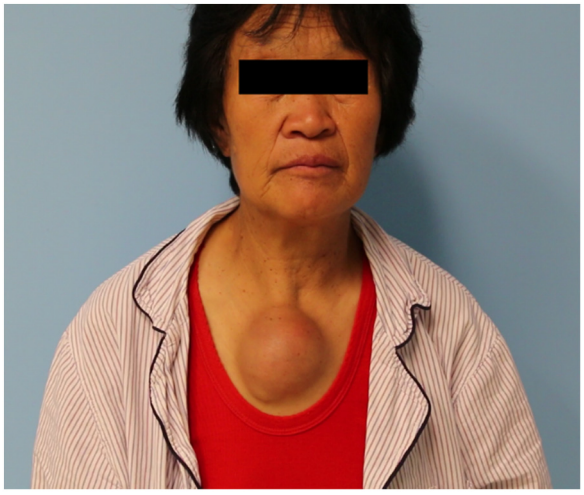

Fig. 1. Frontal photograph of the patient showing the lesion located in front of the sternal region.

Moreover, it was observed to move up and down during swallowing.

An ultrasound scan showed the mass to be cystic. On contrast-enhanced computed tomography (CT) (Fig. 2), a well-circumscribed, non-enhancing 12 cm × 12 cm cystic lesion was noticed in front of the sternum, with a hypodense tract extending from the mass to the piriform fossa. Magnetic resonance imaging (MRI) showed high signal on T2-weighted images and low intensity on T1-weighted images, confirming its cystic nature. The provisional diagnosis was a branchial cleft cyst.

Complete surgical excision is necessary to minimize the chance of recurrence of a branchial cyst. Two incisions were planned to assure complete excision, a superior one in the middle of the neck for the tract, and an inferior one that was elliptical on the surface of the mass. A tunnel was made between the two incisions, so that the intact mass and tract could be taken out. The tract did not pass through the thyroid gland, and as the patient did not have symptoms of suppurative thyroiditis, a partial thyroidectomy was not performed. It was found that the tract originated from the caudal end of the piriform sinus. The recurrent laryngeal and superior laryngeal nerves were not injured, and no complications were encountered.

Histopathological examination showed a fibrous connective tissue wall that included lymphatic tissue with lymphoid follicles, and the cyst wall was lined by non-keratinizing stratified epithelium, which matched the features of BCC (Fig. 3). The patient was discharged without any complication, and there was no evidence of recurrence at the 21-month follow-up.

Discussion

Several theories have been proposed for the development of BCC, such as the branchial apparatus theory, cervical sinus theory, thymopharyngeal theory, and inclusion theory[4]. At present, the most widely accepted theory is that BCC result from incomplete obliteration of the branchial

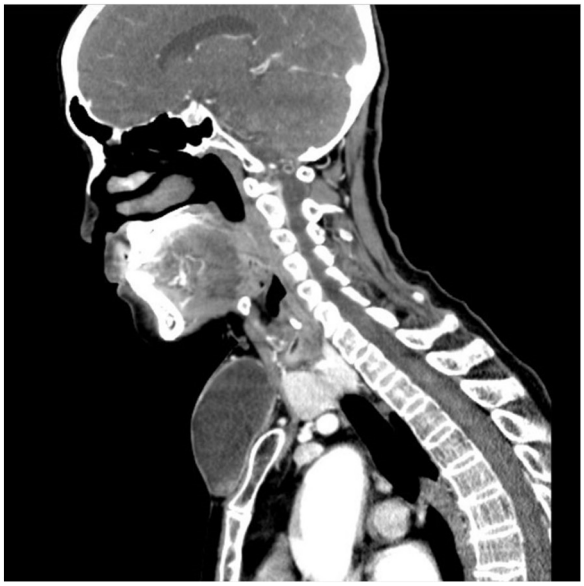

Fig. 2. Contrast-enhanced CT showing the tract extending from the mass to the piriform sinus.

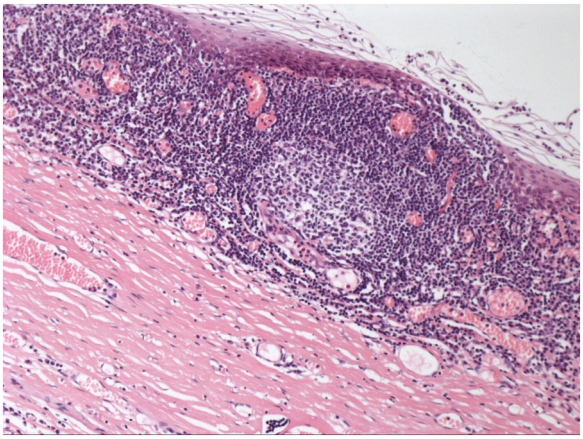

Fig. 3. Lymphoepithelial cyst characterized by a non-keratinized squamous epithelial lining and lymphoid tissue, including an evident germinal follicle within in the immediately surrounding wall.

clefts and pouches during embryogenesis[5].

Both third and fourth branchial cleft cysts are located about two-thirds of the way down the sternocleidomastoid muscle anteriorly. Because of their similar position, distinguishing third and fourth branchial cleft cysts clinically is quite difficult. The fourth branchial pouch sinus may travel inferiorly in the tracheoesophageal groove and descend into the chest to pass around the aortic arch on the left and the subclavian artery on the right . The tract may then change direction, ascending to enter the larynx and then entering the apex of the piriform sinus . A presentation arising from the piriform sinus is a common characteristic of both third and fourth branchial cleft anomalies[6,7]. However, the differences between these two are the origin of the tract and the relationship to the nerves. The third branchial cleft cyst originates from the base (cranial end) of the piriform sinus and passes superficial to both of the superior and recurrent laryngeal nerves. The fourth branchial cleft cyst originates from the apex (caudal end) of the piriform sinus and passes deep to the superior but superficial to the recurrent laryngeal nerves. Accurate differentiation between a third and fourth branchial pouch sinus can only be made by surgical confirmation of the anatomical course.

In most cases, branchial cleft cysts are asymptomatic. However, there are a few key clinical presentations of fourth branchial cleft cyst at different stages of life. In the neonate, as the branchial cleft cyst causes airway stenosis, the main manifestation is respiratory distress. In adults, the most common symptoms are acute suppurative thyroiditis and recurrent cervical abscess. In the case presented, the branchial cleft cyst could be diagnosed by both the medical history and radiological investigations. CT/MRI scans are useful investigations to define the extent of the lesion. Barium swallow study and direct laryngoscopy are more effective methods to show the course of the tract[8]. However, a barium esophagram examination should be performed during a quiescent period. Endoscopy may identify a possible piriform sinus tract opening.

The treatment of fourth branchial cleft cysts, as stated in the literature, may consist of conservative or aggressive therapy, both having their own advantages[9]. Conservative therapy represents a minimally invasive technique using cauterization to obliterate the internal opening of a piriform sinus tract. This may be performed endoscopically using a laser for cauterization and/or combinations of fibrin glue.

The aggressive method refers to completely resecting the tract and cyst by open neck surgery. This remains the gold standard for treatment in many units. Nevertheless, for children who are less than 8 years old, it would seem preferable to undertake conservative therapy and delay surgical neck exploration[10]. For adults, complete excision of the entire fistula tract with a partial thyroidectomy (if the thyroid is involved, or to aid in preserving the recurrent laryngeal nerve) appears to be the best choice for treatment. In consequence, open neck surgery should be reserved for patients older than 8 years of age. In children 8 years or younger, endoscopic treatment is recommended, as using cauterization to obliterate the internal opening of a pyriform sinus tract may yield a lower rate of complications than open neck surgery. Since this tract does not spontaneously regress, there is the potential for recurrence if resection is incomplete.

A review of the literature related to rare locations of branchial cleft cysts was conducted, but no article reporting a pre-sternal location was found. In the case presented here, it could be seen that the tract originated from the apex of the piriform sinus on the MRI/CT scans and intraoperatively. Hence this lesion was identified as a fourth branchial cleft cyst.

In conclusion, a thorough clinical examination should be conducted to evaluate cysts in such a rare location. The examination should include MRI, barium esophagram, direct laryngoscopy, and aspiration biopsy if necessary. In all cases, complete excision including the tract and cyst could reduce the risk of recurrence. And conservative treatment as using cauterization has the advantage of less trauma. The surgical approach needs to be tailored to the type of anomaly or origin of the anomaly.

Patient consent

Written patient consent was obtained to publish the clinical photographs.

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Ethical approval

Ethical approval was obtained from the Ethics Review Board of the Peking Union Medical College Hospital (reference number S-k535).

Competing interests

None.

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Address:
Tao Zhang
Department of Stomatology
Peking Union Medical College Hospital
Chinese Academy of Medical Science
1 Shuaifuyuan
Dongcheng District
Beijing 100730
China
E-mail: drtzhang@126.com