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Case report

Large non-functioning substernal parathyroid cyst: A case report and review of the literature

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ABSTRACT

Objectives: Parathyroid cysts are rare benign lesions of the head and neck that account for less than 1% of cystic neck masses. We present a rare case of a large 6 cm substernal parathyroid cyst.

Presentation of case: An otherwise healthy 65 year-old female presented to the otolaryngology clinic for evaluation of an anterior, midline neck mass. On physical exam, she was noted to have a fullness in the anterior neck extending to the sternal notch. CT demonstrated an enlarged thyroid with a cyst extending to the aortic arch. Initial evaluation suggested a bilateral goiter with substernal extension. The cyst was managed with drainage and observation. After two years of continued growth, the patient underwent a left thyroid lobectomy and mediastinal mass resection via the cervical approach. Final pathology was consistent with a parathyroid cyst.

Conclusions: Parathyroid cysts are a rare cause of neck mass in an adult, and a 6 cm substernal parathyroid cyst represents an unusual site and size for these cysts. Parathyroid cysts are not often considered on the differential of neck and mediastinal cystic lesions. However, appropriate steps should be taken to ensure a proper diagnosis for any cystic lesion in the neck.

1. Introduction

Parathyroid cysts are rare benign lesions of the head and neck that account for less than 1% of cystic neck masses with little over 300 cases reported in the literature [1,2]. Most parathyroid cysts occur in women in their fourth to fifth decade of life. They can be functioning, meaning they secrete parathyroid hormone (PTH), but 90% are non-functioning [3]. Parathyroid cysts can present with symptoms of hypercalcemia or thyroid nodules, but the majority are non-functioning, asymptomatic, and found incidentally.

The clinical diagnosis of parathyroid cysts is challenging, often resembling a thyroid nodule when presenting in the cervical region [4]. Although symptoms are uncommon, the cyst can cause compressive symptoms depending on the size and location, with associated dysphagia, dyspnea, or recurrent laryngeal nerve paralysis [5–7]. The size of parathyroid cysts reported in the literature average 4.8 ± 2.9 cm ranging from 0.5 cm to 15 cm [1].

Location of parathyroid cysts within the head and neck can vary tremendously, from the angle of the mandible all the way down to the mediastinum [1,4]. One review of the literature reported the most common site to be the left thyroid lobe (31.6%) with the superior mediastinum (19.3%) as the second most common, followed by the cervical location at 12.8% [1]. One explanation for mediastinal localization could be the development of parathyroid cysts from ectopic parathyroid glands in the mediastinum. Ectopic parathyroid glands, or any parathyroid gland not immediately adjacent to the thyroid gland, occur in only 15% of patients [8]. Like cervical parathyroid cysts, mediastinal parathyroid cysts can often be resected via a cervical incision, however, sternotomy, thoracotomy, and thoracoscopic approaches are sometimes preferred for intrathoracic mediastinal parathyroid cysts. We report the unusual presence of a large parathyroid cyst measuring 6 cm in length and extending substernally to the aortic arch. The following information has been reported in accordance with SCARE 2020 [9].

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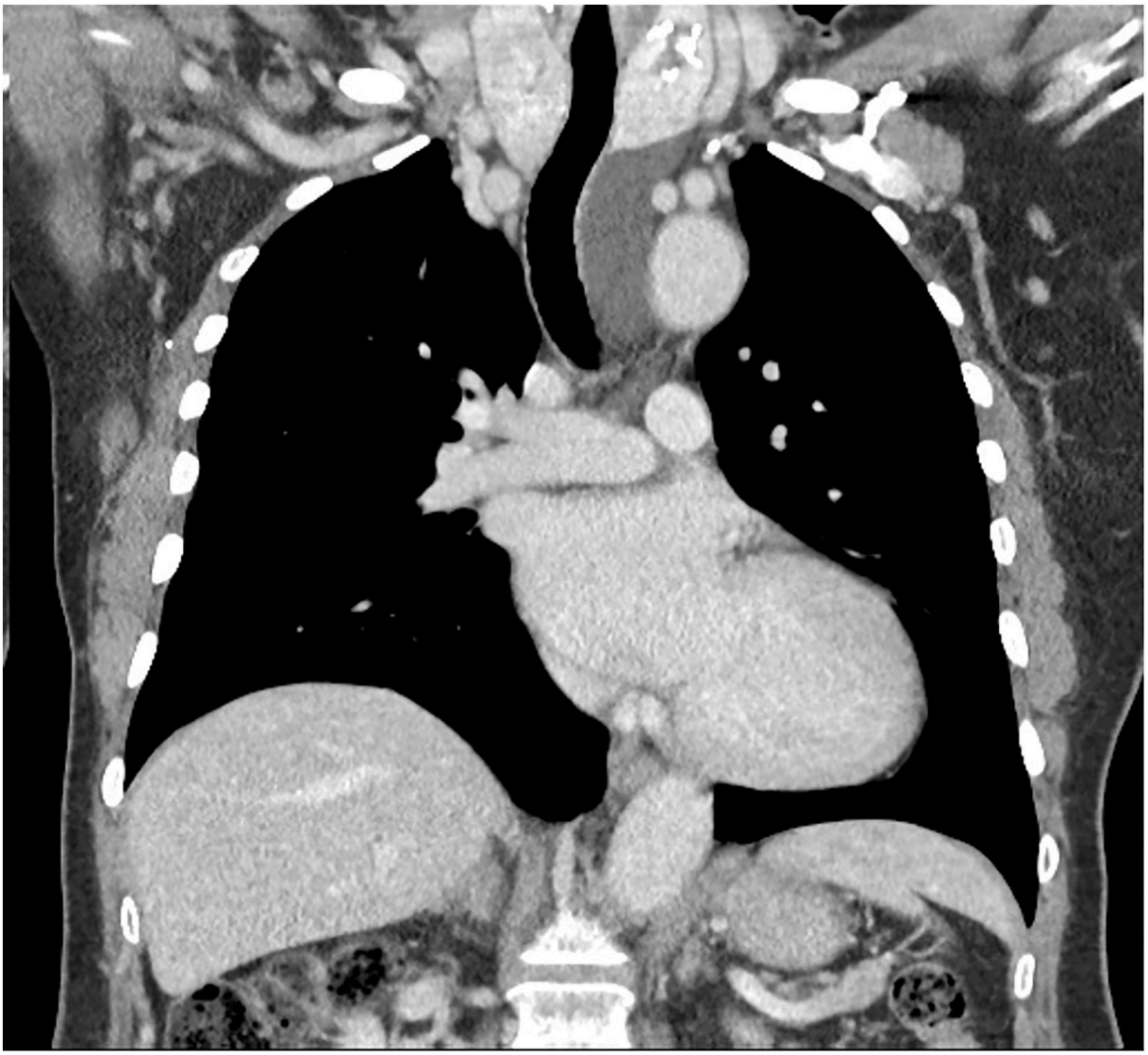

Fig. 1. This coronal CT scan of the chest demonstrates a low-attenuation left paratracheal cystic mass extending from the thoracic inlet down to the aortico-pulmonary window; in close association with the inferior pole of the left thyroid lobe but likely distinct. In addition, a multinodular goiter with cystic and solid nodules with stable calcification of left thyroid lobe and a slight retrosternal extension of right inferior thyroid pole are shown.

2. Presentation of case

A 65 year-old female with a recent diagnosis of substernal goiter requiring urgent thyroidectomy presented to our otolaryngology clinic for a second opinion. The patient was an otherwise healthy 65 year-old female with an asymptomatic anterior neck mass. She reported no dysphagia, odynophagia, shortness of breath, and no symptoms of either hypo- or hyperthyroidism. Her medical history was notable only for papillary carcinoma of the breast. Her family history was notable for head and neck squamous cell carcinoma but negative for endocrine neoplasia or other disorders. On physical exam, she was noted to have a fullness in the anterior neck extending to the sternal notch. Initial evaluation suggested a bilateral goiter with substernal extension. The exam was not concerning at this time for a malignant or infectious process. Although an asymptomatic thyroid enlargement could typically be observed, the substernal extension required further evaluation.

A computerized tomography (CT) scan of the neck was ordered, and this demonstrated an enlarged thyroid, multinodular goiter, and a right airway deviation. A lesion on the left lobe, with mixed cystic and solid components extending below the aortic arch, was noted. The patient was advised that she could undergo a surgical resection or an ultrasound-guided fine needle aspiration (US-FNA) to further characterize the lesion and alleviate any minor compressive symptoms. The patient opted for US-FNA of the largest thyroid nodule as well as decompression of the largest cyst. Using FNA, 15 mL of non-bloody clear fluid were removed from the most dominant thyroid nodule. The fluid was negative for malignancy, and most consistent with a benign colloid nodule. The patient was advised that this was not definitive treatment, and was instructed to return to the clinic in one year or when compressive symptoms developed.

One year later, the patient had begun to experience minor dysphagia, but no other symptoms, and a repeat CT demonstrated further inferior extension of the cyst. The patient was again given the option of surgical resection or continued observation. She was advised that the cyst was not likely to recede and that surgery could become more complex if delayed. The patient again opted for observation. Another year later, now 2 years after initial presentation, she returned for another follow up visit. She still had no symptoms. However, a repeat CT scan showed a continued enlargement of the multinodular goiter, a recurrence of the with a 1 cm increase in length inferiorly, now extending to the aortic arch (Fig. 1). Given the increasing size of the cyst despite drainage and further extension into the mediastinum, the patient now opted for

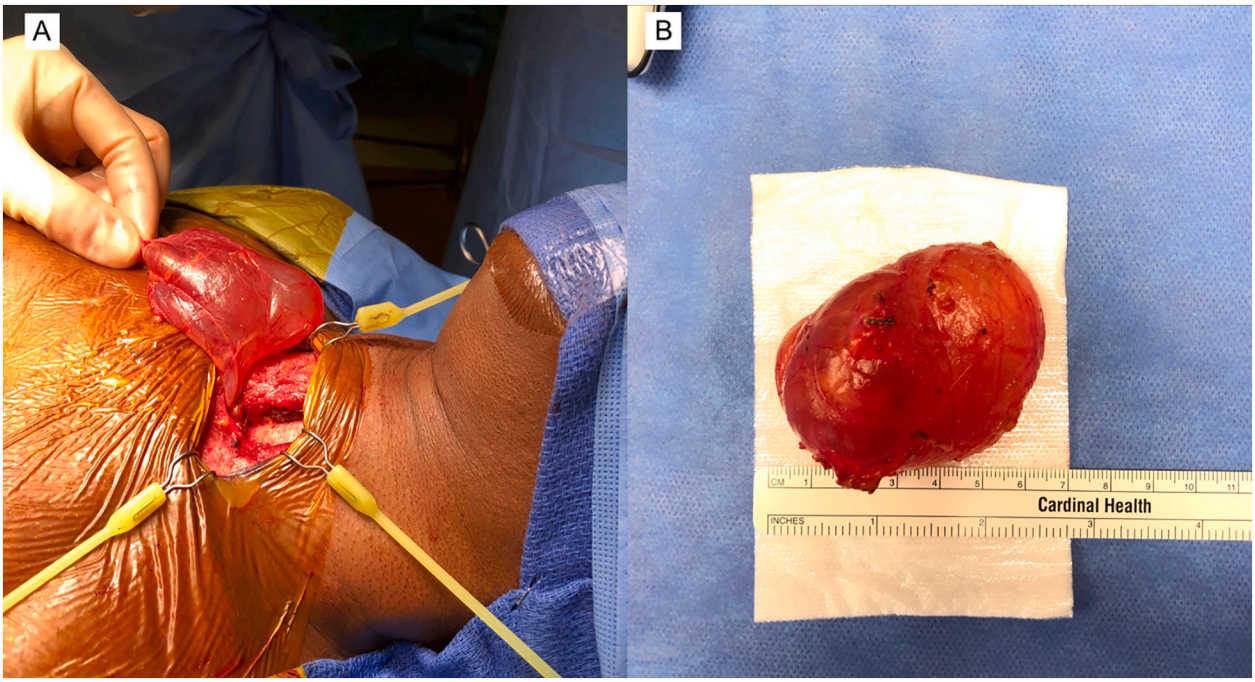

Fig. 2. (A) Gross pathological image of substernal parathyroid cyst during surgical excision. (B) Gross pathological image of cyst after removal measuring 6 cm.

definitive treatment with surgical resection.

A left thyroidectomy and cyst excision was planned as a joint surgery between otolaryngology and thoracic surgery. A standard transcervical approach was utilized. A tissue plane between the thyroid gland and the cyst was identified, and a left thyroid lobectomy was performed, providing increased exposure to cyst. The cyst was found to emanate through the thoracic inlet. It was dissected and excised in its entirety.

Pathologic examination of the left thyroid lobe revealed multiple benign, adenomatous nodules (up to 4.2 cm) and a single benign lymph node (Fig. 2). The mediastinal cyst contained clear fluid and had a smooth wall without any mural nodules. Microscopic examination revealed a fibromuscular cyst wall lined by all the elements of normal parathyroid tissue, including clear cells, chief cells, and oncocytes interspersed with occasional adipocytes (Fig. 3). No mitotic activity or nuclear atypia was present. Overall, these findings were consistent with an enlarged, cystically dilated parathyroid gland. The final diagnosis was a substernal extension of a parathyroid cyst.

The patient tolerated the procedure well and experienced mild vocal cord paresis that resolved within 4 months post operation documented by flexible laryngoscopy. At 12 months follow up, she reported full function of vocal cords without dysphagia.

3. Discussion

This report describes a 65 year-old female presenting with a large 6 cm substernal parathyroid cyst, highlighting the variable size and locations of parathyroid cysts. Since the discovery of the first parathyroid cyst by Sandstrom in 1880 and first surgical excision by Goris in 1905, only around 300 cases have been reported [1,7,8]. While the exact pathogenesis of parathyroid cysts is not entirely understood, several explanations include retention cysts, remnants of brachial clefts, degeneration of adenomas, or a coalescence of numerous small leisons [11,12]. Vestigial remnants of the third or fourth branchial cleft may occur through enlargement or colloid secretion that accumulates within a cuboidal cell lined microcyst within smooth muscle. The cyst walls can have thymus, muscle, lymphoid, or parathyroid tissue. Parathyroid cysts that are thought to derive from the degradation of a parathyroid adenoma are filled with fibrous tissue and nests of parathyroid cells. Finally, the coalescence of microcyst theory suggests that they hypersecrete and form a solitary cyst.

Parathyroid cysts can then be categorized as either functional or non-functional cysts, with non-functional cysts being more common. Non-functioning parathyroid cysts are considered true cysts with an epithelial lining while functional parathyroid cysts show histologic degeneration of an adenoma or hyperplastic gland [13]. Of note, some researchers dispute the histological similarities of functional and non-functional parathyroid cysts and instead consider them to be separate clinical entities. It has been suggested that functioning cysts lead to earlier clinical presentation and are thus smaller in size when discovered and less likely to cause compressive symptoms at the time of detection, as opposed to non-functioning cysts which may not cause symptoms aside from mass effect [13,14]. In our case, the patient experienced only primarily aesthetic symptoms initially, with a notable fullness of her neck anteriorly, only later developing mild dysphagia. It is notable that our patient experienced minimal mass effect despite obvious tracheal deviation.

These cysts can be associated with heterogenous symptoms including hypercalcemia (if functional), dyspnea, or dysphagia due to compression of the trachea or esophagus, recurrent laryngeal nerve palsy, or incidental detection on ultrasound or CT scan. To visualize these masses, imaging using ultrasonography of the neck, CT, or MRI is generally performed. Histopathologic examination demonstrates smooth cystic lesions lined with cuboidal epithelium and parathyroid cells [14]. Since differentiation of parathyroid cysts, especially ectopic cysts, from other cystic lesions of the head and neck can be difficult, FNA of the lesion should be performed. The presence of parathyroid hormone in the aspirate can establish the diagnosis of parathyroid cyst. However, in the case of a non-functional cyst, as in our patient, even an FNA will not elucidate the diagnosis.

In our case, the etiology of the cyst was unclear even after FNA examination. The patient did not have notable lab abnormalities, and the only imaging abnormality was the identification of the cyst on CT. Prior to resection, the cyst was considered most likely to be of thyroid origin, however the differential diagnosis included a cyst of bronchogenic, pericardial, or even esophageal origin. The diagnosis was only later confirmed through histological examination.

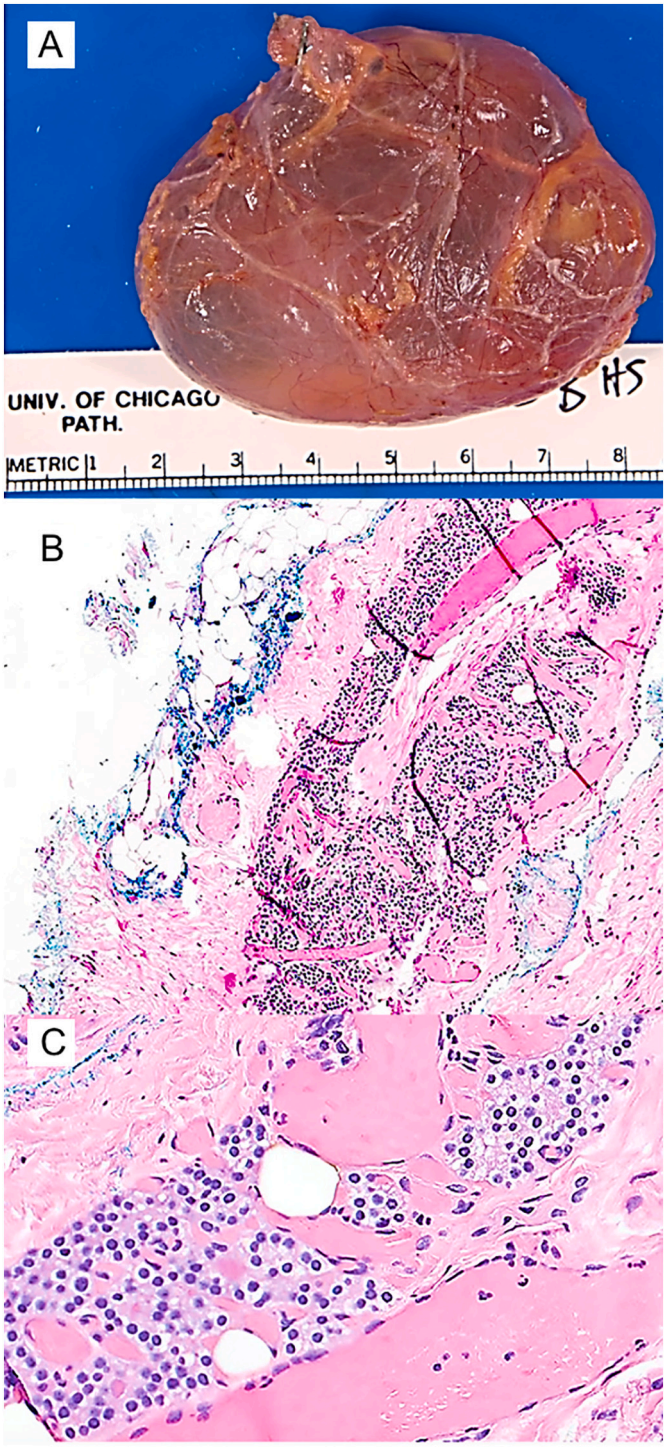

Fig. 3. (A) An intact, smooth walled cyst filled with clear fluid could be appreciated grossly. (B) The cyst wall is comprised of fibroadipose tissue lined by glandular epithelium. (C) High power examination reveals all components of normal parathyroid tissue, including chief cells, clear cells, and occasional adipocytes.

In rare circumstances, ectopic parathyroid cysts can present in the mediastinum. The varying locations of parathyroid cysts are attributed mostly to embryonic development of the parathyroid glands, which can be found anywhere in the anterior neck and superior mediastinum. Infrequently, ectopic parathyroid cysts can develop in the mediastinum. De Quervain first reported a substernal parathyroid cyst in 1925, and since then, fewer than 150 reports of mediastinal parathyroid cysts have been reported worldwide [5]. Moreover, the presentation anywhere from the angle of the mandible to the mediastinum can cause a parathyroid cyst to mimic a thyroid or mediastinal lesion [6]. Parathyroid cysts are most often confused with thyroid cysts in addition to thyroglossal duct cysts, brachial cleft cysts, thyroid adenomas, and parathyroid carcinoma [1].

Parathyroid cysts have been characterized as difficult to diagnose. CT and ultrasound are best used pre-operatively to distinguish a solid or cystic mass, but it cannot differentiate thyroid from parathyroid cysts effectively [15]. Diagnosis is based on the histopathological appearance of the cyst along with calcium and PTH levels in the fluid obtained through FNA or post-surgical excision. FNA is a non-surgical means of diagnosis and possible treatment for an easily accessible, non-functional cyst. If the cyst recurs after aspiration, however, surgery may be the best treatment option [5].

4. Conclusion

In conclusion, we present a case of a large parathyroid cyst extending into the mediastinum. Parathyroid cysts are a rare cause of neck mass in an adult, and a 6 cm substernal parathyroid cyst represents an unusual site and size for these cysts. Parathyroid cysts are not often considered on the differential of neck and mediastinal cystic lesions. However, appropriate steps, including pathology post operatively, should be taken to ensure a proper diagnosis for any cystic lesion in the neck.

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Declaration of competing interest

None.

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Ethical approval

Not applicable.

Consent

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 Editor-in-Chief of this journal on request.

Author contributions

Conception & design: All authors.
Data Acquisition: All authors.
Analysis and interpretation: All authors.
Drafting the manuscript: All authors.
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