Large retropharyngeal undescended inferior parathyroid adenoma masquerading as part of retropharyngeal goitre

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ABSTRACT

Introduction: Primary hyperparathyroidism is the third most common endocrine disorder with an incidence of 3 per 1000 in Europe. An increased prevalence coexistent parathyroid and thyroid disease has been described. A combination of a huge goitre with a scan negative undescended retropharyngeal inferior parathyroid adenoma is presented, which is unusual and currently absent from published literature. Case Report: A 71-year-old female with asymptomatic primary hyperparathyroidism and compressive symptoms from a very large toxic multinodular goitre is presented. Localization studies failed to identify a parathyroid adenoma. Computed tomography scan showed the left thyroid lobe to be larger with a significant retropharyngeal component, extrathoracic tracheal compromise and minor retrosternal extension. A combined total thyroidectomy and parathyroidectomy via a cervical approach was performed. During mobilization of the highly developed superior pole of the left thyroid lobe, a separate retropharyngeal structure was identified. This structure, measuring up to 63 mm, was recognized as a very large adenoma of what we interpret as a non-descended left inferior parathyroid gland. Histology showed a multinodular goitre with an incidentally found 0.2 mm papillary thyroid carcinoma and hyperplasia of all three parathyroid glands, including the very large (21 g) non-descended left inferior gland. Conclusion: We believe this case to be unique in published literature given the huge goitre, unusual nature of the parathyroid disease and coincidentally found microcarcinoma. The primary value of this case lies in the illustration of the difficulties of parathyroid localization in the presence of a large goitre.

Keywords: Hyperparathyroidism, Parathyroid, Parathyroidectomy, Thyroid cancer, Thyroid carcinoma, Thyroid surgery

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INTRODUCTION

Primary hyperparathyroidism is the third most common endocrine disorder [1] with an incidence of 3 per 1000 in Europe [2]. Single gland adenoma is the most common cause accounting for 75–89% of cases [3]. An
increased prevalence of parathyroid adenomas in thyroid disease has been described [4]. Presurgical localization of adenomas with ultrasound imaging and Tc99m-sestamibi scans is used widely with sensitivities of up to 91% for both techniques combined [5]. This case report presents a combination of a huge goitre with a scan negative undescended retropharyngeal inferior parathyroid adenoma in the context of a huge goitre, which is unusual and currently absent from published literature.

CASE REPORT

A 71-year-old female was referred with biochemically proven primary hyperparathyroidism (corrected calcium of 3 mmol/L, parathyroid hormone of 40 pmol/L, 24-hour urinary calcium of 13.3 mmol) associated with a very large symptomatic, toxic multinodular goitre managed with carbimazole 5 mg. The patient had no symptoms attributable to primary hyperparathyroidism.

Examination revealed a very large goitre, more pronounced on left than right, with distension of the external jugular veins suggestive of thoracic inlet compression. Parathyroid localization studies in the form of ultrasound neck and SestaMIBI failed to identify a parathyroid adenoma. Computed tomography scan of neck and upper thorax showed the bilateral thyroid goitre with left lobe larger than right and with extrathoracic tracheal compromise, a large left retropharyngeal component and minor retrosternal extension down to the level of the left brachiocephalic vein (Figure 1).

A combined total thyroidectomy and parathyroidectomy via a cervical approach was performed. The right thyroid lobe was mobilized first, during which an enlarged right superior parathyroid gland was identified and removed. The right inferior parathyroid gland was left in situ. The left lobe of the thyroid was then mobilized and a pathological-looking classically positioned but abnormally enlarged left superior parathyroid gland was found and safely removed. Subsequently, during mobilization of the highly developed superior pole of the left thyroid lobe, a separate retropharyngeal structure, superomedial to the left lobe, was encountered and mobilized. This structure measuring 6.4x4.2x1.3 cm appeared to be separate from the thyroid and was recognized as a very large, adenoma of what we interpret as a non-descended left inferior parathyroid gland (Figure 2).

The histology revealed a large multinodular thyroid containing an incidental 0.2 mm papillary microcarcinoma, confined to the thyroid and completely excised. All excised parathyroid glands were hypercellular, including the very large undescended left inferior gland of 21 g (Figure 3). Postoperative recovery was uneventful and the patient was discharged home on day one with normalized biochemistry (corrected calcium 2.58 mmol/L and PTH 1 pmol/L).

The patient was reviewed 14 weeks after surgery. She was well with a normal voice and a nicely healed scar. Her biochemistry revealed a normal calcium corrected calcium level of 2.5 mmol/L and a normal PTH level of 4.5 pmol/L.

DISCUSSION

Single gland adenoma is the most common cause of primary hyperparathyroidism accounting for 75–89% of cases [3]. Multigland disease is found as hyperplasia or multiple adenomas in approximately 5% and 4% respectively and parathyroid carcinoma is rare accounting for under 1% [6]. As in this case, the histological distinction between parathyroid adenoma and hyperplasia can be challenging in the absence of categorically normal parathyroid tissue associated with the enlarged parathyroid [7]. Multi gland adenomatous disease may be associated with a genetic syndrome such as MEN1 or, less commonly, 2A. This patient had no personal or family history to suggest a genetic disorder but, in the light of her unusual constellation of pathologies, underwent genetic testing and was not found to harbor a recognized mutation.

Figure 1: Computed tomography scan showing a very large multinodular goitre and supposed retropharyngeal extension subsequently found to be huge parathyroid adenoma (marked with an arrow).
A typical parathyroid gland weighs approximately 50 mg making the excised glands in this case many hundreds of times the size of a non-pathological gland. Giant parathyroid adenoma is defined as weighing ≥95th centile or ≥35 g [8]. Both functioning and non-functioning giant adenomas have been reported [9, 10]. Embryologically, the inferior parathyroid glands are derived from the 3rd pharyngeal arch along with the thymus and descend with it after the 5th week, to lie in the inferior neck and superior thorax respectively. The superior parathyroid glands are derived from the 4th pharyngeal arch and descend with the thyroid gland [11]. The large retropharyngeal parathyroid is this case was almost certainly a non-descended inferior gland.

Coexistent thyroid and parathyroid pathology is not unusual with rates of synchronous parathyroid and thyroid surgery in patients with primary hyperparathyroidism reported in up to 29% and synchronous pathology in 92% of those [4]. Hyperparathyroidism and papillary thyroid cancer has also been reported in several individual cases [12] although most as in our case are likely to be incidental papillary thyroid microcarcinomas that are endemic.

The parathyroid pathology in this case was not seen preoperatively on either of the two standard imaging modalities used for parathyroid localization in our department nor interpreted as a possible enlarged retropharyngeal parathyroid on computed tomography scan. The sensitivity of ultrasonography in detecting single parathyroid adenoma ranges from 57–87% [13]. Parathyroid ultrasonography has been reported to appear to more suitable for identifying a concomitant thyroid carcinomas [14]. However, it is largely dependent on the experience of the operator and the detection of multiple nodular thyroid diseases and conditions in silent areas, such as the mediastinum, tracheoesophageal groove, and retroesophageal region, remains unsatisfactory [15]. The radiopharmaceutical technetium-99m methoxyisobutylisonitrile (99mTc-MIBI or 99mTc-sestamibi) has been used for parathyroid imaging since 1989 as a complement to parathyroid planar imaging. A systematic review reported the sensitivity and specificity of parathyroid scintigraphy with 99mTc-MIBI. The parathyroid scintigraphy was found to be 45%/94% for parathyroid localization in primary hyperparathyroidism though the reported sensitivities ranged from 39–90% [16].

CONCLUSION

This case is unique in published literature given the huge goitre, unusual nature of the parathyroid disease and coincidentally found thyroid microcarcinoma. The primary value of this case lays in the illustration of the difficulties of parathyroid localization in the presence of a large goitre and underlines the need for the surgeon to be alert to unexpected operative findings even when previous abnormalities that can account for the biochemistry have been identified.

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Author Contributions

Agata M. Plonczak – Substantial contribution to conception and design, Drafting the article, Revising critically for intellectual content, Final approval of the version to be published

Aimee N. DiMarco – Substantial contribution to conception and design, Revising the article critically for intellectual content, Final approval of the version to be published
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