

A case of persistent hyperparathyroidism after
conventional neck exploration: an unexpectedly simple
solution to a difficult problem

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A 57-year-old man was referred to the University Medical Center Utrecht with persistent hypercalcemia after a bilateral neck exploration for primary hyperparathyroidism (pHPT). The operative notes were reviewed and showed that a thorough and systematic procedure had been performed. During the initial exploration a normal superior parathyroid gland had been identified on the right side of the neck; this was left in situ. A second, slightly enlarged, inferior gland at the same side was resected. On the left side of the neck the superior gland was judged to be normal, but notwithstanding all efforts including thymectomy and thyroidotomy, the inferior gland on this side could not be found, whereafter the exploration was discontinued. Persistent hypercalcemia postoperatively indicated that the causative adenoma had not been identified. Upon referral to our hospital the diagnosis pHPT was confirmed biochemically. Since the patient was symptomatic (nephrolithiasis) with elevated serum calcium levels of 2.95 mmol/L (normal range 2.20-2.60) the indication for reoperation was unavoidable. The initial work-up, as usual for reoperative cases in our institution, consisted of Doppler ultrasonography (US), spiral computed tomography (CT), magnetic resonance imaging (MRI), thallium201-technetium-Tc99m pertechnetate scanning (ThTc) and selective venous parathormone (PTH) sampling. Relying upon the information gained at first exploration, the test results were studied with special attention for the left side of the neck and mediastinum. None of the test results showed any evidence of an adenomatous lesion in the mediastinum. Selective venous PTH sampling showed increased levels of PTH at multiple levels on the right side and only one selective high level of PTH on the left side of the neck. Because this was contrary to what we expected, the possibility of a third gland on the right side was considered. The only positive added value of venous PTH sampling was the presumed cervical (and not mediastinal) location of the lesion. MRI did not show any evidence of an adenoma. Ultrasonography was negative except for one lesion ventral to the carotid sheath at the level of the mandibular angle and the hyoid bone on the left side. The location of this 2-cm oval shaped lesion was typical for a lymphnode. However its intense Doppler flowpattern was atypical for the ultrasonographic characteristics of lymphatic tissue. Therefore, we decided to complete the analysis with fine needle aspiration. Microscopy irrefutably proved the nature of the mass: parathyroid tissue. Revision of the CT images revealed an ectopic adenoma at the location indicated by US, which had been missed during the first evaluation (**Figure 1**). Since the lesion was now proven to be the fourth (missing) parathyroid gland we focused on this lesion alone. The operative plan consisted of a local procedure at the indicated site. A small incision was made at the ventral border of the sternocleidomastoid muscle at the level of the hyoid bone. After only a few minutes of exploration a well vascularised, red-brownish, oval shaped lesion (macroscopically a parathyroid adenoma) was found in the direct vicinity of the submandibular gland. Once the lesion was resected the exploration was terminated. Postoperatively, the calcium level had returned to normal on day one. Histological examination of the specimen



Figure 1 Cross section image of Computed Tomography showing an adenoma at the level the hyoid bone. Adenoma (A), hyoid bone (H), common carotid artery (CC), internal jugular vein (JV), facial vein (FV), sternocleidomastoid muscle (SM).

confirmed the diagnosis: parathyroid tissue. Currently, more than 6 years post-resection, the patient is still normocalcemic.

After a thorough and systematic exploration performed by an experienced endocrine surgeon, persistent hyperparathyroidism still cannot be precluded in all patients as is illustrated by this case. These complications in parathyroid surgery are mostly caused by the variability in location of the glands, rather than by the variety in the number of affected glands. The majority of ectopic parathyroids (adenomas), however, can be found in the vicinity of the lower pole of the thyroid and upper part of the thymus. A rule of thumb is that the further the distance from parathyroid to thyroid the lower the frequency of ectopic parathyroid tissue. A parathyroid adenoma located at the level of the hyoid bone, as in the case presented, is extremely rare (<0.1%)¹². This can be explained by the fact that -in contrast to the superior parathyroid gland whose position remains relatively constant- the inferior parathyroid gland travels a long distance during fetal development

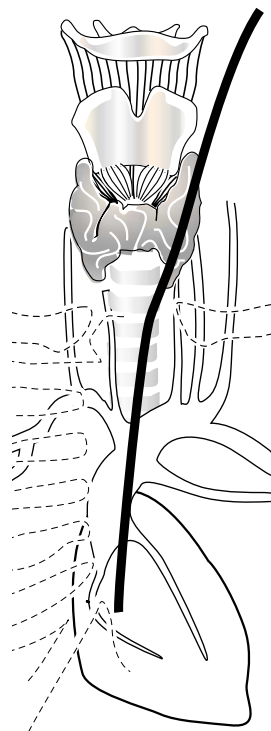


Figure 2 *Descent of the inferior parathyroid gland. The inferior parathyroid gland migrates during fetal life and may end up anywhere along the path of descensus (black line).*

Adapted from Kaplan EL. Thyroid and parathyroid. Chapter 38. Schwartz SI, ed. Principles of Surgery, 5th edition. New York: Mc Graw-Hill, 1989; 1613-1685.

together with the thymus and may thus be found anywhere from the angle of the jaw to the pericardium (**Figure 2**). In a case when the migration of the parathyroid gland is interrupted during development one speaks of a non-descended parathyroid.

The quality and reliability of imaging modalities have improved substantially over the last decade. In this case the missing adenoma was ultimately found on US performed by an experienced, persistent radiologist. All other localizing studies were negative, but for CT. The adenoma was initially overlooked in the CT examination because of its rare ectopic position, rather than its radiological characteristics. The selective venous PTH sampling was inconclusive. With hindsight, the combination of US and CT convincingly localized the adenoma, even in a most surprising position.

In this case the transition from failure to success was realized by a minimally invasive, local procedure relying upon state-of-the-art imaging technology and a direct approach to the lesion. Based on the success of this relatively simple and quick procedure we wondered whether such a direct, minimally invasive strategy could be utilized for the 'standard' case of pHPT as well. We speculated that it might serve as an alternative for the conventional neck exploration. The results of this speculation are reported in this thesis.

References

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- 2 Gaz RD, Doubler PB, Wang CA. The management of 50 unusual hyperfunctioning parathyroid glands. *Surgery* 1987; 102: 949-957