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Parathyroid surgery for inherited endocrinopathies

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Hyperparathyroidism presents as a component of selected familial endocrinopathies including multiple endocrine neoplasia types 1 and 2. These people develop multiple parathyroid adenomas that often present synchronously and can require operative intervention. The goals of the operation are to reduce the functional parathyroid mass to a point that normalizes the serum parathyroid hormone level, to avoid hypoparathyroidism, and to leave the residual parathyroid tissue in a place that can be safely accessed at the time of recurrent hyperparathyroidism. Recurrence is highly likely, as any residual parathyroid tissue has the propensity to develop subsequent adenomatous growth. The 2 main alternatives to accomplish these goals are to either remove all of the evident parathyroid tissue from the neck, and to move it to an accessible position, such as the forearm, as an autograft (total parathyroidectomy and autograft), or to remove nearly all of the parathyroid tissue from the neck, leaving a single focus of well-vascularized, carefully located tissue in the neck (subtotal parathyroidectomy).

Introduction

Primary hyperparathyroidism is caused by resistance of parathyroid tissue to serum calcium suppression of parathyroid hormone secretion. This results in parathyroid hormone levels that are abnormally elevated for the serum level of calcium, and as the excess serum calcium is cleared by the kidneys, the bone stores of calcium are mobilized to maintain the abnormally high serum calcium levels. The net effect is loss of bone calcium through urinary excretion. The effects of primary hyperparathyroidism are (1) gradually diminished bone density; (2) systemic effects of elevated serum levels of calcium, including those due to muscle relaxation such as gastroesophageal reflux, constipation, and leg weakness, as well as the effects on the nervous system, such as fatigue, lethargy, mental fogginess, and depression; (3) diminished renal function due to nephrocalcinosis; and (4) renal stones due to calcium precipitation in the renal collecting system. Most primary hyperparathyroidism is caused by single parathyroid adenoma, and removal of the single adenoma is curative.

Some heritable neoplastic syndromes include primary hyperparathyroidism as a part of the syndrome. The most common and typical is multiple endocrine neoplasia type 1 (MEN-1), but primary hyperparathyroidism also occurs in multiple endocrine neoplasia type 2 (MEN-2) and hyperparathyroidism-jaw tumor syndrome.[1] In each of these syndromes, susceptible tissues, including the parathyroid glands, have an increased rate of development of neoplasms. As time passes, they risk development of multiple parathyroid adenomas within single glands, and in separate glands. As initiation of the tumors occurs at separate times in each parathyroid gland, patients often have multiple involved parathyroid glands (a decreasing number of normal glands with patient age), with tumors of different sizes in each one.[2] This led to the historical misnomer, "asymmetrical hyperplasia." These glands are not hyperplastic, and it would be coincidental for them to be symmetric. In addition, because all parathyroid tissue is at risk of

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developing tumors, these people can develop adenomas in rests of ectopic parathyroid tissue that are normally found in the thymus or carotid sheath. These rests are rarely clinically evident or significant, unless they develop a parathyroid adenoma in the setting of one of these clinical syndromes, or, rarely, as a sporadic event.

This set of circumstances leads to the necessary considerations for managing primary hyperparathyroidism in MEN-1 (which will be used as the prototypical example of heritable primary hyperparathyroidism for this discussion). If these people develop primary hyperparathyroidism, and an indication for intervention, then the planned operation must address each of the parathyroid glands and cervically accessible sites of ectopic parathyroid tissue, must anticipate that any parathyroid tissue left in place will eventually develop additional neoplasms and lead to recurrent hyperparathyroidism, and must recognize that hypoparathyroidism is a significant functional deficit for a patient.[3,4] Opting to risk permanent hypoparathyroidism for a decreased rate of recurrence is not a good option for patients.

Clinical presentation

Patients with MEN-1 hyperparathyroidism present with the same clinical features as those with sporadic disease. There are no distinguishing biochemical features. For patients who are known to be members of MEN-1 kindreds, and who undergo regular biochemical screening, hypercalcemia often develops in the third or fourth decade of life. This can present earlier or later, and it can be the presenting feature for many people. For those who present without known MEN-1, the features that can make one suspicious include young patient age with multiple gland disease and some features in the family history that suggest possible MEN-1 components.[5,6]

Patient selection

The indications for intervention in primary hyperparathyroidism associated with MEN-1 are the same as for those without the familial basis for the disease. If the patient has symptoms attributable to the hyperparathyroidism (fatigue, lethargy, depression, renal stone, and fragility fractures), then they should strongly consider operation. If they do not have clear symptoms, then criteria that were first introduced after a National Institutes of Health consensus conference in 1990 are generally used. Those criteria include young age (less than 50 years), markedly elevated serum calcium (>1 mg/dL above the upper limit of normal), or end-organ effects (osteoporosis by dual energy X-ray absorptiometry scan, or renal function impairment with elevated serum level of creatinine). For recurrent disease in this setting, the criteria are generally stricter, with evidence of functional impairment by symptoms or end-organ effect being most convincing. Once a decision to intervene is settled, then the operative strategy can be selected.

Imaging

Preoperative imaging has become common as a part of the operative planning for sporadic hyperparathyroidism. However, it is not useful, in general, for planning of initial cervical operations in familial disease. Because each parathyroid gland must be identified in the operating room for these patients anyway, the imaging does not limit the planned exploration as it does for focused procedures in the sporadic setting. For reoperations, imaging may be helpful to identify the site of recurrence and to plan the operation.

Surgical exploration

The operative exploration for a patient with known MEN-1 hyperparathyroidism proceeds in a standard fashion, no matter the selected operative strategy for maintaining parathyroid function while preparing for the expected later recurrence.

A typical incision (Figure 1) over the isthmus of the thyroid gland provides access to the central neck (region bounded by the carotid sheaths from submandibular glands to upper mediastinum) where abnormal parathyroid tissue is likely to reside. The length of the incision can be determined by the patient's body shape and by the size of the thyroid gland. Subplatysmal flaps (Figure 2) can be raised to enable a smaller incision to provide access to adjacent parts of the neck. The tracheoesophageal groove is exposed sequentially on each side as diagramed (Figure 3), demonstrating all of

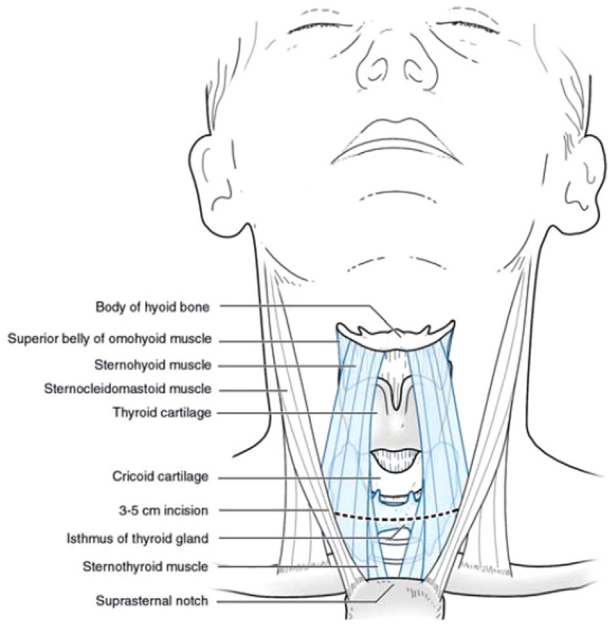

Figure 1 With the patient position supine, neck extended, and upper body raised above the level of the abdomen ("beach chair"), a transverse incision is made over the isthmus of the thyroid gland, or within a nearby skin crease. (Modified with permission from Doherty GM, Minter RM (eds): Current Procedures Surgery. New York, NY, McGraw-Hill Companies, 2010.) (Color version of figure is available online.)