Recurrent hyperparathyroidism caused by parathyromatosis

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INTRODUCTION

Persistent hypercalcaemia after surgical treatment for primary hyperparathyroidism is uncommon. It is most often the result of misdiagnosis, failure to locate a parathyroid adenoma or failure to recognize primary chief-cell hyperplasia involving all four parathyroid glands (Palmer et al, 1975). This case demonstrates another cause, parathyromatosis, which despite being rare must be considered, particularly with the increasing use of minimally invasive surgery.

DISCUSSION

In this case report the authors demonstrate that cervical re-exploration is a valuable diagnostic and therapeutic tool in treating patients with unexplained recurrent hyperparathyroidism.

Parathyromatosis is described as macroscopic nodules of hyperfunctioning parathyroid tissue scattered in the soft tissues of the neck and/or mediastinum. It is recognized as a rare cause of persistent hyperparathyroidism postoperatively (defined as hypercalcaemia occurring within 6 months of initial parathyroid surgery) and truly recurrent hyperparathyroidism (recurring after 6 months of normocalcaemia) (Sokel et al, 1993).

The two proposed theories for the phenomenon of parathyromatosis are seeding of hypercellular parathyroid glands during surgical excision and overgrowth of parathyroid rests left behind during ontogenesis (Palmer et al, 1975; Riddick et al, 1977; Fitko et al, 1990; Kollmorgen et al, 1994).

As nodules were found throughout the site of the previous neck dissection, including subcutaneous tissue and superficial to the sternocleidomastoid muscles, it was agreed that seeding at the time of primary surgery was the most likely cause of the parathyromatosis. This diagnosis was supported by the histology report which stated that the larger specimen excised was only partially encapsulated.

More commonly persistent hypercalcaemia is caused by misdiagnosis of primary hyperparathyroidism, failure to locate a parathyroid adenoma, often because of an unusual site, or failure to recognize the presence of primary chief-cell hyperplasia involving all four parathyroid glands (Palmer et al, 1975). The incidence of reoperation for persistent secondary hyperparathyroidism varies from 2.5–20% (Sokel et al, 1993).

In order to prevent parathyromatosis, a number of recommendations with regard to techniques involved in parathyroid surgery have been made. For example, it is essential the parathyroid glands should not be fractured during removal and there should be no capsular disruption from grasping with clamp, traction or suture (Sokel et al, 1993).

Recent advances in parathyroid surgery, including the unilateral approach, endoscopic procedures and minimally invasive video-assisted parathyroidectomy (Reeve et al, 2000), necessitate the removal of the parathyroid glands through either a small

CASE REPORT

In 1987, an Indian lady, aged 62 years, first presented complaining of a 1 year history of weight loss, poluria, polydipsia and lethargy. On examination of the neck there was a palpable nodule arising from the lower aspect of the right thyroid lobe. Bloods revealed raised corrected calcium of 3.62 mmol/L (normal 2.18–2.47 mmol/L). Neck ultrasound scan showed a large nodule in the right lower lobe of the thyroid, thought unlikely to originate from the parathyroid glands due to its size.

After failed conservative treatment the patient underwent cervical exploration. A bilobed nodule was removed from the right side with a total weight of 12.5 g. Two specimens were sent for frozen section and histology confirmed the diagnosis of a parathyroid adenoma (weights 0.91 g and 11.60 g respectively). The larger specimen was documented as partially encapsulated, cystic and degenerative. Post operatively normocalcaemia was achieved with resolution of preoperative symptoms.

Fourteen years later, in 2001, the lady presented with low back pain. She was found to have hypercalcaemia (adjusted calcium of 2.93 mmol/L) and hyperparathyroidism (parathyroid hormone level 46.7 pmol/L, normal values 1.1–6.9 pmol/L). Initial investigations, including a neck ultrasound scan, sestamibi scan, magnetic resonance imaging (MRI) scan of the neck and chest and parathyroid venous sampling were unable to localize the source of the hyperparathyroidism. In 2003, as a result of increasingly problematic symptoms, further localisation studies were performed. Ultrasound showed an enlarged parathyroid gland inferior to the right lobe of the thyroid. The decision was then made to proceed to re-exploration of the right side of the neck.

At the repeat exploration, aided by methylene blue localization, multiple nodules of parathyroid tissue were identified in the following areas: subcutaneous tissue (within the midline); superficial and lateral to left sternocleidomastoid muscle; Adherent to the right recurrent laryngeal nerve; within the left sternothyroid muscle; right paratracheal and paraoesophageal areas; bilaterally in the thymus glands.

All macroscopic nodules were excised and bilateral cervical thymectomies performed with no visible residual disease remaining. Histology confirmed multiple well-circumscribed nodules of parathyroid tissue similar to that originally excised and a diagnosis of parathyromatosis was recorded. The patient's subsequent calcium levels have fallen, her symptoms have improved and she is currently well.

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