CASE REPORT

Exceptional localisation of an ectopic parathyroid adenoma at the level of the hyoid bone

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SUMMARY

Primary hyperparathyroidism is characterised by high parathyroid hormone (PTH) levels and concomitant hypercalcaemia. Ninety per cent is caused by solitary parathyroid adenomas, 10% by multiple adenomas, hyperplasia and carcinomas. Less than 0.1% of the inferior parathyroid glands are ectopically located. This case is of a 54-year-old woman with hypercalcaemia 3.05 mmol/L and high PTH 22 pmol/L. Scintigraphy and CT scan of the neck in 2009 showed no signs of ectopic tissue. After treatment with Aredia (pamidronate disodium) infusion, calcium decreased to 2.73 mmol/L. In 2010 the serum calcium levels increased again to 3 mmol/L. Repeated CT showed no ectopic tissue. However, revision of the CT by the multidisciplinary team including an experienced endocrine surgeon revealed an ectopic parathyroid gland at the level of the hyoid bone, which was surgically removed in 2011. Preoperatively the PTH level was 16.8 mmol/L, postoperatively it decreased to 2.9 mmol/L, proving a successful excision. In case of suspicion of a parathyroid adenoma a CT of the neck should be thoroughly assessed by a dedicated multidisciplinary team, including the head and neck surgeon, as clinical data and radiological findings must be combined since ectopic parathyroid adenomas are a rare entity. Thereafter the adenoma can be surgically removed and the patient cured.

BACKGROUND

In case of suspicion of a parathyroid adenoma a CT of the neck should be thoroughly assessed by a dedicated multidisciplinary team, including the head and neck surgeon to exclude any ectopically located parathyroid adenomas. These can easily be surgically removed and are therefore important to diagnose.

CASE PRESENTATION

A 54-year-old woman was admitted to the hospital in 2009 at the internal medicine department because of hypercalcaemia. She had no remarkable medical history apart from a uterus extirpation in 2001. She now presented with decreased concentration, fatigue, sleeplessness, weakness of the upper extremities, nycturia, polydipsia and obstipation. Physical examination showed no abnormalities, except for an enlarged thyroid. Laboratory data showed an increased blood sedimentation rate 41 (1–20 mmol/h), an increased calcium 3.05 (2.20–2.65 mmol/L) and parathyroid hormone (PTH) of 22 (1.9–11.3 pmol/L) and a decreased vitamin D 1.25 of 41 (48–161 nmol/L). An ultrasound of the neck in August 2009 (because of suspected primary hyperparathyroidism) showed no pathology except a minimally enlarged thyroid gland with two small nodules in the left lobe. In

Figure 1 (A) A 54-year-old woman with ectopic parathyroid adenoma; CT scan of the neck 2009. HB, hyoid bone; MM, masseter muscle; IJV, internal jugular vein; ICA, internal carotid artery; red arrow, ectopic parathyroid adenoma. (B) A 54-year-old woman with ectopic parathyroid adenoma; CT scan of the neck 2011. HB, hyoid bone; MM, masseter muscle; IJV, internal jugular vein; ICA, internal carotid artery; red arrow, ectopic parathyroid adenoma.

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November 2009 a sestamibi scan using radioactive-labelled technetium was performed because of suspicion of primary hyperparathyroidism, which showed retention at the superior left thyroid lobe and no signs of ectopic tissue. The following CT of the neck showed a normal aspect of the thyroid and no signs of ectopic tissue. The ultrasound was repeated in November 2009 and showed no pathology. However, the patient was treated for hypercalcaemia with aminopropylidene diphosphonate (APD) infusion. (APD (3-amino-1-hydroxypropylidene)-1,1-bisphosphonic acid is a drug analogue of pyrophosphate and blocks bone resorption of calcium and therefore lowers calcium levels in case of parathyroid disease.)

The calcium levels decreased to 2.73 mmol/L and the patient was scheduled for a minimally invasive parathyroid excision in January, which was cancelled by the patient. She returned in 2011 once again with an increased calcium level of 3 mmol/L and a PTH of 25 pmol/L, while using Mimpara (also known as Cinacalcet, a calcimimetic agent which directly lowers PTH levels by increasing the sensitivity of the calcium sensing receptor to extracellular calcium).

A CT of the neck was repeated, which radiology reported as normal (figure 1A). Revision of this CT of the neck at a multi-disciplinary consultation showed a suspected abnormality on the left side at the level of the hyoid bone. As the patient showed clear signs of hyperparathyroid disease and the mass was identified in an anatomical region where ectopic parathyroids are found, the suspicion was raised. Retrospectively this abnormality was also visible on the CT scan in 2009, the lesion was unchanged having the same size and contrast enhancement in 2009 and 2011 (figure 1A, B). However this could have been a lymphnode or other nodular disease. Although there was no fatty centre, indicative of a lymphnode, additional imaging in the form of an ultrasound was necessary to strengthen the diagnosis. An additional ultrasound was performed to identify the suspected lesion. The colour Doppler ultrasound was used to identify a vascular pedicle favouring the diagnosis of a parathyroid adenoma and the lesion was mobile during swallowing which allows differentiation from structures such as paracarotid cervical lymph nodes or the longus colli muscle. A minimally invasive excision of the parathyroid adenoma at the level of the hyoid bone was performed. The patient was placed in supine position with the head in hyperextension. After intubation the surgeon repeated the ultrasound to determine the exact position. A small incision at the level of the hyoid bone was made, followed by incision of the platysma. The carotid artery was identified and on the

medial side the suspected adenoma was identified and removed (figure 2).

Preoperatively a parathyroid sampling was performed using our standard protocol, which means sampling PTH at several moments preoperatively and postoperatively (T0-weighted pre-operatively, T1-weighted pre-excision, T2-weighted 3 min after excision, T3-weighted 6 min after excision, T4-weighted 9 min after excision, T5-weighted 12 min after excision). The PTH showed a fast diminution with a half-life of 3 min (table 1).

One day after excision the calcium normalised to 2.26 nmol/L. Pathology confirmed that the tissue removed was parathyroid tissue, from which can be concluded that the excised gland was indeed an ectopically located parathyroid adenoma at the level of the hyoid bone.

INVESTIGATIONS

Patients with primary hyperparathyroidism may present with physical symptoms such as kidney stones, polyuria, nycturia, renal colic, bone loss, osteitis fibrosis cystica, muscle weakness, fatigue, depression, nervousness, cognitive dysfunction, psychosis, confusion, headache, peptic disease, pancreatitis, cholelithiasis, nausea, vomiting, loss of appetite, constipation, abdominal pain, hypertension and arrhythmias. This wide variety of physical symptoms makes it difficult to follow purely on clinical judgement. Biochemical and medical imaging are of paramount importance in this diagnosis.¹ High PTH and calcium levels can be determined in blood diagnostics. There are several non-invasive medical imaging possibilities; scintigraphy, ultrasound, CT and MRI and invasive diagnostics; arteriography, parathyroid venous sampling, dual-phase scintigraphy with technetium. Recent studies concerning a new diagnostic tool single-photon emission CT (SPECT; CT and technetium-99m sestamibi), have shown good results. SPECT correctly identified uniglandular disease in patients with negative sonographic finding.²

DIFFERENTIAL DIAGNOSIS

Primary hyperparathyroidism is sometimes associated with multiple endocrine neoplasia. Further diagnostics in these cases are at place for endocrine tumours in family history.³

TREATMENT

The golden standard of treatment is surgical excision of the adenoma. When surgery is contraindicated the disease can be treated by oestrogen supplements and alendronate. Supplemented by bone density monitoring at regular intervals.

Figure 2 A 54-year-old woman with ectopic parathyroid adenoma; minimal invasive excision of the parathyroid adenoma. Incision followed by spreading the platysma muscle (A), showing the macroscopic parathyroid adenoma (B). After removing the adenoma (1.5 cm) (C) the incision was closed with Monocryl 3.0 (D).

Table 1 PTH-values preoperative and postoperative

Preoperative	16.8
Postoperative (min)	
3	8.8
6	3.5
9	3.1
12	2.9

PTH, parathyroid hormone.

OUTCOME AND FOLLOW-UP

The prognosis of surgical excision is very good, 95–97% is cured. Re-exploration surgeries have a slightly lower success rate (85–90%).⁴

DISCUSSION

Primary hyperparathyroidism is characterised by high PTH levels and hypercalcaemia. In 90% of the patients this is caused by a solitary parathyroid adenoma and in 10% by multiple adenomas, hyperplasia or carcinomas.⁵ Prevalence of primary hyperparathyroidism is 0.1–0.4% and 2–3 times higher for women at an average age of 55 years.⁶ Less than 0.1% of the lower parathyroid adenomas are ectopically located.⁷ The study of Tardin *et al* included 105 patients with parathyroid adenomas of which 20 were ectopically located. The locations of the parathyroid adenomas are divided over several levels; 45% was

located paraoesophageally, 25% at the level of the cervical thymus, 20% at the level of the cervical mediastinum, 5% at the level of the anterior portion of the mediastinum and 5% at the level of the thymus.⁸ A study of Mendoza *et al* included 145 patients with parathyroid adenomas of which 13 were ectopically located. Thirteen per cent was located in the tracheal oesophageal sulcus, 31% intrathymal, 15% intrathyroidal, 7.5% in the aortal pulmonary window, 7.5% in the anterior mediastinum and 7.5% in the submaxillary region.⁹ The superior is also known as parathyroid IV, because they arise from the dorsal wing of the fourth pharyngeal pouch, they migrate minimally caudally and their final resting point is usually on the dorsal surface of the thyroid gland. This is in contradiction to the inferior parathyroid gland which arises from the dorsal wing of the third pharyngeal pouch. Of which the ventral wing becomes the thymus. The thymus then migrates caudally and medially, pulling the parathyroid glands with it. And therefore they can be found anywhere between the jaw and the pericardium¹⁰ (figure 3).

Learning points

- ▶ Less than 0.1% of the inferior parathyroid glands are ectopically located.
- ▶ Thorough assessment of a CT-scan in combination with clinical data leading to the suspicion of an ectopic parathyroid adenoma is of paramount importance.
- ▶ Localisation even at the level of the hyoid bone is possible for a parathyroid adenoma.

Figure 3 Migration pattern of the inferior and superior parathyroid glands.⁶

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Competing interests None.

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