Parathyromatosis

(Disorders of the parathyroid glands, calcium metabolism and bone)

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Parathyromatosis is a rare but clinically relevant disease that is characterized by ectopic hormone secreting parathyroid tissue. The occurrence is related to spill of parathyroid cells after parathyroid surgery in the medical history. Primary parathyromatosis has incidentally been described as result of hyperplasia of parathyroid rests from embryologic development. Continuous parathyroid stimulation, specifically in chronic renal failure, is related to increased incidence of parathyromatosis. Treatment is surgical by complete resection of all lesions.

We describe a 72-year- old woman with recurrent hyperparathyroidism, caused by parathyromatosis.

At the age of 45 the patient underwent a hemi-thyreoidectomy of the right thyroid lobe because of euthyroid goiter and a parathyroidectomy of one parathyroid gland on the right side because of an enlarged cystic adenoma. Recently, she was admitted to the hospital because of a gradual swelling of the right side of the neck and a hypercalciemic nefropathy. On physical examination a non-solid, not fixated process of 3 cm was palpated. The laboratory results showed an elevated serum calcium of 3.41 mmol/L, a normal phosphate level of 0.74 mmol/L, an elevated PTH of 346 pmol/L and an elevated kreatinin level of 147 μmol/L. An ultrasonogram and MRI scan of the neck showed a large process on the right side, adjacent to the remaining thyroid tissue. An Iodine-123 and Technetium-99-Tetrafosmin scan showed increased uptake in the same region. On surgery a large, soft tumour of 8x6x3 cm was removed. Histological examination revealed a multinodular hyperplasia matching the diagnosis of parathyromatosis. One month after surgery the serum calcium level normalized but the PTH level remained elevated (41.6 pmol/L). We now consider the use of a calcimimetic agent (cinacalcet) as a possible useful adjuvant treatment to prevent further growth.