Airway obstruction due to giant non-parathyroid hormone-producing parathyroid adenoma

ABSTRACT

We present a case of a 39-year-old female patient with acute stridor due to a large tumor located at the level of the upper third of her thoracic esophagus. Parathyroid gland tumors are unusual in the differential diagnosis of mediastinal tumors. This tumor was removed via a thoracocervical approach, which offers multiple advantages when used for tumors in this location. The eventual diagnosis on histology was parathyroid adenoma. The patient had no clinical evidence of metabolic abnormalities and her pre- and postoperative calcium and postoperative parathyroid hormone (PTH) levels were within normal limits. This case poses the interesting question of whether identification of elevated PTH levels is an absolute prerequisite for diagnosing parathyroid adenomas. It is an example of a difficult diagnostic and therapeutic problem.

KEY WORDS: Mediastinal tumor, parathyroid adenoma

INTRODUCTION

Parathyroid glands are known for occurring in aberrant positions, characteristically in the thyroid parenchyma and, less commonly, in the mediastinum.[1] The inferior parathyroid glands are more frequently found in ectopic positions than the superior pair. Adenomas of the parathyroid usually manifest with symptoms related to hypercalcemia[2] and are, therefore, one of the most common causes of primary hyperparathyroidism. There is little literature available on non-parathyroid hormone (PTH)-producing adenomas.

CASE REPORT

In February 2005, a 39-year-old female presented to a district general hospital with acute stridor and mild dysphagia; she had no voice change. She reported that she had been experiencing an irritating sensation in her throat and some dysphagia during the last 6 months and these symptoms had deteriorated significantly in the past 5 days. She was admitted and treated conservatively with dexamethasone and she improved clinically. Hematological and biochemical results were all within normal limits. She had no other medical history; she was a nonsmoker and was not habituated to alcohol.

A CT scan of her neck and chest revealed a large solid mass at the thoracic inlet, extending into the superior mediastinum to below the level of the aortic arch. It measured 4 × 2.5 cm and had a smooth contour. It compressed and displaced the esophagus to the left at the level of the first rib. The mass was separate from the thyroid gland and was seen to be compressing the trachea. The other mediastinal structures were normal and unremarkable.

A transesophageal fine needle aspiration under ultrasound guidance yielded benign-appearing thyroid epithelial cells. Detailed examination of the head, neck, bronchi, and upper gastrointestinal tract by bronchoscopy and esophagogastroscopy was unremarkable. As the position of the mass was thought to be unusual for an ectopic thyroid gland, and in order to completely exclude the possibility of malignancy, the patient was referred to our institution, the Royal Marsden Hospital, in June 2005.

We repeated a CT scan, which demonstrated the presence of a soft-tissue smooth, solid tumor in the cervical esophagus at the level of the thoracic inlet; the tumor was predominately in the right posterior lateral side, starting just below the level of the thyroid. It had features that were consistent with a leiomyoma. The esophagus was displaced to the left. In October 2005 she underwent excision of the mass via a thoracocervical approach and a partial longitudinal sternal split. The tumor was found to be located in the outer part of the right wall of the esophagus. It measured 7 × 6 × 5 cm and was soft in consistency and oval in shape with a pale...
surface. The muscular wall of the esophagus was preserved and there was no mucosal breach. The thyroid and thymus glands were identified and were normal in appearance. The patient had an unremarkable postoperative recovery and was discharged after 3 days. Histology reported small cells set in tubules, acini and focally in confluent sheets [Figures 1 and 2]. The cells were medium in size, with round to oval nuclei and moderately eosinophilic cytoplasm. There were no mitoses or cytological atypia. Immunohistochemistry was positive for PTH, AE1/AE3, CD4, CK7, and focally for chromogranin. There was no staining for synaptophysin, CD56, SMA, CD117, TTF-1, thyroglobulin, calcitonin, MB-1, or cyclin D1 [Figure 3]. The features were characteristic of parathyroid adenoma. Her serum calcium was 2.42 mmol/l preoperatively and 2.17 mmol/l postoperatively and her postoperative parathormone was 4.8 pmol/l.

**DISCUSSION**

Parathyroid adenomas are the most common cause of primary hyperparathyroidism. Most of them are solitary, though multiple adenomas do present in 6% of cases; in most cases, they arise from the inferior parathyroids. The biggest adenoma that has been reported to date measured 8 × 5 × 3.5 cm and was described as a giant adenoma. The excised tumor in our case should perhaps be considered a giant adenoma as well.

Ectopic parathyroids are known to exist and have been reported with a frequency of 7–46%. Adenomas can develop in any of the various ectopic positions. Mediastinal adenomas present in 2.7–4% of cases. Most of these unusual locations are explained by parathyroid gland embryology, and can be related to anomalous branchial cleft position.

Parathyroid adenomas without elevations of PTH or serum calcium are infrequent. We found very limited literature reporting data about parathyroid adenomas without elevated PTH levels; some of the reported tumors displayed features of cystic degeneration and hemorrhage. The fact that in our case the large mass did not produce PTH raises the question of whether all parathyroid adenomas produce PTH and, consequently, whether PTH is an adequately reliable marker for these lesions.

It is very interesting to note that when the patient was later questioned about her previous medical history, she mentioned that for several years she had had arthralgia, malaise, lethargy, and mood swings, all suggestive of primary hyperparathyroidism. This symptomatology occurring without elevation of PTH is exceptional. At her 3-month follow-up appointment she reported that all of these symptoms had completely disappeared. Her serum calcium was again found to be normal.

Surgical excision is the treatment of choice in parathyroid adenoma. In our case we had to use an unusual thoracocervical approach via a T-shaped skin incision and a partial sternal vertical split of the manubrium in order to gain sufficient...
access to the tumor. This is an uncommon approach but has the advantage of providing excellent access to upper mediastinal masses, causing less postoperative pain, and resulting in a smaller scar as it leaves the larger part of the sternum intact.

CONCLUSION

Ectopic parathyroid adenomas may be responsible for interesting diagnostic problems and may mimic other tumors. Although the majority are of small size, some tumors can reach gigantic dimensions. Elevated PTH and serum calcium are not always detected in parathyroid adenomas. Further research is needed to identify the frequency of non-PTH-secreting adenomas. We advocate the use of the thoracocervical approach for benign tumors of the upper mediastinum.

REFERENCES


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