 

CASE REPORT

# Functional ectopic cystic parathyroid adenomas: case reports and literature review

Nigora Rasulova*, Qaisar Hussain Siraj, Amir Javaid, Anwar Al-Banna

Department of Nuclear Medicine, Farwania Hospital Kuwait

## Abstract

Parathyroid adenomas are the commonest cause of primary hyperparathyroidism. About 10 percent of the parathyroid adenomas are ectopic in location. Cystic degeneration in parathyroid adenomas is seen in 4% of the cases and represents 1-2% of the cases with primary hyperparathyroidism. Hence, the combination of parathyroid cysts, which are both ectopic and functional is extremely rare. Functional parathyroid cysts can be either "silent" or associated with a wide range of clinical symptoms. We present two cases of functional mediastinal parathyroid cysts, one with and the other without clinical manifestations.

## Introduction

Cysts of parathyroid glands occurs in less than 0.001% cases of the neck mass [1]. The majority of parathyroid cysts (PCs) are non-functioning and presents as an asymptomatic nodule(s) in variable locations extending from the cervical to the mediastinal regions, with 10% of the ectopic PCs located in the mediastinum [2].

Parathyroid cysts do not have a specific sonographic appearance [3] and in some cases definitive differentiation of these rare lesions on the basis of sonographic appearance and location alone may not be possible [4]. Since the ectopic locations are variable and the results of radiographic and cytologic modalities may lead to confusion, in patients with hypercalcaemia and hyperthyroidism, dual-phase technetium-99m sestamibi ($^{99m}$Tc-MIBI) scan is the method of choice for the accurate localization of ectopic functional parathyroid cysts. The treatment of choice is complete surgical removal of the cyst and therefore pre-surgical localization of the functional parathyroid cysts by dual-phase MIBI scan can be extremely helpful to the surgeon. $^{99m}$Tc-MIBI parathyroid scintigraphy accurately localizes the tumour in 90% of cases and simplifies the surgical management [5].

We present 2 cases with ectopic mediastinal functional parathyroid cysts both with and without clinical manifestations.

***Key words***: *$^{99m}$Tc-sestamibi, SPECT/CT, parathyroid adenoma, parathyroid cyst*

***Correspondence**

Dr. Nigora Rasulova
Nuclear Medicine Department
Farwania Hospital
PO Box 18373
Kuwait 81004
Email: niga_r@mail.ru

## Case 1

A 46-year-old lady presented in the emergency department with severe abdominal pain associated with vomiting and fever. A diagnosis of acute pancreatitis was established and the patient managed conservatively. Her routine laboratory results raised WBC counts and raised CRP, and also showed high serum calcium 4.14 mmol/L (normal range: 2.2-2.6). Serum parathormone (PTH) level was raised at 25.1 pmol/L (normal range: 1-7.5). Chest x-ray showed a mass in the right mediastinum (Figure 1). Contrast-enhanced CT (Figure 2) showed a fairly well-defined cystic lesion in the right paratracheal region measuring 5.5x4.0x3.7 cm with marginal enhancement and fine internal septations .

The patient's MIBI parathyroid scan (Figure 3) showed a large doughnut-shaped lesion in the right upper mediastinum characterized by increased uptake at the periphery, which was seen to correspond to the lesion seen on CT. Pre-surgical serial PTH levels showed a rising pattern with the highest value of 125.40 pmol/L. A diagnosis of bronchogenic cyst was made and video-assisted thoracoscopic surgery performed. Excision biopsy of the lesion confirmed parathyroid adenoma. PTH level fell to 38.3 following surgery. A repeat dual-phase MIBI scan was negative (Figure 4).

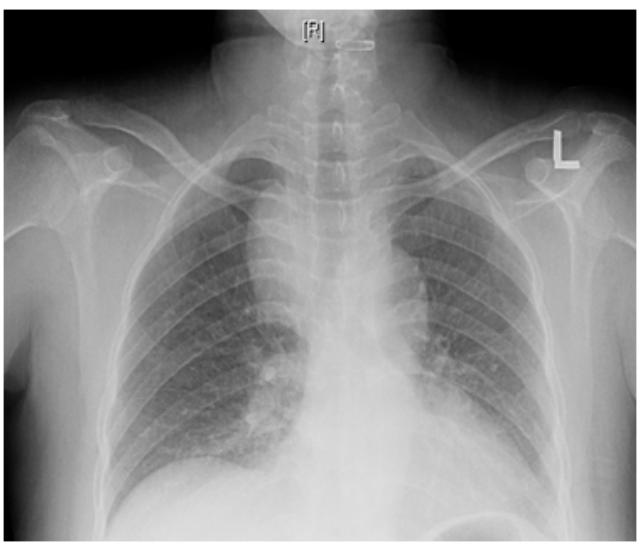


***Figure 1*** *Chest x-ray of Case 1*

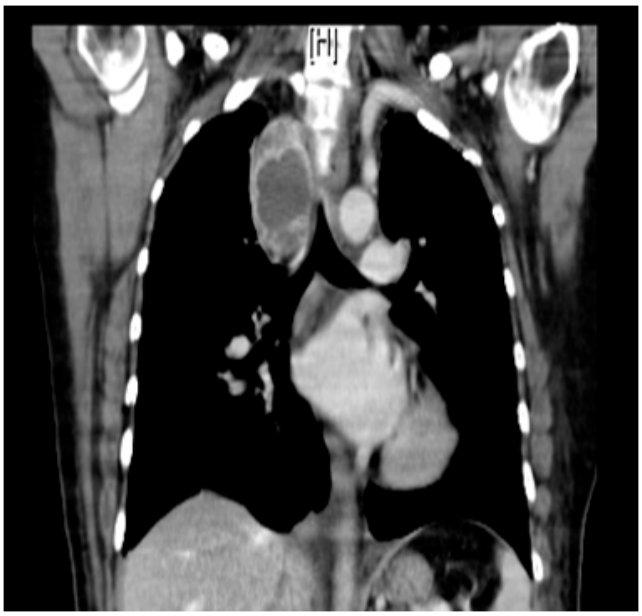

***Figure 2*** *Contrast-enhanced CT scan showing the mediastinal cyst*

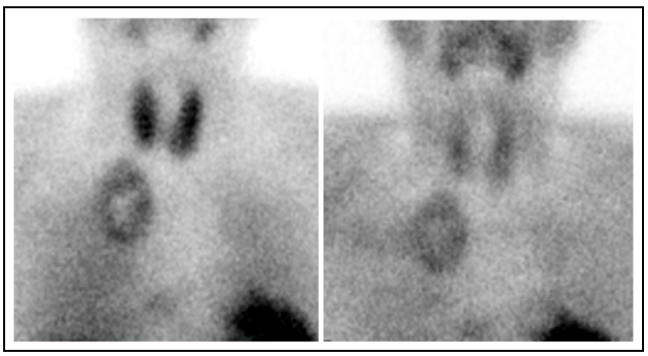

***Figure 3*** *Early (left) and late (right) baseline $^{99m}$Tc-MIBI scans showing a doughnut-shaped mediastinal lesion*

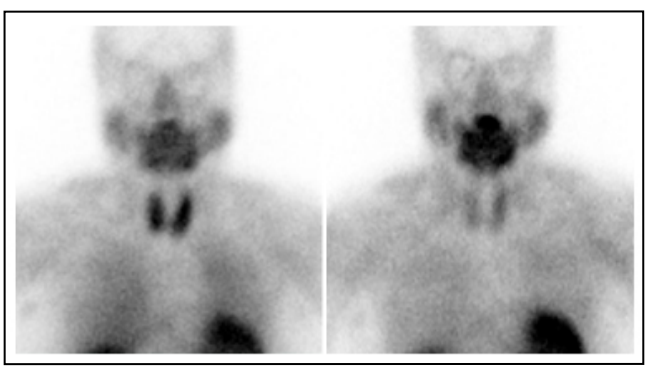

***Figure 3*** *Early (left) and delayed (right) postoperative $^{99m}$Tc-MIBI scan showing normal uptake*

## Case 2

A 16-year-old male was found to be hypercalcaemic with a corrected serum calcium of 4.14 mmol/L (normal range: 2.2-2.6). Serum parathormone (PHT) level was subsequently also found to be raised at 50 pmol/L (normal range: 1-7.5). Chest X-ray showed mediastinal widening and a right sided shadow suggesting a tumour (Figure 5).

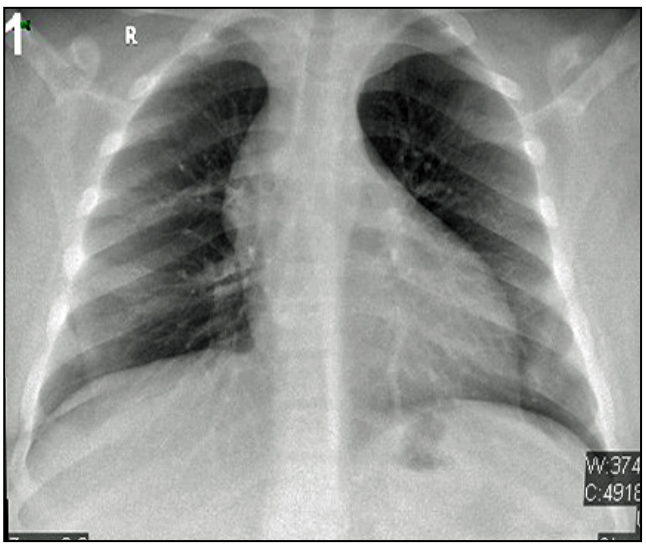


***Figure 5*** *Chest x-ray showing mediastinal widening*

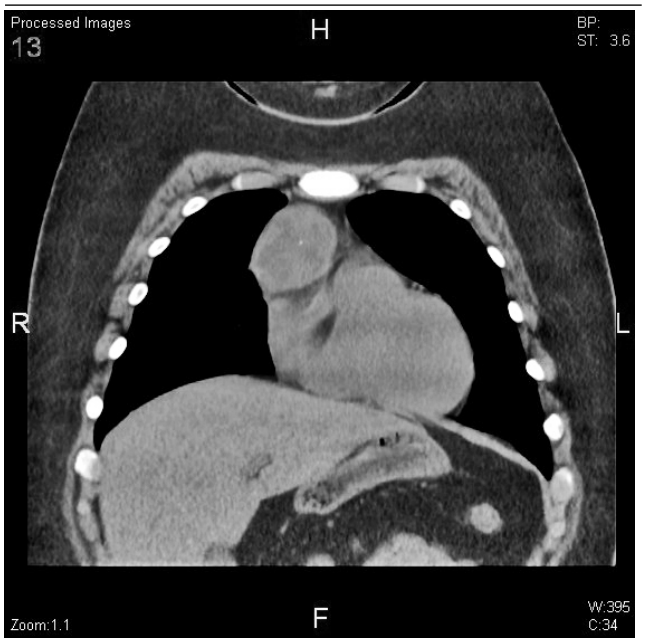


***Figure 6*** *Contrast-enhanced CT scan showing a mediastinal mass*

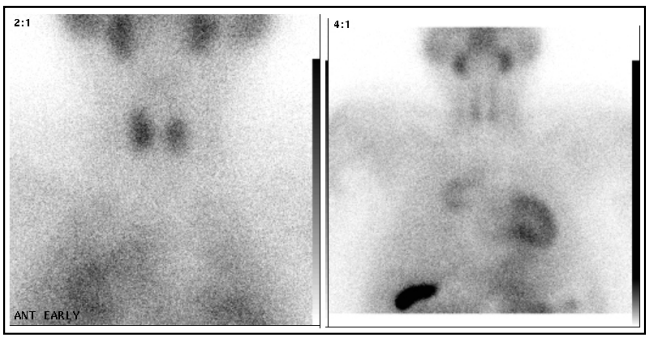


***Figure 7*** *Planar early (left) and late (right) MIBI scan showing a half doughnut lesion in the mediastinum*

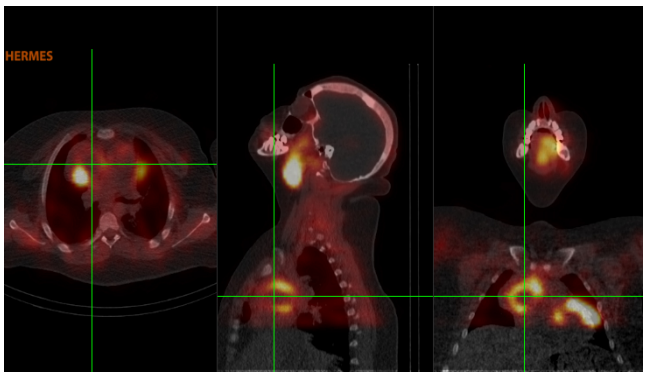


***Figure 8*** *Fused SPEC/CT scan showing a peripheral rim of increased MIBI uptake in*

The contrast CT showed a multicystic mass lesion in the mediastinum measuring 6x5x7 cm located between the sternum and the great vessels, with well-defined low-attenuation fluid density areas and tiny foci of fat density suggesting a thymic neoplasm (Figure 6).

The patient's planar $^{99m}$Tc-MIBI scan images showed a large eccentric rim of increased uptake, with the SPECT/CT scan (Figure 7) showing central photopaenia with increased uptake in periphery corresponding to the multicystic mass on the CT component (Figure 8).

## Discussion

Only about 10% parathyroid cysts (PCs) are reported to be functional associated with hypercalcaemia and primary hyper-parathyroidism which can manifest as fatigue,

weakness, polydepsia, polyuria, depression, nephrolithiasis, osteoporosis, peptic ulcer disease, abdominal pain, in some cases resulting to parathyroid crisis [6, 7].

These so-called functional parathyroid cysts are in essence parathyroid adenomas with secondary cystic changes within the adenoma. The non-functional parathyroid cysts are not associated with increased serum calcium and PTH levels but are identified by a raised level of PTH in the cystic fluid [8]. These functional PCs have reported to present as parathryroid crises [A, B]. There are only a few published case reports on this entity in the radiology literature [6, 7].

The heterogeneous clinical presentation of PCs is determined by their hormonal activity, size and location. The clinical manifestation of disease in case 1 was acute pancreatitis. Acute pancreatitis secondary to hypercalcemia is an uncommon presentation of primary hyper-parathyroidism and has been reported in 1-8% of cases [10-13]. Previous publications about acute pancreatitis induced by primary hyperparathyroidism suggest that the relationship between the two clinical conditions is not incidental [14-21]. Hypercalcaemia induces pancreatic injury via a secretory block, accumulation of secretory proteins, and possibly activation of proteases [22]. In cases of metabolic pancreatitis, in addition to standard routine management of pancreatitis, a careful monitoring of metabolic abnormalities is crucial due to the danger recurrent bouts of acute pancreatitis which may be life threatening [23]. Increased levels of serum calcium documented in a patient presenting with an episode of acute pancreatitis should raise the suspicion of primary hyperparathyroidism [24].

In Case 2 patient, who was clinically asymptomatic, routine pre-operative screening x-ray showed mediastinal widening with routine biochemical testing showing incidentally raised serum PTH and calcium levels. CT examination however was suggestive of a possible thymic neoplasm, The presence of parathyroid and thymic tissue in the same locations may be explained on embryological basis as thymus and inferior parathyroids both usually develop from the third pharyngeal pouch with the inferior parathyroids separating from the thymic tissue but remaining close to lower pole of the thyroid, whereas the thymus descends into the mediastinum. During thymic migration, small fragments of thymus may separate and attach themselves to any site along this route, with the parathyroid tissue either close to or embedded within the thymus as a result of their common origin and path of descent [25].

Functional hybrid imagine technique such a SPECT/CT with $^{99m}$Tc sestamibi of the neck and chest allows accurate preoperative localization of ectopic parathyroid adenomas, both cystic and non-cystic [26]. In our cases Tc99m Sestamibi SPECT/CT scan showed increased tracer uptake in periphery with photopenic defects at the centre confirmed by the CT component. In both cases $^{99m}$Tc-sestamibi SPECT/CT was more specific than x-ray and CT scan.

## Conclusion

Functional mediastinal parathyroid cysts are an extremely rare cause of primary hyperparathyroidism, which in our opinion can only be diagnosed correctly *in vivo* by functional radionuclide imaging. The dual-phase MIBI parathyroid scintigraphic technique is valuable not only in pre-surgical localization but also helps in the differential diagnosis.

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