

A Large Parathyroid Adenoma- A Textbook Case in Pathology

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Introduction

In 1963, I entered my residency in General Surgery at the University of Puerto Rico and Affiliated Hospitals in San Juan, Puerto Rico. At the time, Dr. Francisco L. Raffucci was Chairman of Surgery, and Dr. Leo Cuello Mainardi, Program Director in Surgery.

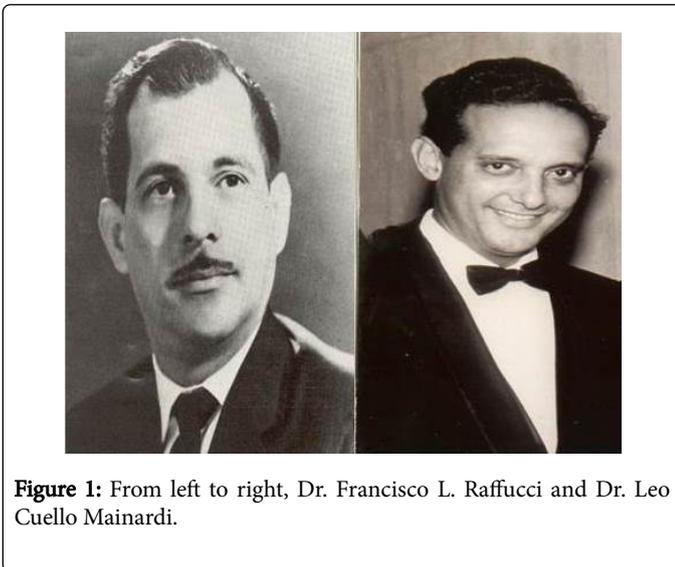


Figure 1: From left to right, Dr. Francisco L. Raffucci and Dr. Leo Cuello Mainardi.

Dr. Francisco L. Raffucci, a pioneer in cardiovascular surgery in Puerto Rico, was also involved in treating patients with portal hypertension, liver failure, and pancreatic diseases. In 1961, he removed the first parathyroid adenoma in Puerto Rico.

My interest in parathyroid surgery was prompted by an assignment from Dr. Cuello Mainardi in 1964. He required that I conduct a seminar on the topic of hypercalcemia. For this purpose, I prepared a paper for the house staff on the differential diagnosis of hypercalcemia, which includes: sarcoidosis, milk-alkali syndrome, multiple myeloma, hypernephroma, metastatic bone disease, and hyperparathyroidism. The event was pivotal to what became a life-long career as a thyroid and parathyroid surgeon.

Upon completion of my training in 1968, and after serving a two-year tour in the US Army, in 1970, I joined the faculty of the University of Puerto Rico School of Medicine's Department of Surgery.

Case Presentation

My first operation for hyperparathyroidism was performed in September 1970. The patient was a 41-year-old female who presented anorexia, weight loss, polydipsia, polyuria, and constipation, bone pain particularly in the pelvis, lacrimation, and burning sensation of the

eyes. She also had repeated urinary infections. Her calcium levels ranged from 16.4 mg/dL to 18.1 mg/dL. The chest film was interpreted as normal, the intravenous pyelogram revealed nephrocalcinosis, and X-rays of the pelvis uncovered a pelvic bone cyst. A bone scan showed generalized demineralization. The neck was explored on September 21, 1970. No neck adenomas were detected. A large lower neck and mediastinal mass was found, requiring a median sternotomy for its removal. A large 12 cm × 5 cm × 3 cm mass, weighing 113.3 grams, was removed. Although it was my first case as an attending surgeon, there was no way I could possibly miss such a large adenoma.

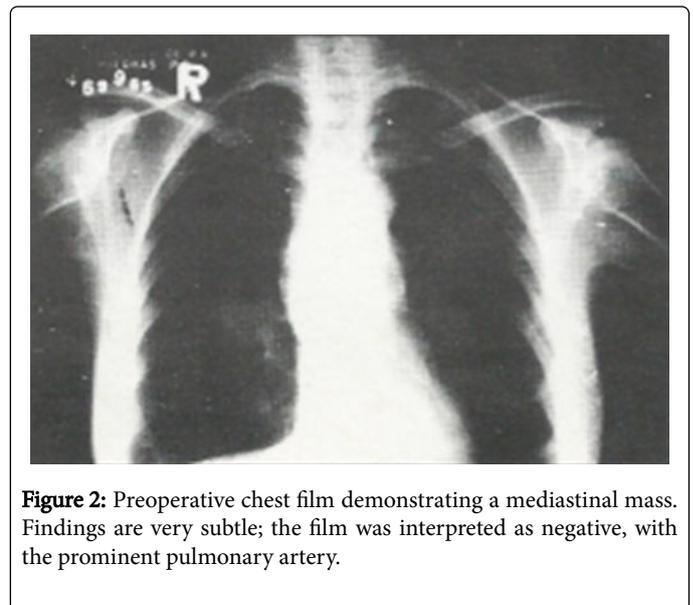


Figure 2: Preoperative chest film demonstrating a mediastinal mass. Findings are very subtle; the film was interpreted as negative, with the prominent pulmonary artery.

Histologic examination showed a monotonous field of parathyroid chief cells. A postoperative chest film confirmed absence of the previously undiagnosed mediastinal mass. The patient remained asymptomatic until 1990, when she was found to have mild hypercalcemia. Both the PTH-C terminal and the PTH intact tests were elevated as well.

To this day, this is the second largest parathyroid adenoma ever reported in the medical literature. The following a table showing the parathyroid adenomas weighing over 50 grams reported in the medical literature.

Discussion

This patient presented severe manifestations of hyperparathyroidism. At the time, in 1970, the diagnosis of hyperparathyroidism was made by excluding other causes of hypercalcemia, since PTH assays, thyroid sonograms, or CT scans

were not available as diagnostic tools. (1) Nowadays, hyperparathyroidism can be diagnosed early either with mild symptoms or no symptoms at all.



Figure 3: X-rays of the pelvis showing a bone cyst.



Figure 4: Lebsche sternal chisel and hammer.

The above photo shows the instruments used to perform the sternotomy on this patient. At that time, the electric saw nowadays used for median sternotomies was not yet in use.



Figure 5: Pathologic specimen removed from patient 1, measuring 12 × 5 × 3 cm and weighing 113 grams.

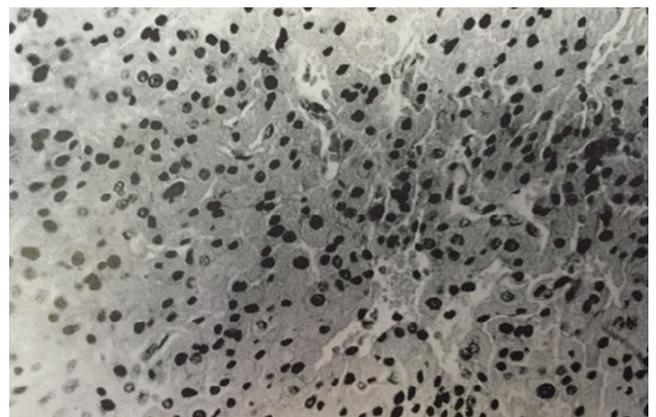


Figure 6: Histologic examination shows a monotonous field of parathyroid chief cells, compatible with adenoma.

A review of the literature revealed that the largest parathyroid adenoma ever removed weighed 120 grams; the procedure was done by Sharpe in 1939; however, it was smaller in size than the one in our case. Usually, adenomas weigh less than one or two grams. Our patient's tumor weighed one hundred times the usual parathyroid lesion: 113 grams, the second heaviest tumor reported so far in the literature.

Hyperparathyroidism is the most frequent cause of hypercalcemia in non-hospitalized patients, and the second most common cause of hypercalcemia after malignancy among those hospitalized patients. (2) In approximately 85% of cases the etiology of hyperparathyroidism is an adenoma; 10%-12% of cases are due to hyperplasia of all four parathyroid glands, and in approximately less than 3% of cases the cause is a parathyroid carcinoma. (3) Multiple adenomas are very rare (4,5). The patient in question is alive 45 years after her operation. She is now 86 years old. Her most recent calcium level is 10 mg/dL and her PTH 72.8 pg/mL. Her serum creatinine is elevated, 1.8 mg/dL, and her

creatinine clearance is low, 24.93 mL/min. She has no kidney stones. This woman still works as an assistant cook at a home for the elderly.

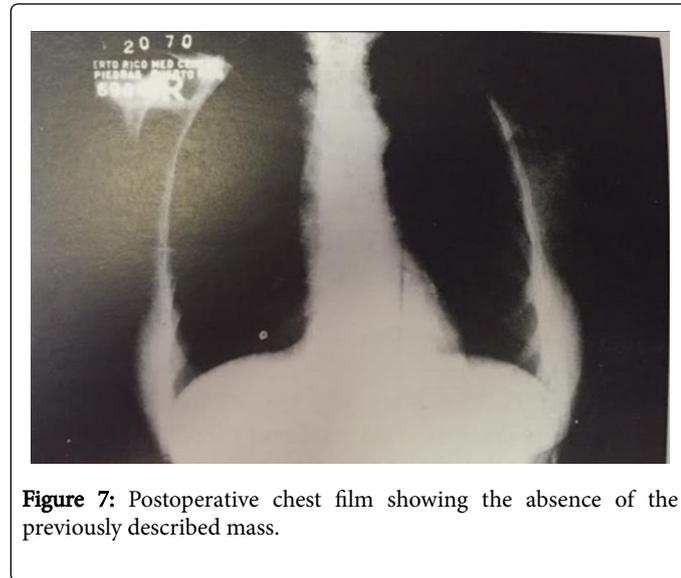


Figure 7: Postoperative chest film showing the absence of the previously described mass.

Surgeon	Year	Location	Size (cm)	Weight (g)
Dresser	1931	R	6.5 × 5.0 × 3.0	53.0
Castleman	1936	R	6.5 × 5.0 × 3.5	53.2
Earll	1969	LL	7 × 5 × 3	85.0
Snell	1936	RL	6 × 6 × 5	101.0

Vazquez	1970	M	12 × 6 × 3.5	113.3
Sharpe	1939	LU	7 × 6.5 × 4	120.0

Table 1: Parathyroid adenomas over 50 grams in weight.

Summary

We have presented a patient with severe symptoms of hyperparathyroidism, markedly elevated calcium levels, bony cyst, nephrocalcinosis, and a large mediastinal adenoma. Forty-five years after the surgery, she is now 86 years old, with normal calcium and PTH intact levels. At the time of her operation, there were no localizing tests, nor the electric saw to open the sternum. This demonstrates that in the past, good results could be obtained with the tools that were available.

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