

Giant mediastinal parathyroid adenoma

A case report



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Giant mediastinal parathyroid adenoma: a case report

A 65 years old female patient suffering from multinodular goitre and from hypercalcaemic syndrome which had been and was being treated with drugs for a number of years. The patient undergoes total thyroidectomy. Surgical neck and anterior mediastinum exploration was negative for sick parathyroids. Neck and chest TC and ^{99m}Tc sestamibi scintigraphy showed the presence of a 7 cm hyperfunctional ectopic parathyroid tissue in the postero-superior mediastinum infiltrating oesophagus. Video bronchoscopy revealed an extrinsic compression of the trachea.

The patient underwent surgery. Through right posterolateral thoracotomy and mediastinal pleura incision, the adenoma excision was carried out.

The histological examination revealed a potentially malignant parathyroid adenoma with infiltration, without overreaching the capsule.

The surgical treatment resulted in an immediate resolution of the clinical symptomatology. One year later, the follow up has shown no observable relapse with normal calcium and PTH values.

Today, the tendency to minimally invasive surgery, an accurate preoperative localization of the adenoma and the use of techniques such as intrasurgical nuclear mapping through manual gamma probes, selective angiography, venous intraoperative dosage of PTH and intrasurgical ultrasound scan.

KEY WORDS: Adenoma, Hyperparathyroidism, Mediastinum.

Introduction

Parathyroids originate in the endoderm between gestation weeks 5 and 6 and precisely the superior ones out of the fourth branchial pouch, the inferior ones out of the dorsal area of the third branchial pouch. The thymus originates in the ventral area of the third branchial pouch.

Parathyroids associated with the offshoot of their respective pouches migrate, during the embryo development, to their final destination at the end of gestation week 8.

As for the inferior parathyroidism their seat is finally found in the area in which the inferior thyroidal artery enters the thyroidal parenchyma.

The superior glands' final seat is represented by the context of the adipose tissue at the back of the upper pole of the thyroid close to the area where the recurrent nerve enters the larynx.

Since the path of such a migration is extremely variable it's easy to understand the possible glandular ectopy or the discovery of a supernumerary gland.

The different origin of the parathyroids from the third and fourth branchial pouch determines their different migration. The inferior parathyroids in fact, can migrate to the anterior mediastinum remaining more or less closely connected to the thymus, whereas the superior parathyroids can migrate to the posterior mediastinum remaining closely connected to the lateral walls of the oesophagus¹⁻².

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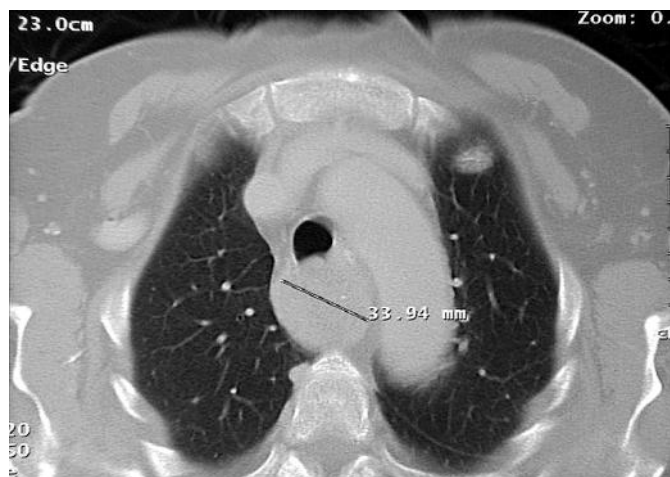


Fig. 1

Primary hyperparathyroidism arises, in most cases (80-90%) due to the presence of a single adenoma.

The parathyroid adenoma appears at a macroscopic level as brownish in colour and of elastic texture, only exceptionally weighs more than 1.5 g and features a connective capsule. The histological picture is constituted of main cells increased in volume and arranged in trabecular structures. The main characteristic of these cells is their over-produc-

tion of parathormone (PTH); the clinical consequence - though not always present - is an increased concentration of seric calcium in the blood. The therapy in most cases is surgical and the prognosis is excellent.

Clinical case

A 65 years old female patient suffering from multinodular goitre presented an hypercalcaemic syndrome. The clinical picture was characterized by migraine, anxiety, depression, dysphagia, chronic bilateral nephrolithiasis, moderate renal failure, erosive gastritis, and osteoporosis which had been and has being treated with drugs for a number of years. Calcium plasmatic level would fluctuate between 12.5 and 15 mg/dl and the serum PTH reaching values up to 1500 pg/nl.

The patient therefore undergoes total thyroidectomy and surgical exploration of the neck.

This exploration led to the discovery of four parathyroids which looked in aspect and size.

Negative was the extensive cervical exploration of the thymic cavity.

Post-surgery calcium and PTH plasmatic values were observed to remain unchanged.



Fig. 2

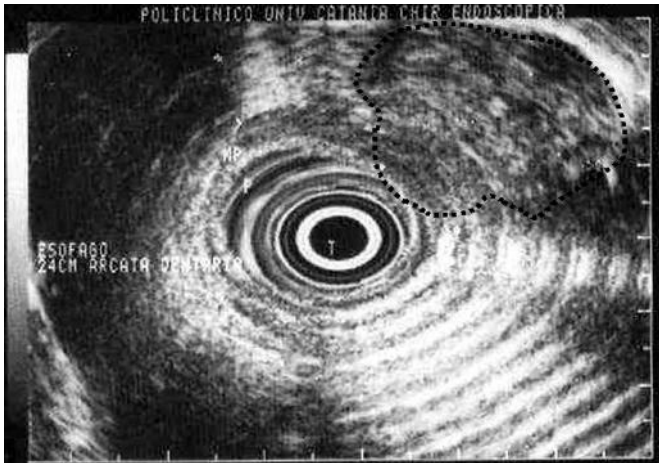


Fig. 3

An in-depth diagnostic analysis followed, making use of the neck and chest CT, showed the presence of an egg-shaped anomalous formation with a diameter reaching about 70 mm in its widest area, localized in the postero-superior mediastinum and infiltrating the oesophagus (Fig. 1, 2).

⁹⁹Tc sesta-mibi scintigraphy (Fig. 3) confirmed the presence of a massive binding area of the radioactive drug in the mediastinal site, therefore relating to hyperfunctional ectopic parathyroid tissue.

The suspected infiltration of the mediastinal structures led us to complete the study with the oesophagogastroscopy which highlighted, at about 24 cm from the dental arch, an extrinsic compression of the lower middle third of the oesophagus, covered with a normal looking mucosa. Also the ultra-sound endoscopy (Fig. 4) confirmed, in the same site, the presence of an hypoeco-

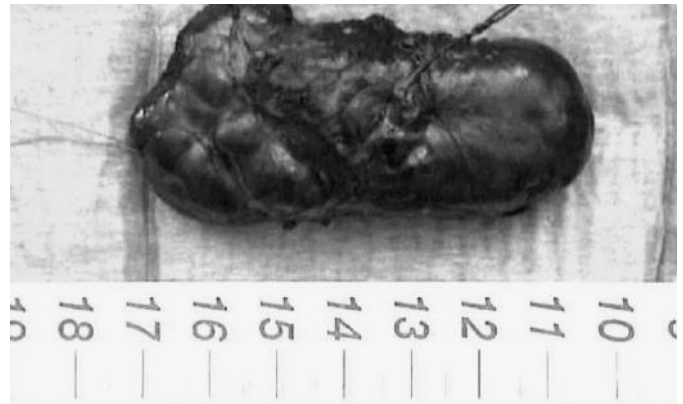


Fig. 5

genic formation starting from the muscular layer of about 3.5 x 3.5 cm.

Video bronchoscopy revealed an extrinsic compression on the pars membranacea of the trachea far away about 2 cm from the carina without signs of infiltration.

The patient underwent a surgical operation under general anaesthesia with selective endobronchial intubation.

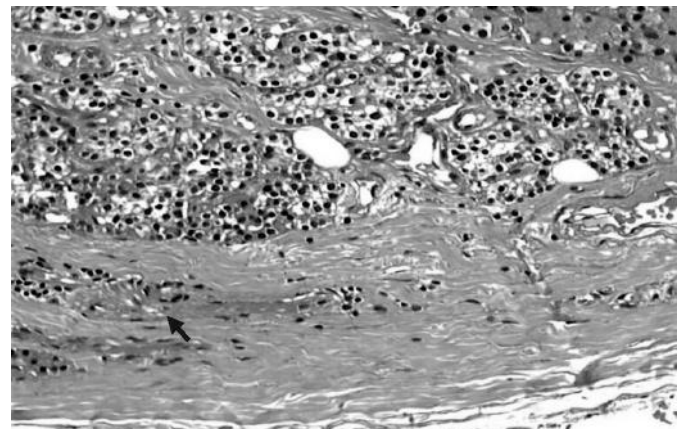


Fig. 6

After the right-side posterolateral thoracotomy and due to its relative pulmonary exclusion it was highlighted the new formation (Fig. 5) emerging from the retro-azygotic and paracaval mediastinal side, the extension of which reaching the anonymous venous trunk of ipsilateral side. The mediastinal pleura incision highlighted the presence of a cleavage plane which persisted throughout the extension of the lesion although without infiltrations of neighbouring structures.

The excision was carried out with blunt and careful dissection of the adenoma, not to damage the oesophageal mucosa. The mass in fact marked a compression upon the oesophagus without infiltrating it but causing a distasis of the muscle fibres. The surgical specimen appeared

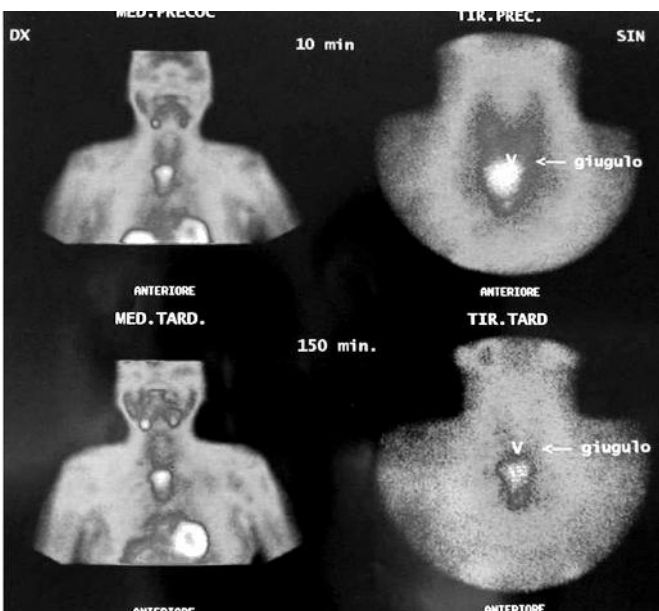


Fig. 4

red roundish and capsulated, the size of 7.1 x 4.3 x 2.8 cm and weighing 95 g (Fig. 6).

The histological examination revealed a parathyroid adenoma potentially malignant with infiltration, without overreaching the capsule.

The surgical treatment resulted in an immediate resolution of the clinical symptomatology.

The serous monitoring of the PTH values showed rapid decrease right after the operation, accompanied by an abnormally low blood calcium levels, necessitating immediate pharmacological corrective measures.

One year later, the follow-up has shown no observable relapse of the disendocrine condition with normal calcium and PTH values.

Discussion

A literature review shows that the chosen treatment for primary hyperparathyroidism is doubtless surgical, which is successful in 95% of the cases.

Drug treatment therefore should be chosen only for those patients extremely debilitated and pre-operatively for patients showing very high calcium levels. Such values can be checked through rehydration and administration of thiazide diuretics, diphosphonate and calcitonin.

Surgical treatment involves bilateral neck exploration which in 97% of the cases is the seat of the pathology³.

Pre-operative localization techniques^{4,5} should be always used being variable the final seat of the parathyroid in the neck, in the antero-superior mediastinum⁶, in the retro oesophageal area, along the carotid axes and in the thyroidal context⁷.

Today, the tendence to minimally invasive surgery, imposes an accurate pre-operative localization of the adenoma and the use of techniques such as intra-surgical nuclear mapping through manual gamma probes⁸, selective angiography⁹, venous intra-operative dosage of PTH^{10,11,12} and intra-surgical ultra-sound scan.

In 67% of the cases¹³ similar study reveals the presence of new growths bearing upon parathyroid adenoma localized in the anterosuperior mediastinum, whereas in only 2% of the cases¹⁴ such pathology is localized in the posterior mediastinal site, which is in any case inaccessible via cervicotomy.

Once identified the site, the access route is chosen. It is advisable the V.A.T.S.¹⁵ surgical approach over traditional surgical techniques^{16,9,17,18,13,19}.

Conclusion

The hyperfunctional hyperparathyroid glandular ectopy is a fairly rare event. From literature review it turns out that its occurrence is between 11-25% of all cas-

es of primitive hyperparathyroidism^{20,21} and in only 2% of instances its seat is in the mediastinum^{18,19}.

The presence of a supernumerary hyperfunctioning gland of massive size bearing upon the posterior mediastinum is certainly an exceptional event, taking - on the other hand- into account its uncertain degree of malignancy.

We do agree with most Authors that failure to detect the disease upon surgical exploration of the neck but with calcium values persistently above 11.5 mg/dl it's necessary a reevaluation of the case. Re-intervention is subordinated to localizing the gland a ⁹⁹Tc sesta-Mibi scintigraphy with CT and/or neck and thorax MNR⁵. Once the site has been identified, the chosen treatment, especially for benign lesions is the video-thoracoscopy better if the dissection is guided by either ultrasound scanning probe or gamma rays camera.

The finding of an hypersecreting gland of considerable size has to alert us to the possibility of being in the presence of a malignant neoplastic degeneration.

Typical of such neoplasia is its remarkable topically invasive character with a high percentage of relapses and few lymph node metastases.

Though agreeing with a mininvasive approach, given the site, the size and suspected oesophageal infiltration, we have chosen the thoracotomy access.

Riassunto

L'ectopia ghiandolare paratiroidea iperfunzionante è un'evenienza molto rara. Da una revisione della letteratura risulta che l'ectopia paratiroidea si riscontra nel 11-25% di tutti gli iperparatiroidismi.

Noi presentiamo il caso di una sindrome ipercalcemica conclamata (sintomi neurologici, nefrolitiasi recidivante) con assenza, all'esplorazione cervicale, di paratiroidi iperplastiche. Persistendo la sintomatologia ed i livelli plasmatici elevati di calcio e PTH, si procede ad ulteriori approfondimenti diagnostici dai quali emerge la presenza di una grossa formazione mediastinica posteriore, scintigraficamente ipercaptante, infiltrante la parete esofagea.

La presenza di una ghiandola iperfunzionante soprannumeraria, di dimensioni enormi a livello del mediastino posteriore è sicuramente un evento eccezionale, considerato l'incerto grado di malignità.

Il riscontro di una ghiandola ipersecernente di grosse dimensioni ci deve far sospettare la degenerazione neoplastica maligna. Tali neoplasie sono caratterizzate da una invasività locale molto spiccata ed alta percentuale di recidiva e, meno frequentemente, da metastasi linfonodali locoregionali e a distanza.

Pur condividendo un approccio chirurgico mininvasivo, nel caso descritto la sede, le dimensioni della lesione e la sospetta infiltrazione delle strutture mediastiniche ci hanno indotto a praticare un accesso toracotomico.

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