Parathyroid cysts: description of two cases and review of the literature

Cisti paratiroidee: descrizione di due casi e revisione della letteratura

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Summary

Parathyroid cysts are a relatively rare cause of neck swelling, and their anatomo-pathological and clinical features are still under debate. Two types of parathyroid cysts have been recognized: the non-functioning or essential forms, which are more frequent, and the adenomatous or functioning parathyroid cysts, which are rarer and cause hyperparathyroidism. Two cases of parathyroid cysts are reported and the treatment is described bearing in mind the various therapeutic procedures proposed in the literature. Attention is focused on pre-operative differential diagnosis from other masses, in the same area, and, to this end, the importance is stressed of assaying parathyroid hormone levels in the cystic fluid, obtained by fine-needle aspiration, and correlating these values with serum levels of the hormone.

Riassunto

Le cisti paratiroidee costituiscono una entità clinica relativamente rara il cui inquadramento anatomopatologico e clinico è fonte di discussione. Sono suddivise in forme non-funzionanti o essenziali, le più frequenti, e le cisti funzionanti o adenomatose, più rare e causa di iperparatiroidismo. Vengono presentati due casi di cisti paratiroidee e ne viene discusso l'approccio clinico-diagnostico e il trattamento alla luce delle più recenti proposte della letteratura. In particolare si pone il rilievo sulle possibilità diagnostiche preoperatorie con altre neoformazioni del distretto cervicale e, a questo fine, viene sottolineata l'importanza del dosaggio nel liquido cistico del paratormone, ottenuto durante lo studio con agobiopsia ecoguidata, e le sue correlazioni con i corrispondenti livelli serici.

Introduction

Parathyroid cysts are an unusual cause of neck swelling, and, indeed, only about 250 cases have been reported in the literature [1-3].

These cysts usually present in the fourth or fifth decade of life and are classified as functioning or adenomatous cysts and non-functioning or essential cysts in relation to whether parathyroid hormone (PTH) secretion is increased. A single case of malignant transformation has been described [4].

Non-functioning cysts (80% of cases) are more common in females, while the functioning forms are more common in males. While parathyroid cysts are usually solitary, there have been several reports of more extensive disease involving more than one of the parathyroid glands. This is more commonly the case for the functioning forms, as if the multiple sites were an expression of a parathyroid endocrine disorder [5 6].

The functioning cysts are generally considered the result of cystic degeneration of a parathyroid adenoma [7 8].

Various hypotheses have been advanced to explain the non-functioning forms but an unequivocal conclusion has not been reached:

- Dysembryogenic residues of the 3rd and 4th branchial pouches. However, these cysts are rarely reported in children, a fact which has further added to the criticisms of this hypothesis [9].
- Forms resulting from the fusion of several microcysts normally present in the parenchyma of the gland.
- Retention cysts of parathyroid hormone resulting from overactivity of the gland, not accompanied by an increase in plasma levels of the hormone.

These swellings are almost always found in close proximity to the inferior pole of a thyroid lobe although other cervical and mediastinal locations have been reported [10 11].

Symptoms of the non-functioning forms are limited to the those caused by compression of neighbouring structures, while the problems associated with the functioning forms are related to excessive secretion of parathyroid hormone. In the former, neck swelling may be associated with dysphagia, odynophagia,

dyspnoea, and paralysis of the recurrent laryngeal nerve [12-14]. The functioning forms may produce not only the same symptoms but also the typical manifestations of hyperparathyroidism, which are the more important symptoms in this case.
In the event of a clinical finding of a swelling, usually located in the inferior pole of the thyroid, confirmed with imaging procedures (ultrasound (US), computed tomography (CT), magnetic nuclear resonance (MNR)) to be cystic and in a juxtathyroid location, the diagnostic dilemma can, in most cases, be resolved by examination of the intracystic fluid, obtained by fine-needle aspiration. This fluid is watery and colourless in simple parathyroid cysts, whereas it is brown or reddish in the most common thyroid nodules, and yellowish and viscous in branchial cysts.
Evaluation of the intracystic level of parathyroid hormone is, nevertheless, even more important than the physical characteristics of the cystic fluid. PTH levels are increased in all parathyroid cysts independently of whether they are non-functioning or functioning [15 16]. PTH in cystic fluid is almost entirely in the form of the non-active C-terminal fragment, only very small quantities of whole PTH being present since whole PTH is rapidly broken down in the cystic fluid. In the functioning forms, in addition to the cyst, the surrounding adenomatous tissue is very active. Thus, there are the same high levels of PTH (inactive) in the cystic fluid samples but also serum PTH (active) levels are increased due to secretion by the adenomatous cells. PTH levels should be correlated with serum levels of calcium and phosphorus.

Case 1

FF, a 47-year-old male, presented with a 2-year history of dysphagia and dyspnoea, upon effort, accompanied by a tracheal whistle which had become more severe in the 2 months prior to coming to our attention.
A recent examination had revealed the swelling which had the consistency of parenchyma and appeared to involve the caudal part of the left lobe of the thyroid. Laryngoscopic findings were normal.
US evaluation of the neck showed a hypoechoic oval-shaped formation with well-defined margins in close continuity with the posterior part of the left thyroid lobe. A CT scan was performed which showed that the formation extended downwards to the jugular vein and compressed the trachea, the lumen of which was decreased by about one third.
Thyroid and parathyroid function tests failed to reveal any abnormalities.
The excised mass had the morphological appearance of a cyst, approximately 4 x 3 cm in size.
PTH assays (C-terminal fragment) showed a concentration of 0.34 ng/ml in the serum and 8.5 ng/ml in the fluid aspirated post-operatively from the cyst.
Histological examination revealed a parathyroid cyst with a capsule adhering to the residual parathyroid parenchyma and fibroadipose tissue (Fig. 1).

Fig. 1. Case 1: Histological examination reveals fibrous wall surrounding a cystic space filled, focally, with islands of parathyroid chief cells, with no evidence of necrosis, mitosis or atypia (White arrow). Focally, a monolayered lining of similar epithelial cells lined the inner surface of wall (Black arrow). H&E 100 X.

Case 2

CA, a 37-year-old female, complained of a 6-year history of a roundish swelling which, depending on the patient's position, breathing or physical efforts, emerged from the retrosternal region filling the jugular fossa. According to the patient, the mass had become larger and caused episodes of dyspnoea, with a sensation of tracheal compression. Upon examination, a roundish mass, about 4 cm in diameter, could be felt at the jugular. Under pressure, it seemed to disappear, sliding behind the sternum. CT scan revealed a mass behind the manubrium sternum. The oblong, apparently cystic, mass was connected to the inferior pole of the left thyroid gland (Fig. 2). A needle biopsy, with fluid aspiration, was performed and the contents of the cyst were submitted to cytochemical analysis. The level of PTH in the fluid aspirate was 8.9 ng/ml. The serum values were, however, within the normal range and this led us to suspect a parathyroid cyst. Excision of the mass was planned. The cyst was connected by a fibrous stalk to the lower pole of the left thyroid lobe and was closely connected to the parathyroid tissue which was preserved and left *in situ*. Subsequent histological analysis confirmed the diagnosis of a parathyroid cyst with marked giant cell inflammation (Fig. 3).

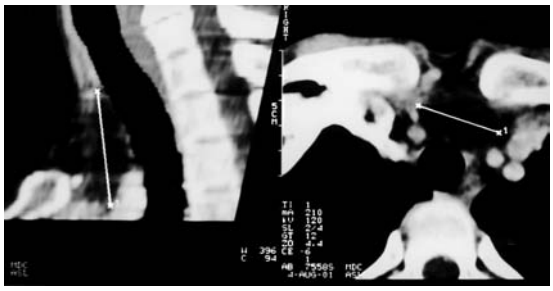

Fig. 2. Case 2: CT image revealing evidence of new cystic growth located in jugular fossa extending retrosternally. Transverse diameter 4 cm, longitudinal diameter 5 cm (reconstruction of sagittal sections on left and axial sections on right).

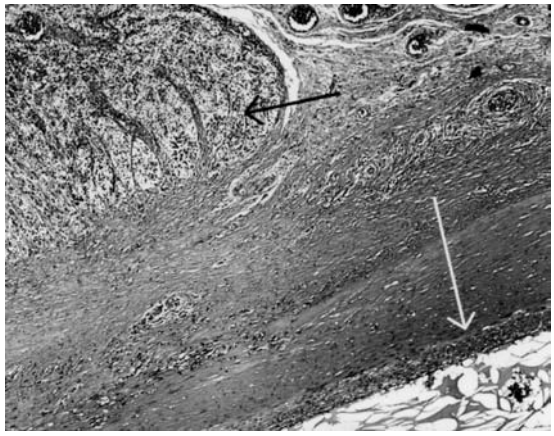

Fig. 3. Histological examination showing multiple fibrous septa; surrounding large pseudocystic spaces (White arrow) filled with inflammatory follicular lymphocyte infiltrate, with widespread giant-cell granulomas containing cholesterol clefts, and, focally, islands of parathyroid chief cells, with no evidence of necrosis, mitosis or atypia (Black arrow). H&E 100 X.

Discussion

Taking into consideration the more recent reports in the literature and our experience in the two cases presented here, the correct procedure, in the case of a juxtathyroid cystic swelling, is to perform US examination and collect a specimen, by means of fine needle aspiration. for routine cytological assessment and, if the fluid is watery and colourless, assay the various components of PTH. This information should be correlated to the serum levels of PTH, calcium and phosphorus. Diagnosis in the first case described here was made post-operatively since we carried out only a cytological examination of the intracystic fluid instead of measuring the PTH content.

CT scan is necessary only in cases with mediastinal extension or when there are symptoms of compression of adjacent structures.

Likewise, scintigraphic investigations, with double contrast agents, are not considered necessary in forms with normal PTH and calcium values. These are, however, advisable in the hypersecretory forms in order to clearly identify the site and number of lesions prior to surgery [17].

Management of the uncomplicated forms can be limited to US-guided aspiration. In some cases, aspiration can be followed by introduction of a sclerosing agent, although this procedure may lead to complications, such as recurrent laryngeal nerve paralysis, on account of neurotoxicity of the substance injected [18].

Surgical excision is always indicated in the functioning forms, in the non-functioning form in cases of recurrece after simple aspiration and in patients complaining of significant symptoms (dysphagia, dyspnoea, recurrent laryngeal nerve paralysis, etc.).

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