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CASE REPORT

Parathyroid Carcinoma: A Surgeon's Suspicion

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Abstract

Parathyroid carcinoma is a rare clinical entity. Its incidence is less than 1% of all primary hyperparathyroidism. It is a clinical challenge to distinguish between adenoma and carcinoma preoperatively; usually it is a intra operative and histological diagnosis. We present a patient with parathyroid carcinoma evaluated in our clinical setting and its management

Keywords: Parathyroid carcinoma, Hyperparathyroidism, Hypercalcaemia, Adenoma

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Introduction

Parathyroid cancer (PC) is one of the most uncommon tumors, accounting for 0.005% of all malignancies and fewer than 1% of parathyroid abnormalities. Only a few thousand cases have been recorded in the literature since De Quervain's first description in 1904 [1].

Even though most occurrences are sporadic, it has been linked to hereditary diseases such as hyperparathyroidism-jaw tumor syndrome (HPT-JT), multiple endocrine neoplasia-1 and 2A, and familial isolated hyperparathyroidism. The overlapping range of clinical and biochemical criteria makes it difficult to distinguish between benign and malignant tumors, and postoperative histology is frequently used to make the diagnosis [1].

The clinical appearance of parathyroid cancer is similar to that of primary hyperparathyroidism, and it includes hypercalcemia-related symptoms such as tiredness, bone and joint pain, nephrolithiasis, reduced glomerular filtration rate, osteoporosis, fragility fractures, and neurocognitive impairment [2].

Case presentation

A 39-year-old male residing in Tamilnadu, India with no comorbidities, referred to general surgery department by endocrinology specialist in suspicion of parathyroid adenoma causing primary hyperparathyroidism. Clinically he was asymptomatic; he had no family history of hypercalcemia or metabolic conditions. A physical examination revealed no palpable neck swelling. Laboratory investigation showed increased levels of parathyroid hormone (1098pg/ml) and calcium (11.6mg/dl). Tc99m sestamibi parathyroid scintigraphy showed well defined nodular soft tissue lesion measuring 3.1 in diameter posterior to left lobe of thyroid; features suggestive of functioning parathyroid lesion. The computed tomography (CT) scan of the neck revealed Heterogeneously differentially enhancing nodular lesions measuring 1.5cm and 1.7 cm in diameter, both the nodules are seen on the posterior aspect of the left inferior lobe of thyroid, and appear en masse as a single lobulated lesion, causing indentation on the posterior border of thyroid; suggestive of left parathyroid adenoma.

He was evaluated for surgery, after pre anesthetic and endocrinology clearance Left parathyroidectomy was performed.

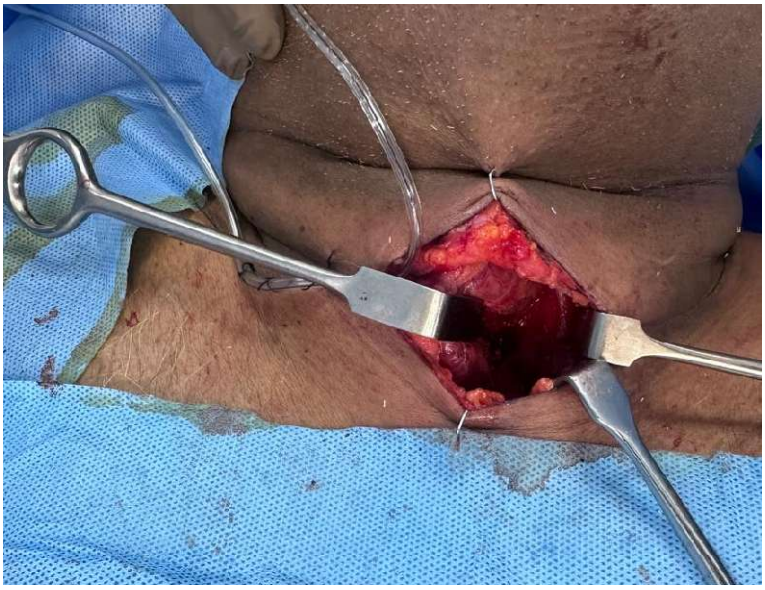

Figure 1. Site of skin incision

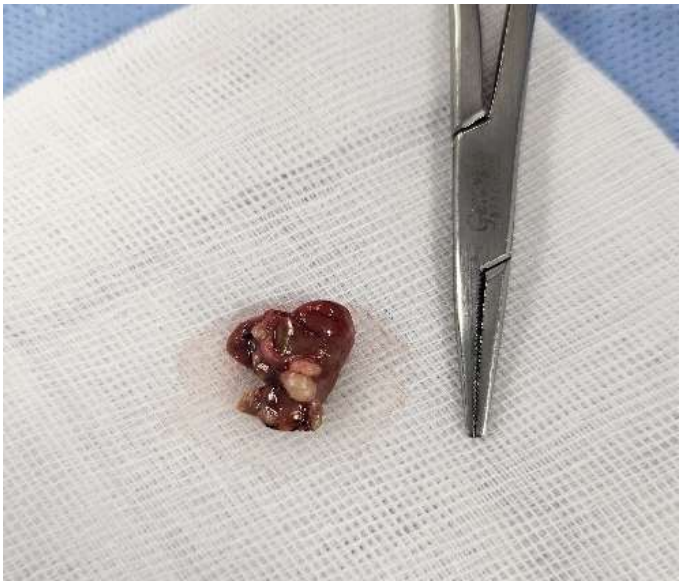

Figure 2. Postoperative specimen of parathyroid lesion

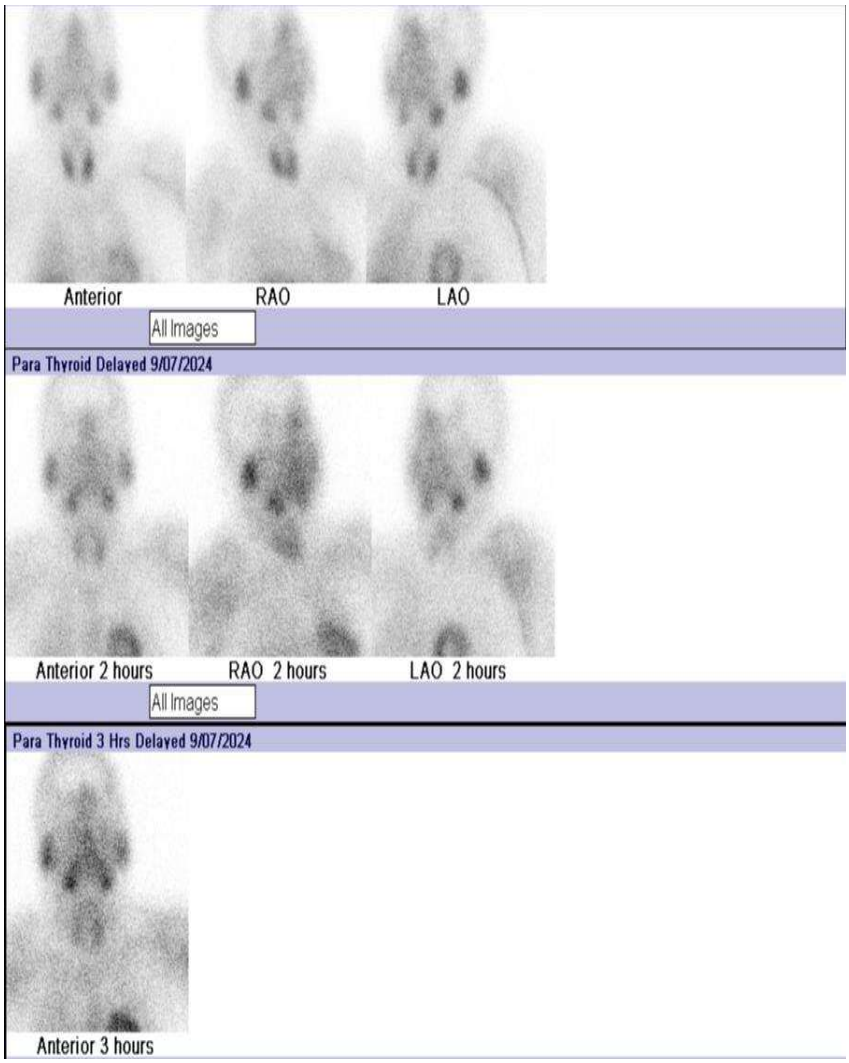

Figure 3. 99mTc-MIBI scan

Discussion

Parathyroid carcinoma is a challenging clinical diagnosis that is nearly always acquired only after a postoperative histological study, as in this case. PC is an uncommon condition that accounts for 0.1–5% of pHPT cases. Higher incidence is observed in Japan, with reports exceeding 5%. In Western nations, it typically accounts for less than 1% of pHPT cases; nevertheless, an Italian investigation found a PC incidence of 5.2% in patients undergoing pHPT.PC incidence has remained generally stable, with the exception of a minor rise in the number of patients with moderately symptomatic PC, most likely due to the discovery of hypercalcemia during biochemistry investigation [3].

The majority of PCs are sporadic; however, they can also occur in hereditary/syndromic settings [3]. The most prevalent mutation is in the cell division cycle 73 (CDC73) or hyperparathyroidism type 2 (HRPT2) genes, which were initially identified in 2002. In addition to being observed in occasional cases, it has shown a

syndromic connection which includes hyperparathyroidism-jaw tumor syndrome, familial isolated hyperparathyroidism, MEN2A and retinoblastoma genes [1].

The PC is frequently diagnosed following surgery based on histology, and the clinical spectrum is similar to that of its benign counterpart. In contrast to parathyroid adenoma, which manifests ten years later and is more common in women, parathyroid malignancies often manifest in the fourth to fifth decade without a sex predilection. Approximately 90% of PCs have hypercalcemia symptoms and are functionally active. These include skeletal symptoms like bone aches, pathological fractures, and brown's tumor; neurological symptoms like exhaustion, sadness, and anxiety; renal manifestations like nephrolithiasis and impaired renal function; and gastrointestinal dysfunction like bloating, constipation, or pancreatitis. In cancer, hypercalcemia and its related symptoms are typically more severe [1].

Patients should be assessed for parathyroid pathology if they exhibit hypercalcemia symptoms and signs, whether or not they have a palpable neck lump. Although there are no strict distinctions between benign parathyroid diseases and malignancies, PCs often have higher laboratory results. A cancer rather than an adenoma should be suspected if serum calcium levels are greater than 14 mg/dl and parathrmone (PTH) levels are more than five times normal. With PC, alkaline phosphatase levels are often greater than 300 IU/L. The imaging supports the malignancy diagnosis and aids in disease localization. It also aids in the detection of metastases and recurrence. [1].

Even while imaging tests (ultrasound, CT scan, MRI, PET) cannot definitively distinguish between adenoma and parathyroid cancer, higher-resolution anatomical studies are very helpful when malignancy is suspected. Contrast-enhanced computed tomography (CT) can show swollen lymph nodes and invasion of adjacent structures in addition to providing great details on the lesion's location and relationship to nearby structures. The best detail on the soft tissues of the neck can be obtained with magnetic resonance imaging (MRI) that uses gadolinium and fat suppression. This type of MRI can also be used to supplement other information, particularly when preoperative evaluation is being conducted [3].

It is presently advised to use Tc-MIBI scintigraphy as the initial radionuclide imaging technique. Cervical ultrasonography and 99mTc-MIBI together have an 81%–95% sensitivity in identifying hyper functioning parathyroid lesions. SPECT/CT is unable to visualize the smallest diseased glands; however, PET/CT can help detect them. 18FDG-PET/CT offers a high sensitivity for PC identification throughout all stages of the disease. Previous studies have employed 11C-methionine (MET) PET/CT as a second-line imaging modality following conventional imaging that yielded negative or equivocal results [2].

It is recommended to manage these uncommon tumors with a multidisciplinary team. Surgery is the primary treatment for parathyroid neoplasms; however, compared to adenoma, the extent of resection in PC is

more radical [7]. Sometimes it is challenging to distinguish between adenoma and cancer before surgery. In many situations, the postoperative diagnosis is a histological surprise. In order to undertake appropriate surgery at the time of first therapy, the surgeon must be alert to detect the sinister characteristics of malignancy intraoperatively. Carcinomas are typically larger, irregular, white masses with a fibrous capsule that can attach to or penetrate the thyroid lobe or adjacent fibro fatty tissue. Metastatic nodes are a pathognomonic sign of cancer. [6]. These patients should have their serum calcium levels regularly checked and supplemented as needed. As the bones are re-mineralized and the surviving parathyroid take up the function, the demand decreases [1].

Conclusion

Parathyroid carcinoma is an uncommon endocrine cancer. Although preoperative diagnosis is difficult, a neck tumor with symptoms and signs of local invasion and hypercalcemia may raise suspicions. The greatest probability of recovery is provided by complete surgical excision during the initial procedure. It is unclear what part adjuvant treatments like chemotherapy and radiation play. Recurrence is frequent, and the preferred course of treatment is surgical salvage of recurring or metastatic disease combined with medication control of hypercalcemia [1]. Prospective studies and trials must be established to establish management guidelines and enhance outcomes, despite the fact that this is difficult given the rarity of the condition.

Statements and Declarations

Conflicts of interest

The authors declare that they do not have conflict of interest.

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