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Case Report .

Parathyroid Carcinoma Masking Behind Primary Hyperparathyroidism: A Case Report with Review of Literature

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ABSTRACT

Parathyroid carcinoma is a rare disease. It accounts for less than 1% of all case of primary hyperparathyroidism and is usually not detected until the time of surgery or thereafter. Preoperative staging is not available for most patients. Hence a radical excision remains the standard management. Usually the disease has an indolent but slowly progressive course. Most of the patients suffer due to complications of hypercalcemia, rather than direct tumor invasion or metastases. The management of PC is difficult in terms of diagnosis, treatment and follow up. Here is a case report that proves a diagnostic challenge to both the clinicians and pathologists and is discussed below.

Keywords: Parathyroid Carcinoma, Hyperparathyroidism, Hypercalcemia.

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CASE REPORT

We report a case of 50 year old male with chief complaints of pain abdomen, vomiting and anorexia since past 3 months. Patient's symptoms started with a history of trauma thereby developing pain in lower limb for which he got operated 3 years back. Thereafter he had recurrent bony pains and developed pathological fracture of shaft of left femur. X-ray showed lytic lesion in mid shaft left femur but no such lesions in X-ray of skull. Bone marrow aspiration ruled out any malignancy or secondaries. Protein electrophoresis indicated hypoalbuminemia and negative for M spike, ruling out Multiple myeloma. For complete work up ultrasound neck was done with Doppler .This showed a solid lesion measuring 4.2 x 2.5 x 2.4 cm just posterior and inferior to the left lobe of thyroid with well-defined margins and vascularity from the inferior thyroid artery. Also serum calcium levels were increased to 15.6 mg/dl (Normal S Calcium level-9 -11 mg/dl).Hence a provisional diagnosis of Parathyroid adenoma was given but patient was non-compliant for surgery. Three months later patient again came with the presenting complaints of pain abdomen. Abdominopelvic ultrasound showed large Pseudocyst of the pancreas at its body and tail & bilateral nephrocalcinosis with calcific specs in both the organs along with hydroureteronephrosis of left kidney.

Serum calcium levels had further increased to value of 17.7 mg/dl. Serum parathormone also was markedly increased to 886 pg/ml (Normal PTH level – 10- 69 pg/ ml). Serum amylase was raised to 188 IU/L (Normal S. amylase level – 40 -140 IU/L). Also patient complained of voice change. Henceforth excision of parathyroid adenoma with cystogastrostomy was done .Per operative haemorrhagic fluid was drained from the pancreatic pseudocyst .The very next day post op serum calcium level came back to normal level.

We received the gross specimen of parathyroid adenoma as a nodular solid grey white mass measuring 3x3x1cm with attached thyroid tissue (Fig 2a and b). Capsule could not be made out. Cut section of the mass showed solid grey white areas with focal yellow specs. Microscopically seen was a neoplasm with lobules and solid nests of clear cells and chief cells separated by thick fibrous septae.(Fig 3). Marked nuclear pleomorphism and hyperchromatism was noted with many bizarre cells and few multinucleated giant cells (Fig 4). Focal area of necrosis with large areas of metastatic calcification was noted (Fig 5). Capsular and extensive vascular invasion was seen with tumour cells invading adjacent thyroid (Fig 6 and 7). With all these features morphological diagnosis of Parathyroid carcinoma was offered.

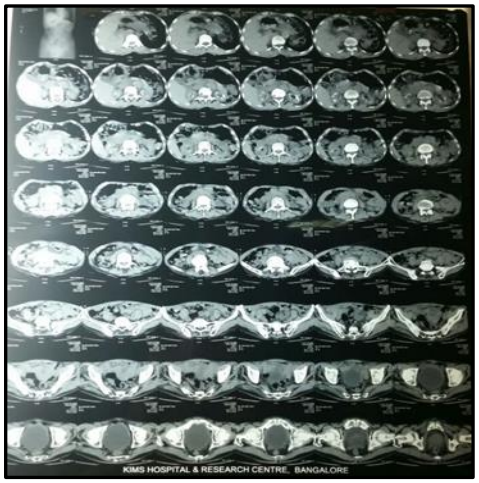
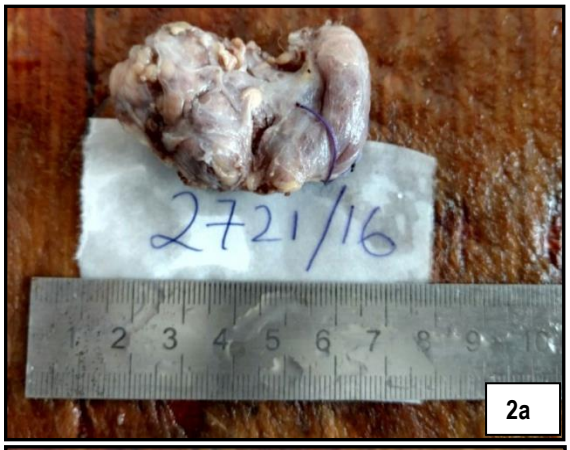


Fig 1: CT scan pseudocyst of pancreas



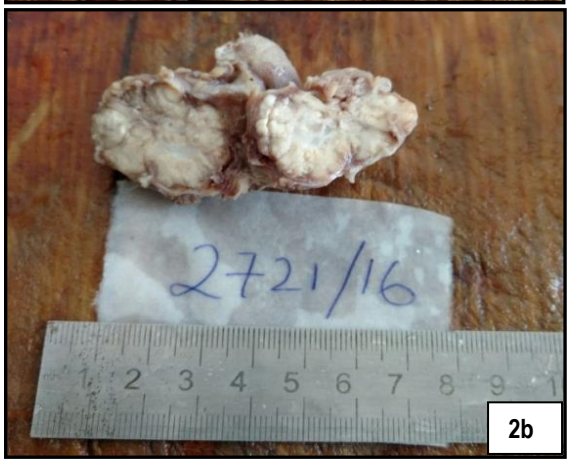


Fig 2a and 2b: Gross picture of Tumour (Specimen)

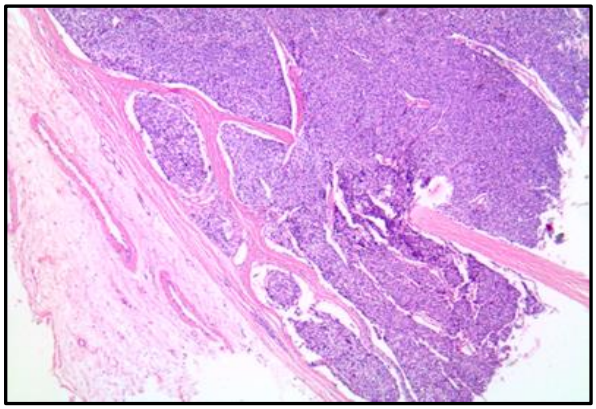


Fig 3: Scanner view of Neoplastic cells

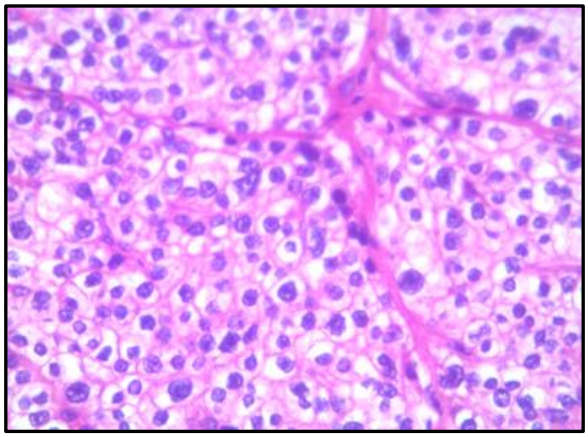


Fig 4: High power (Nests of Neoplastic Cells)

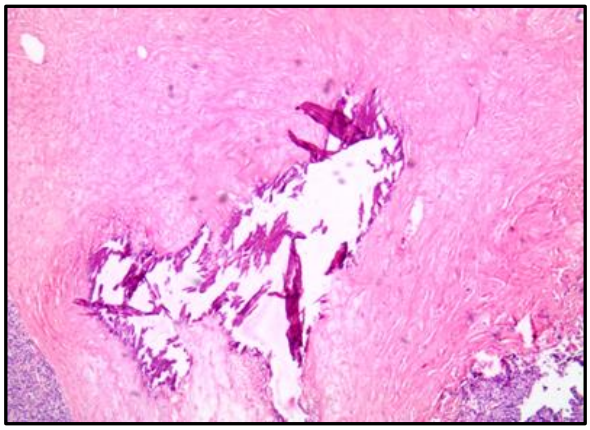


Fig 5: Metastatic Calcification

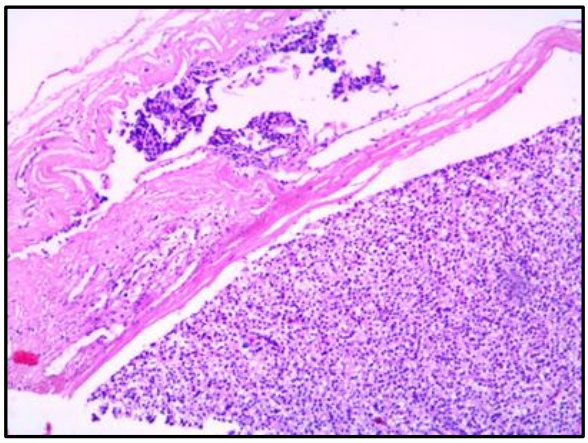


Fig 6: Capsular invasion in Low power

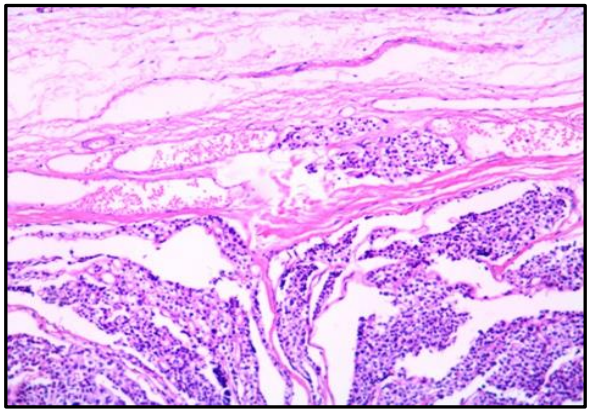


Fig 7: Vascular Invasion in Low power

Table 1: Clinical presentations of Parathyroid neoplasms[9]

Feature	Parathyroid adenoma	Parathyroid Carcinoma
1. Incidence	Much higher (~85% of primary hyperparathyroidism)	Rare (~1% of primary hyperparathyroidism)
2. Age	Mean 56-62 yr	Mean 44-55 yr (one decade younger)
3. Sex	F>M (3:1)	F=M
4. Skeletal involvement	14-20%	Common (63-91%)
5. Renal involvement(renal stone)	10-30%	Commoner (48%-56%)
6. Simultaneous renal and skeletal involvement	Rare (<5%)	Commoner (32%-53%)
7. Palpable neck mass	Rare (<2%)	Commoner (35-45%)
8. Hypercalcemia	Present (2.75-3mmol)	More severe (3.5-4mmol)
9. Weight of tumour	Usually <1 g	Large tumour , average 12 g

Table 2: Histological Criteria for diagnosing of malignancy in a Parathyroid Neoplasm.[10]

Absolute criteria of malignancy	Features associated with malignancy
Presence of any one of the following criteria is sufficient for diagnosis of malignancy	In the absence of absolute criteria, a diagnosis of malignancy should be made with great caution(eg . more than three of the following features are to be present)
1. Invasion of surrounding soft tissue	Capsular invasion (without extension into surrounding tissues)
2. Invasion of surrounding vital structures eg thyroid, esophagus, pharynx, larynx, trachea, recurrent laryngeal nerve, carotid artery	2. Readily identifiable mitotic figures (>5/10 hpf)
3. Vascular invasion	Broad intratumoral fibrous bands splitting the parenchyma and separating the expansile nodules
4. Perineural invasion	Coagulative tumour necrosis(to be distinguished from infarction, which can occur in parathyroid adenoma)
5. Histologically documented regional or distant metastasis	Diffuse sheet like monotonous small cells with high nucleus to cytoplasmic ratio
	6. Diffuse cellular atypia
	7. Macronucleoli present in many tumour cells

DISCUSSION

The most common causes of primary hyperparathyroidism are parathyroid adenoma (approximately 85%), primary parathyroid hyperplasia – eg MEN type 1, 2 etc (approximately 14%), parathyroid carcinoma (approx 1%) (Table 1).[1] Parathyroid carcinomas (PTC) are much rarer than adenomas and affect patients at slightly younger age group equally in both genders compared to adenomas. Majority of such cases are functional with the presentations of hypercalcemia, weakness, renal colic, bone pain and pathological fracture and recurrent pancreatitis. The above features, either present alone or in varied combinations, are particularly suggestive of a diagnosis of PTC over parathyroid adenoma. These include very severe hypercalcemia, simultaneous presence of renal and bone disease, and presence of palpable neck mass. Some cases may be misdiagnosed initially as adenoma, but the malignant nature of the tumor does manifest years later because of recurrence or metastasis.

Most PTC are sporadic, but some cases may occur in the setting of hyperparathyroidism–jaw tumor syndrome, multiple endocrine neoplasia (MEN syndrome) or familial isolated hyperparathyroidism. PTC can also occasionally supervene on parathyroid adenoma or hyperplasia as seen in this case.[2-4]

PTC can be a challenging histological diagnosis but its recognition is of utmost importance. Some PTC are deceptively bland looking whereas some are frankly malignant, as seen in this particular case, with invasive growth, diffuse cellular atypia, and readily identified mitotic figures. The principle histologic features that distinguish parathyroid carcinoma from adenoma are a trabecular pattern**,** mitotic figures, thick fibrous bands (Fig. 4), and capsular and blood vessel invasion (Fig.7).[5] The histological diagnostic criteria is summarized in Table 2. Vascular and capsular invasion with tumour extending beyond the confines of fibrous capsule is quite evident in most cases. The presence of broad fibrous septa throughout the tumor is a characteristic feature of malignancy, although this has to be distinguished from focal scarring due to previous hemorrhage or surgery. This feature is significant only when accompanied by expansile nodules.[6]

The most controversial feature is the presence of mitotic figures. Mitotic figures can often occur in benign parathyroid lesions (71% of parathyroid adenomas and 80% of hyperplasias, the count ranging from 1 single mitosis in several sections to 5 mitoses /10 high-power fields) but its significance has been disputed.[7,8] In this case too mitotic activity was not much significant to be qualified as one of the diagnostic histological criteria.

CONCLUSION

PC is one of the rarest tumours that can remain misdiagnosed for a long time. Patients with persistent unexplained hypercalcemia should be suspected and investigated. However, Prognosis is not predictable, because when malignancy is confirmed, recurrences occur in most case. The average time of recurrence is approximately 3 years, although longer intervals up to 20 years have been reported.[11]

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