Parathyroid Carcinoma

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A 48 year old patient was diagnosed with parathyroid cancer. Blood work revealed severe hypercalcemia (16 mg/dl) and emergency parathyroidectomy was performed. Despite surgical intervention, the patient required monthly pharmacologic intervention to lower serum calcium using biphosphonates. Computed tomography demonstrated both lung and bone metastases. The patient developed acute pancreatitis and suffered multiple pathologic fractures.

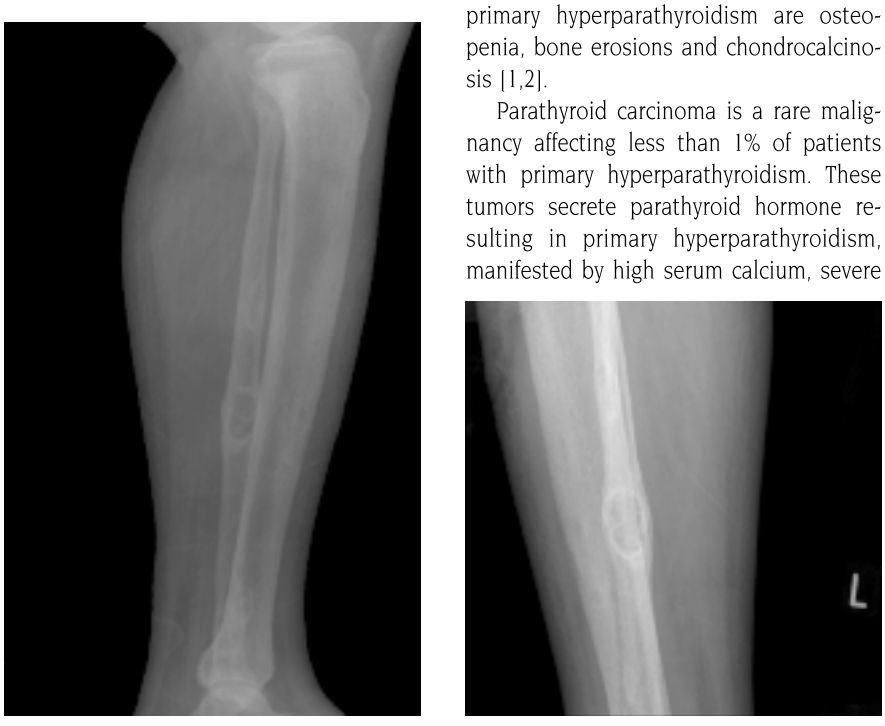
The radiograph of the left leg [Figure A] shows two focal lytic lesions in the diaphysis of the fibula. The largest one is expansile and well demarcated with a thin sclerotic rim. The other lesion, more proximal, has the same characteristics but is eccentric and less expansile. In addition, there are multiple small, lytic, well-defined lesions cortically located in the tibia and distal part of the fibula.

The enlarged figure [Figure B] of the diaphyses of the bones demonstrates, in addition to the cortical lytic lesions, irregularity of the cortical margin, especially in the fibula. This represents bone resorption, shown here as a sub-periostal resorption and intracortical resorption.

The findings in this patient represent the classical picture of primary hyperparathyroidism. One of the effects of the high levels of parathormone is resorption of bone – osteitis fibrosa cystica. Sub-periostal resorption, pathognomonic of hyperparathyroidism, affects particularly the radial aspect of middle phalanges of second and third digits but also the long bones, clavicles, ribs and teeth. The lytic lesions shown here represent Brown tumors.

Brown tumors (osteoclastomas) are lytic, expansile lesions resulting also from intense osteoclastic activity and replacement of bone by fibrous tissue. They occur most often in the metaphyses and diaphyses of long bones, pelvis, jaw and axial skeleton. Those lesions can mimic true tumors or metastases. Other findings in primary hyperparathyroidism are osteopenia, bone erosions and chondrocalcinosis [1,2].

Parathyroid carcinoma is a rare malignancy affecting less than 1% of patients with primary hyperparathyroidism. These tumors secrete parathyroid hormone resulting in primary hyperparathyroidism, manifested by high serum calcium, severe bone disease (90%), renal stones (50–80%), and psychiatric disturbances. One-third of the patients present with a palpable mass in the neck. It essentially occurs in adults above the age of 30.



[A] Radiograph of the left leg. **[B]** Enlarged figure of the leg.

Diagnosis

Diagnosis is determined by a composite of pathologically high calcium levels, which are markedly higher than those of benign primary hyperparathyroidism, and high serum parathyroid hormone levels. Although there are no specific imaging tests, hand films are typical. Definitive diagnosis is based on histology, clinical symptoms and tumor recurrence.

Therapy

Patients often undergo parathyroidectomy in an attempt to control hypercalcemia that is often unresponsive to long-term medical management. The usual causes of death are hypercalcemia, extensive metastases, renal disease, and acute pancreatitis.

Prognosis

The overall 5 year survival rate is 50–70%; however, many die after 5 years. Recurrence occurs in two-thirds of patients after the initial operation, sometimes up to 20 years later. Recurrence within 2 years portends a poor prognosis.

References

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