Severe Idiopathic Hypercalcemia In A Young Female Demonstrated On Skeletal Scintigraphy

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Citation

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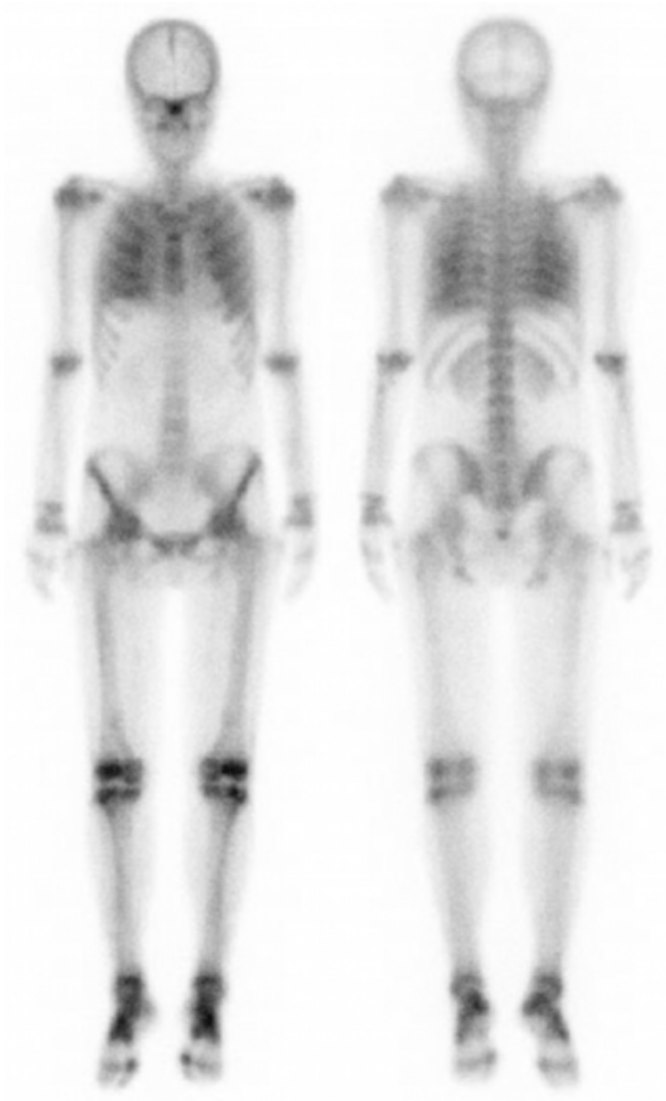
Abstract

A 30 year old female developed nodules on the extensor services of the forearms, knees and back and was noted to have hypercalcemia. Skeletal scintigraphy demonstrated increased uptake throughout the lung fields diffusely and in multiple joints. Multiple investigations were unsuccessful in revealing a cause of the hypercalcemia but confirmed widespread organ calcification. This case is unusual due to the young age of the patient, severity of manifestations on imaging techniques and the lack of an underlying aetiology.

Figure 1

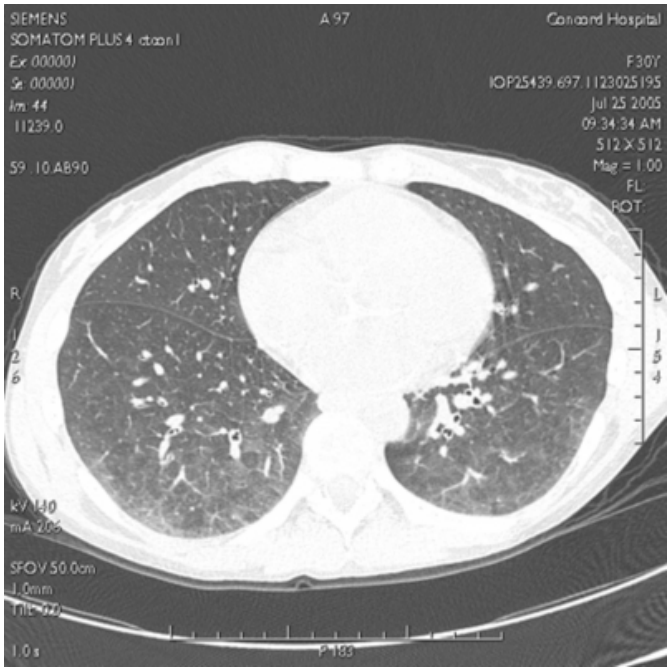
Figure 1: A 30 year old female presented with nodule formation on the extensor services of the forearms, knees and back.



Investigations revealed hypercalcemia 3.32 mmol/L corrected (NR 2.1-2.6 mmol/L) and renal failure Cr 215 µmol (NR 50-90 µmol/L). Her parathyroid hormone, ACE and urinary Calcium levels were normal. She was referred for a bone scan to exclude metastatic bone disease as a cause of her hypercalcemia. Whole body scintigraphy performed 3 hours following administration of 25 mCi (1000 MBq) 99m MDP revealed diffuse intense tracer uptake by the lungs and throughout the joints. Linearly increased uptake was noted in the midline of the skull.

Figure 2

Figure 2: A high resolution Chest CT demonstrated subtle ground glass opacities and centrilobular nodules throughout the lungs.



Lung function tests and bronchoscopy revealed calcification of airways.

Renal biopsy showed nephrocalcinosis with deposition of calcium in tubules and glomeruli with chronic inflammatory changes.

A skeletal survey and bone marrow biopsy were normal.

Figure 3

Figure 3: CT of the Brain demonstrated calcification of the falx cerebri and tentorium cerebelli (a). Calcification was also seen in the sclera bilaterally(b), presumably secondary to the known hypercalcemia. No parenchymal calcification or focal lesion was identified.

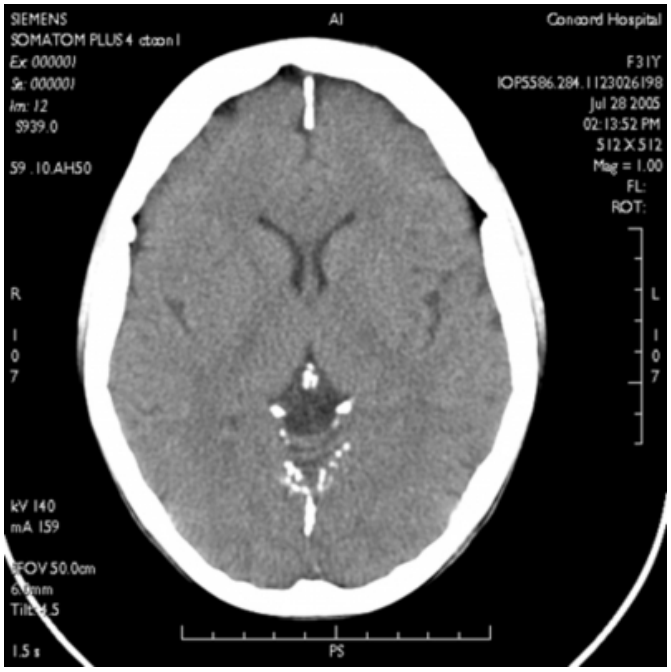
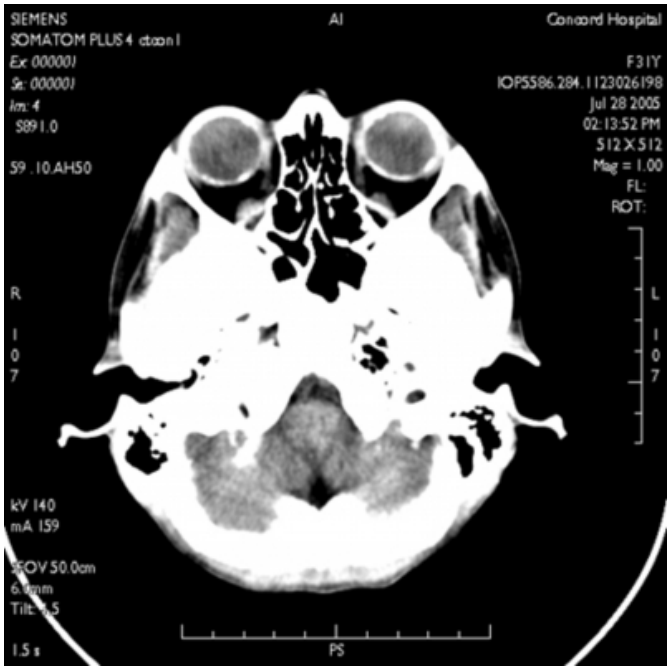


Figure 4



The patient was diagnosed with idiopathic hypercalcemia in the absence of an underlying pathology and commenced on high dose corticosteroid therapy with good initial response.

DISCUSSION

Hypercalcemia can be a manifestation of a serious illness such as malignancy ([1]) or can be detected incidentally by laboratory testing in a patient with no obvious illness.

Whenever hypercalcemia is confirmed, a definitive diagnosis must be established. Although hyperparathyroidism, a frequent cause of asymptomatic hypercalcemia, is a chronic disorder in which manifestations, if any, may be expressed only after months or years, hypercalcemia ([2]) can also be the earliest manifestation of malignancy, the second most common cause of hypercalcemia in the adult. The causes of hypercalcemia are numerous, though hyperparathyroidism and cancer ([3], [4]) account for 90% of cases.

Hypercalcemia in an adult who is asymptomatic is usually due to primary hyperparathyroidism. In malignancy-associated hypercalcemia the disease is usually not occult; rather, symptoms of malignancy bring the patient to the physician, and hypercalcemia is discovered during the evaluation. In such patients the interval between detection of hypercalcemia and death is often <6 months. Accordingly, if an asymptomatic individual has had hypercalcemia or some manifestation of hypercalcemia, such as kidney stones, for >1 or 2 years, it is unlikely that malignancy is the cause. Nevertheless, differentiating primary hyperparathyroidism from occult malignancy can occasionally be difficult, and careful evaluation is required, particularly when the duration of the hypercalcemia is unknown. Hypercalcemia ([6]) not due to hyperparathyroidism or malignancy can result from excessive vitamin D action, high bone turnover from any of several causes, or from renal failure ([8]). Idiopathic hypercalcemia of infancy though rare has been previously described, true idiopathic hypercalcemia of adults is exceedingly rare and Treatment of the hypercalcemia of malignancy is first directed to control of tumor; reduction of tumor mass usually corrects hypercalcemia. If a patient has severe hypercalcemia yet has a good chance for effective tumor therapy, treatment of the hypercalcemia should be vigorous while awaiting the results of definitive therapy ([7]). If hypercalcemia occurs in the late stages of a tumor that is resistant to anti-tumor therapy, the treatment of the hypercalcemia should be judicious as high calcium levels can have a mild sedating effect.

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