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UNUSUAL PRESENTATION OF PRIMARY HYPER PARATHYROIDISM

Radio Diagnosis

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ABSTRACT

Hyperparathyroidism is a condition where usually elevated secretion of parathyroid hormone either primary or secondary. Primary hyperparathyroidism is due to solitary adenoma in 80% cases. Patients with normal intact parathyroid hormone (iPTH) with skeletal features are less well known. Awareness of the unusual phenotype of normohormonic primary hyperparathyroidism may facilitate earlier diagnosis and surgery. We present two cases of primary hyperparathyroidism because of their unusual presentation. Both the patients showed classical radiographic features in skeletal survey and parathyroid adenoma. Biochemical investigations revealed elevated parathyroid hormone and calcium in the first case to suggest primary hyperparathyroidism. Biochemical set up was not suggestive of hyperparathyroidism in the second case and revealed normal level of parathyroid hormone suggestive of normohormonic primary hyperparathyroidism.

KEYWORDS

Hyperparathyroidism, Brown tumour, Parathyroid adenoma, Nephrocalcinosis, Osteoporosis

INTRODUCTION:

CASE REPORT:

CASE 1

Patient was referred for chest radiograph as a pre-operative evaluation for hysterectomy. Incidentally detected an expansile lytic lesion involving lateral third of clavicle and mild irregularity in the lateral end of right clavicle. Advised further work up to rule out hyperparathyroidism. Eight months later patient was referred for radiograph lumbosacral spine to evaluate the cause for severe back pain with clinical diagnosis of lumbar canal stenosis. Radiograph lumbosacral spine including skeletal survey revealed classical features of hyperparathyroidism. HRUSG and MDCT Neck revealed left parathyroid adenoma. USG abdomen shows features of bilateral nephrocalcinosis. Biochemical investigations revealed elevated parathyroid hormone and calcium. Patient improved and discharged after parathyroid adenoidectomy.

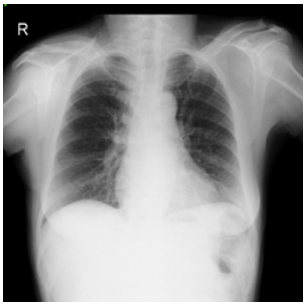

Figure 1: Chest radiograph PA view: (As a pre-op workup for hysterectomy), unremarkable except for expansile lytic lesion involving lateral third of clavicle and mild irregularity in the lateral end of right clavicle.

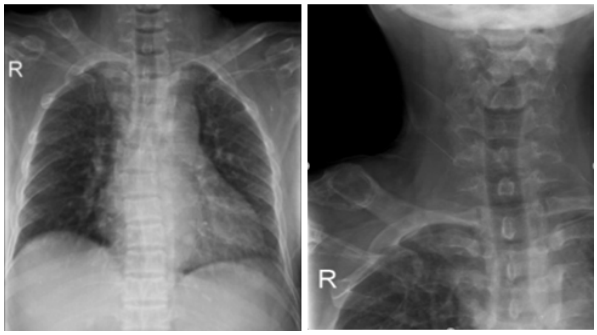

Figure 2 and 3: Radiograph Chest AP and Cervical Spine - A.P. view (8 months later)

There is evidence of expansile lytic lesion in the mid third of right clavicle suggestive of brown tumour. Generalised osteoporotic changes with ill defined coarse trabeculae noted involving cervical and dorsal vertebrae and the ribs.

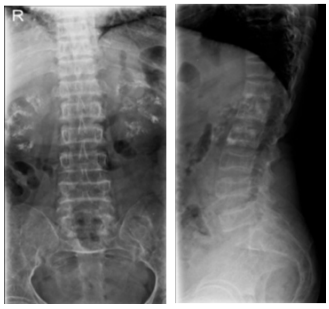

Figure 4 and 5 : Radiograph Lumbosacral Spine - A.P & Lateral views

Extensive generalised osteoporosis with ill defined coarse trabeculae along with vertical compression of lumbar vertebrae. Evidence of nephrocalcinosis is seen bilaterally.

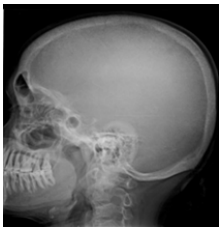

Figure 6: X-Ray Skull - Lateral View: The skull vault shows fine stippled osteoporotic areas giving classical "pepper pot" appearance.

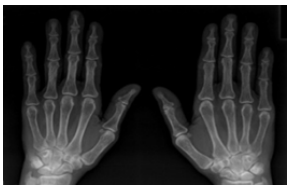

Figure 7: Radiograph Both Hands - A.P view : Phalanges of both hands show classical appearance of subperiosteal resorption.

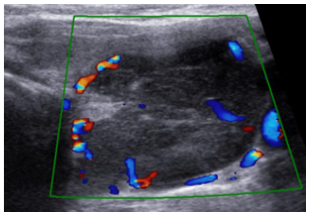

Figure 8: High Resolution Ultrasonography Of Neck : Hypoechoic mass lesion with peripheral and internal vascularity on left side – suggestive of left parathyroid adenoma.

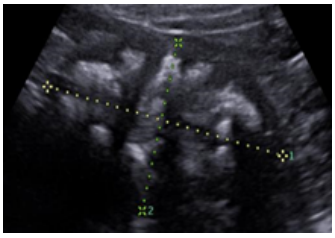

Figure 9: USG abdomen shows features of Medullary Nephrocalcinosis.

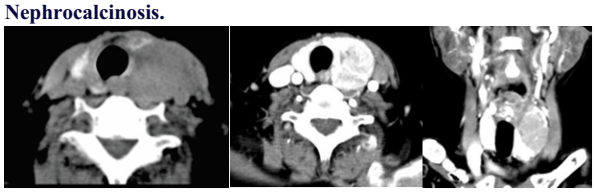

Figure 10,11,12: CECT neck revealed solid enhancing mass lesion in left parathyroid suggestive of parathyroid adenoma of Hyperparathyroidism.

CASE 2:

Patient presented with body weakness and disability of right hip since 4 years, referred to radiology department for the following radiographs and it revealed features suggestive of hyperparathyroidism. HRUSG neck shows hypoechoic mass lesion with increased perilesional and intralesional vascularity on colour doppler study suggestive of left parathyroid adenoma. Biochemical set up was not suggestive during this stage and revealed normal level of parathyroid hormone suggestive of normohormonic primary hyperparathyroidism.

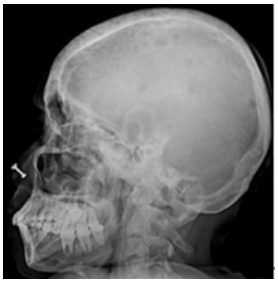

Figure 13: X-ray skull - lateral view: The skull vault shows fine stippled osteoporotic areas giving classical "pepper pot" appearance

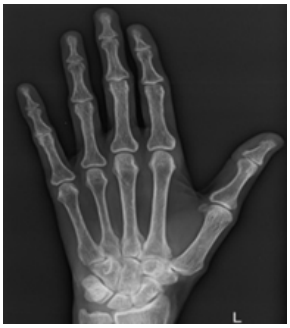

Figure 14: Radiograph of hand – AP view : Mild generalised osteoporosis with minimal subperiosteal resorption – Suggestive of hyperparathyroidism.

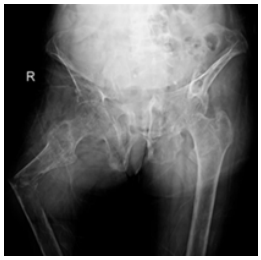

Figure 15: Radiograph pelvis with proximal part of thigh - A.P view
Generalized osteoporosis and right femur shows gross thinning of cortex and endosteal scalloping with pathological fracture in proximal 3rd

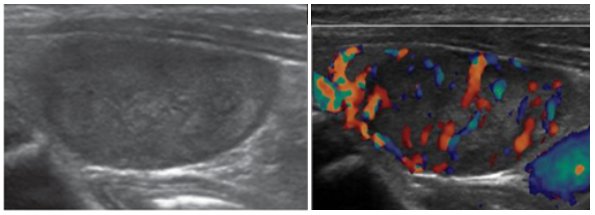

Figure 16 and 17: HRUSG neck : Evidence of left parathyroid adenoma noted with increased perilesional and intralesional vascularity on colour doppler study.

DISCUSSION:

- Primary hyperparathyroidism is diagnosed in patients with hypercalcemia and high intact serum parathyroid hormone level. The incidence of PHPT is nearly two or three times more common in women than in men. It is an endocrine condition categorized as primary, secondary, or tertiary. Primary HPT (PHPT) is characterized by hypersecretion of PTH, which may be caused by an adenoma (solitary or multiple), idiopathic hyperplasia, or a parathyroid carcinoma. Secondary HPT is caused by hypocalcaemia or vitamin D deficiency acting as a stimulus of excessive PTH production. Chronic renal failure is the main cause of secondary HPT. Tertiary HPT is caused by the development of autonomous parathyroid hyperplasia after long-standing secondary HPT and it is most often in patients with renal failure.
- Most of the patients are asymptomatic and they present with various manifestations including recurrent nephrolithiasis, bone pain, peptic ulcers, depression, polyuria, polydipsia etc. However, with great awareness of the disease, wider use of multiphasic screening tests the diagnosis is frequently made in patients who have no symptoms /minimal symptoms. Classical roentgenographic features are subperiosteal cortical bone erosions, osteopenia, salt and pepper' mottling of skull, local destructive bone lesions ("brown tumour") and calcification of the soft tissue.
- Increased parathyroid activity liberates parathormone, which exerts a strong osteoclastic effect on the skeleton. The most common radiologic finding in primary hyperparathyroidism is osteopenia, which may be generalised or asymmetric. Bone resorption may occur at many different anatomic sites and can be classified as subperiosteal, intracortical, trabecular, endosteal, subchondral, subligamentous or subtendinous. Subperiosteal resorption is an early and pathognomonic sign of hyperparathyroidism. The earliest bone changes are visible in the hands, particularly in the phalanges, symphysis pubis, distal clavicle, vertebral bodies, lamina dura and calvaria. The most common site in hyperparathyroidism is the radial aspect of middle phalanges of the index and middle fingers.
- Radiological features in the spine include osteopenia, trabecular accentuation, endplate concavities, rugger jersey spine, widened sacroiliac joints. Diffuse granular deossification produces a mottled appearance to the calvaria gives "salt and pepper" or "pepper pot skull". A characteristic sign is resorption of the cortical bone (lamina dura) surrounding the tooth socket. Soft tissue changes include nephrocalcinosis, renal calculi, chondrocalcinosis, and calcification in various periarticular tissues and visceral organs.
- Primary hyperparathyroidism results in bone resorption and tissue change known as osteitis fibrosa cystica, which is the end stage and is associated with the development of brown tumours (Osteoclastoma). Brown tumours are not true neoplasms, but they can be locally aggressive and mimic malignancies. The term "brown tumour" is derived from the characteristic appearance of a brown-coloured material within the cystic lesion, which is caused by high vascularity, haemorrhage, and hemosiderin deposits. The most common sites of occurrence include mandible, pelvis, ribs, and femora, although any bone can be affected. On imaging, brown tumours seen as osteolytic or sclerotic lesions with regular borders, and no cortical disruption, periosteal reaction, or inflammatory signs. It shows fluid-fluid level and enhancement on post contrast MRI. Brown tumours are hypervascular and appears intensely active in bone scintigraphy.
- Manifestations of hypercalcemia will appear as nephrocalcinosis and renal calculi. This is seen in up to 75% of hyperparathyroidism patients and sonography helps in detection of medullary nephrocalcinosis and renal calculi.

- Primary hyperparathyroidism caused by adenoma or hyperplasia can be cured surgically with a high rate of success. Sonography and Tc-99m sestamibi scintigraphy are useful in the pre-operative evaluation of primary hyperparathyroidism. USG is the primary modality used to localise parathyroid tumours. The size of the adenoma is usually correlated with the degree of parathyroid elevation. Adenoma appears as well defined hypoechoic lesions with cystic or necrotic areas. Colour and power doppler imaging commonly show a characteristic polar feeding vessel, which gives "arc or rim of vascularity". Nuclear imaging (Sestamibi scan) have highest accuracy rate, widely available and useful in detection of diseased glands outside of the neck. CT imaging shows intense enhancement of adenoma and scanning from skull base through the mediastinum helps in detecting most ectopic glands. MRI can be used to evaluate ectopic parathyroid adenomas. On T1W images, adenomas appears as low signal intensity masses, whereas intermediate or high signal intensity is seen on T2W images. Gadolinium enhancement with fat suppression results in diffuse enhancement in adenomas.
- Treatment of HPT depends on the etiology of the condition. Once PHPT is diagnosed, the only cure is surgical removal of the parathyroid lesion and the best treatment of brown tumours is cure of the underlying PHPT.
- Normohormonal primary hyperparathyroidism (NHPHP) is a distinct entity of parathyroid disease that manifests normal PTH. Possible mechanism for NHPHP include pulsatile PTH secretion, post translational modification of the PTH molecule altering its measurement but not function, presence of unmeasured but active PTH fragments, presence of a circulating entity interfering with the assay, the presence of PTH and increased peripheral sensitivity to a normal PTH molecule.

CONCLUSION:

- Hyperparathyroidism is a condition where usually elevated secretion of parathyroid hormone either primary or secondary. About 90% of the patients with primary hyperparathyroidism have elevated parathyroid hormone (PTH) levels. Patients with normal intact parathyroid hormone (iPTH) with skeletal features are less well known. Awareness of the unusual phenotype of normohormonic primary hyperparathyroidism may facilitate earlier diagnosis and surgery. We must be alert in evaluating the radiographs and keeping in mind the possibility of high suspicion of hyperparathyroidism.
- Radiographic findings of subperiosteal resorption are most specific for primary hyperparathyroidism. Ultrasonography is approximately 75% sensitive in identifying adenoma, but this technique has low sensitivity in identifying ectopic lesions. The advantage of sestamibi scan are its wide availability and the ability to evaluate for diseased glands outside of the neck at the same time. MRI and CT also can be used as diagnostic modalities to evaluate ectopic parathyroid adenomas.
- A definitive diagnosis of primary hyperparathyroidism is only possible after complete evaluation of medical history, radiographic findings and biochemical analysis. It will help to achieve a correct diagnosis and avoid unnecessary bone resections in patients with primary hyperparathyroidism.

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