Abstract:
A 41 year old male presented with complaints of progressively increasing pain over the left leg for duration of one year and progressive swelling over the left leg for nine months. Patient gave history of polyuria, vomiting, polydipsia, lethargy and fatigue for a period of 6 months on eliciting further history. Patient had a pathological fracture of right humerus two years back and left hydronephrosis two years back, diagnosed as left pelviureteric stone, treated with ESWL. Patient was not a smoker, not alcoholic and vegetarian. There was no other significant history. Physical examination revealed two hard non mobile masses of size 6x6 cm and 2x2 cm over the left tibia(Fig-1) and 2x2 cm hard swelling over the first interphalangeal joint of left fourth finger(Fig-2). Neck examination didn’t reveal any palpable mass(Fig-3).

Keyword: Parathyroid adenoma, Von Recklinghausens disease, osteitis fibrosa cystica
Urine examination showed elevated 24 hour urine calcium. Blood investigations revealed elevated serum calcium level (11.9mg/dl), serum parathormone (1774.3pg/ml) and serum alkaline phosphatase (2240 U/L) suggestive of hyperparathyroidism. Blood urea and creatinine were raised suggestive of chronic kidney disease. High frequency ultrasonography implied 2.3x1.7x1.5cm hypoechoic, well defined mass lesion near the superior pole of left lobe of thyroid suggestive of parathyroid adenoma and no evidence of suspicious neck nodes. 99mTc- Sestamibi scan revealed a picture suggestive of left superior parathyroid adenoma(Fig-4).

Computed tomography of chest revealed no mediastinal lymphadenopathy. Sonography of abdomen revealed bilateral multiple renal calculi. Roentgenogram of skull showed multiple osteolytic lesions sub-osteal bone resorption with pepper pot appearance. Roentgenogram of leg, hand revealed osteolytic lesions over the swellings(Fig-5,6).

Fig.1

Fig.2

Fig.3

Fig.4
Histopathology of biopsy taken from tibial lesions suggested features of non ossifying fibroma of tibia. The patient was diagnosed to be having left superior parathyroid adenoma with severe complications osteitis fibrosa cystica and renal osteodystrophy. Left superior parathyroidectomy was done (Fig-7).

On third day postoperatively, patient developed features of hungry bone syndrome diagnosed with features of hypocalcaemia and was treated with intravenous calcium preparations and oral calcitriol supplementation. The symptoms improved gradually and the skeletal lesions regressed. Post operative histopathology confirmed as parathyroid adenoma.

Discussion:
Spectrum of hyperparathyroidism varies from fatigue, myalgia, renal stones, polyuria, polydipsia, skeletal manifestation, and psychiatric disturbances. The causes of hyper parathyroidism include parathyroid adenoma, parathyroid hyperplasia, parathyroid carcinoma, hereditary syndromes and renal osteodystrophy. Osteitis fibrosa cystica was first described by Gerhard Engel and then explained by famous Friedrich Daniel Von Recklinghausen in 1890. Osteitis Fibrosa Cystica is a severe complication of hyperparathyroidism in which bone turns soft and become deformed. It is caused by surplus PTH from overactive parathyroid glands. The pathophysiology includes the hyper functioning of osteoclasts resulting in bone resorption causing osteolysis which is replaced by peritrabecular fibrosis and formation of brown tumors. The diagnosis includes the radiologic investigations, diagnosis of hyper parathyroidism with blood investigations and nuclear imaging-Sestsmibi-Tc99. The skeletal manifestations improved after surgery. The post operative complication includes the hungry bone syndrome due to functional or relative hypoparathyroidism after surgery resulting in transient hypocalcaemia which recovers in a week. Calcium supplementation in the form of calcitriol leads to regression of skeletal manifestations. With the advent of imaging / nuclear imaging modalities, hyperparathyroidism is diagnosed early before the appearance of OFC in western countries whereas hyperparathyroidism is often seen with OFC in India.
References:


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