International Journal Dental and Medical Sciences Research
Volume 3, Issue 6, Nov-Dec 2021 pp 291-292www.ijdmsrjournal.com ISSN: 2582-6018

CLINICAL IMAGE

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Submitted: 01-12-2021 Revised: 11-12-2021 Accepted: 14-12-2021

Case presentation: 50 years old female presented with painless swelling in right lower jaw for 5 years which was insidiously increasing in size. Patient gave history of difficulty in chewing with facial disfigurement. No similar swellings in other parts of body were present. No fractures and renal stones history were present with no family history of similar illness. On examination single hard, irregular, non-tender swelling of size 12 x 8 cm involving rt side of jaw along the mandibular angle was present (image 1). Her biochemical investigation showed S. Calcium 11.2 mg/dl (8.5-11mg/dl), S. phosphorus 2.1 mg/dl (2.5-7mg/dl), S. alkaline phosphatase 579U/L (30-120U), iPTH 781 pg/ml(12-72pg/ml). S creatinine 0.9 mg/dl (0.5-1.2mg/dl). Xay done was suggestive of expansile lesion in mandible right side-image 2. **What is the diagnosis?**

Answer: Ossifying fibroma of jaw in hyperparathyroidism jaw tumor (HPT-JT) syndrome. HPT JT syndrome is autosomal dominant disorder due to mutation in HPRT2 tumour suppressor gene (also known as CDC 73) located on chromosome 1q25-q31 that regulates parafibromin expression. This syndrome is characterised by ossifying fibrosis of jaw, parathyroid adenoma or carcinoma along with renal and uterine tumours. Ossifying fibroma usually is the first clinical manifestations and its commonly involves mandible and maxilla. These are benign tumours with <0.5% chances of malignant transformation and may be single or multiple. X-ray findings are of radiolucent lesion usually unilocular may be multilocular with peripheral sclerotic rim with teeth splaying however root resorption is not seen. These tumours arise from mesenchymal cells of periodontal ligament and mainly comprises of fibrous tissue. Main differential diagnoses are osseous fibroma which has ground glass appearance on X-ray, brown tumour which is purely lytic lesion with no sclerotic rim, ameloblastoma which are associated with root resorption and osteosarcoma which shows aggressive periosteal reaction. Present patient`s ultrasonography neck was suggestive of single left inferior parathyroid gland adenoma. Due to financial constraints nuclear imaging was not done and patient was planned for subtotal parathyroidectomy followed by resection of mandibular lesion and reconstruction. In addition, screening of family members was advised.

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International Journal Dental and Medical Sciences Research
Volume 3, Issue 6, Nov-Dec 2021 pp 291-292www.ijdmsrjournal.com ISSN: 2582-6018

Image 1

Image 2