Renal osteodystrophy presenting as Brown tumour at the paranasal zone: A rare case report

Vikash Khandelia[1*], Umashanker Nama[2], Ashok Mahawar[3], Sharwan Kumar Sharma[4]

[1]Professor and HOD, [2-4]Junior Resident, Dept. of Nephrology, Govt. Medical College, Kota, Rajasthan, India

***Corresponding Author: Vikash Khandelia**
Email: vikasnephro@gmail.com

Abstract
Hyperparathyroidism causes disturbance in the mineral and bone assimilation process which might produce skeletal as well as extra skeletal changes. Renal insufficiency disorders might disturb the homeostasis of calcium, vitamin D and bone metabolism causing secondary hyperparathyroidism. A complication of hyperparathyroidism is Browns tumour, which is a result of disorganised osteoclast activities. It is a type of osteitis fibrosa cystica caused due to the excess osteoclast activity during hyperparathyroidism. It can be located in any part of the body but rarely is it seen in the craniofacial area. Here also it commonly affects the mandible than the maxilla. Here is such a rare case of Browns tumour in the paranasal region in a 15-year-old boy with stunted growth. His renal parameters were deranged raising a suspicion for renal insufficiency causing secondary hyperparathyroidism, which could have been the cause for his stunted growth too. The aim to present this case is to create awareness amongst the clinicians while treating cases of stunted growth or paranasal cystic swellings, to rule out any renal disease or hyperparathyroidism. As in such instances early management might minimise the chances of surgery and reducing the mortality rate.

Keywords: Browns tumour, High turnover bone disease.

Introduction
Renal osteodystrophy (ROD) is a known complication of chronic kidney disease (CKD), which is caused due to derangement in the mineral and bone metabolism producing skeletal changes. Secondary hyperparathyroidism is a consequence of the failing kidney functions, which in rare instances produces browns tumour. Brown tumour consists of fibrous tissue, woven bone with adjacent blood vessels but without any matrix. The deposition of hemosiderin in the osteocystsgives it the characteristic brown colour.[1] These are frequently located in the ribs, clavicles, extremities but rarely are it seen in the craniofacial bones.[2]

Evidence of ROD is seen only in histological findings of moderate renal failure cases but as the renal failure deteriorates, the severity and presentation of ROD worsens too. In almost 90-100% of cases with ESRD maintained with haemodialysis, ROD is evident when the GFR begins to fall from 60 ml/min.[3,4]

Here, we present a case is of high turnover ROD which represented as a cystic swelling around the paranasal sinus in a 15-year-old boy with short stature.

Case Study
A 15-year-old boy presented with history of bowing of legs and short stature for 2 years (Figure 1). An orthopaedic was consulted and calcium, vitamin D supplements and non-steroidal anti-inflammatory drugs were prescribed for the same. Patient was on antihypertensive for arterial hypertension. Since 3 months, the patient developed swelling around the nose and within the mouth on the hard palate (Figure 2), for which he was consulted to an ENT physician who advised CT scan of the area. The findings of the CT scan of the paranasal sinuses were suggestive of cystic, lytic lesions of left upper alveolus (Figure 3).

On examination a prominent bulge was seen on the hard palate with mild swelling around the left nostril. The patient's renal parameters were also deranged with increased blood urea (99mg/dL) and creatinine (4.0mg/dL), for which a nephrologist was consulted. His abdominal ultrasound revealed bilateral small kidneys with cystitis and insignificant post-void residual urine (PVRU).

On further laboratory workup, there were elevated parathyroid levels (PTH: 616.6 pg/ml), high turnover bone disease but with preserved calcium (9.4mg/dL) and phosphorus levels (4.2 mg/dL). Patient even showed signs of metabolic acidosis with pH of 7.23, bicarbonate 10.6 mEq/L with high base excess of 13.4 mmol/L. With all the above findings he was diagnosed as chronic kidney disease with renal osteodystrophy (mineral and bone disorder). Patient was treated with antihypertensive and diuretics along with calcium and iron supplements. He was advised restricted dietary phosphorus.

On his follow up visit after 5 months, there was increase in the swelling (Figure 4) so fine needle aspiration cytology (FNAC) was advised. The findings were of inflammatory cells, RBC and cystic fluid indicating infected cystic lesion. Even his blood pressure was high (160/120 mmHg) with increased bone cell activity (serum alkaline phosphatase: 1114.0 U/L). Hence a calcium channel blocker was added (Amlodipine 5 mg/day) along with phosphate lowering agent (calcium acetate 675 mg)

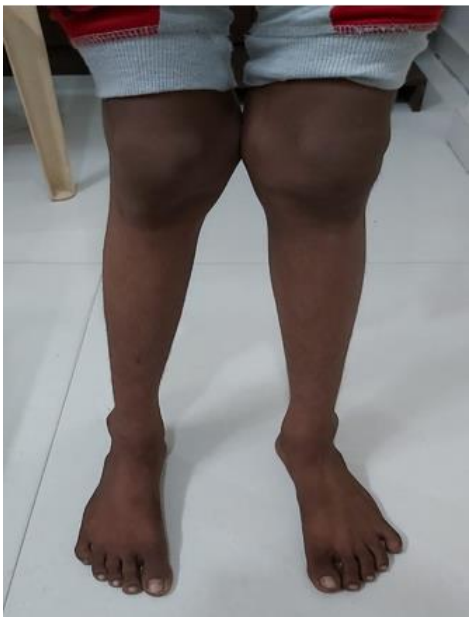

Fig. 1: Bowing of the knees

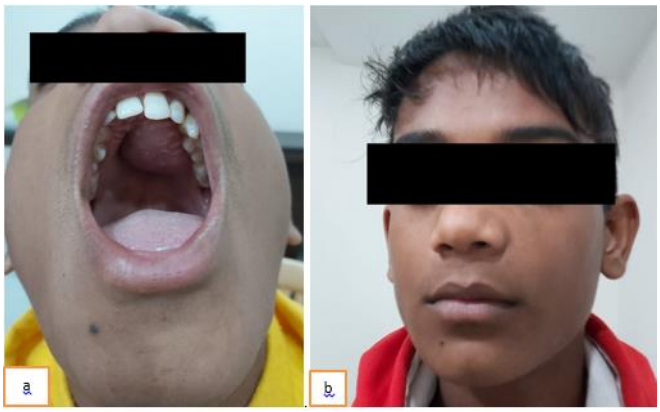

Fig. 2: (a) Mass on the hard palate; (b) Facial asymmetry due to the mass around left nostril

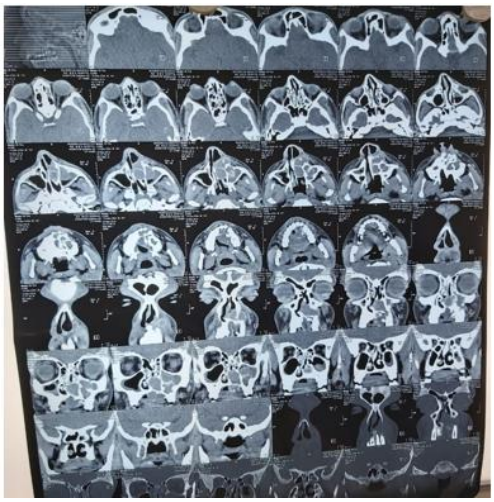

Fig 3: CT Scan of the paranasal sinus revealing cystic, lytic lesion the left upper alveolus

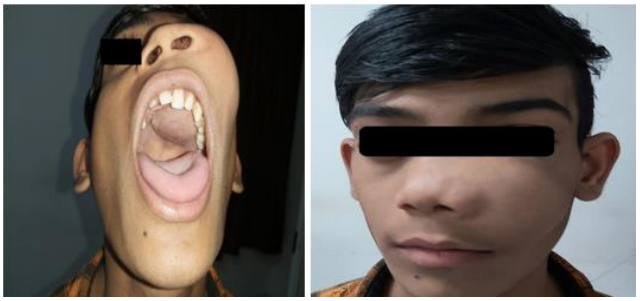

Fig. 4: Follow up image showing increase in the size of the tumour

Discussion

Apart from excretion of metabolic wastes, kidneys play a vital role in metabolism of calcium and certain hormones in the body by synthesising vitamin D. The vitamin D in association with other minerals and hormones helps in absorption of dietary calcium from the intestines which are further used in bone metabolism and maintenance of the serum calcium levels.[5] In end stage renal disease (ESRD) or chronic kidney disease (CKD) due to severe damage to the structure and functions of the kidneys, there is a decrease in the production of Vitamin D which in turn decreases the calcium absorption. This hypocalcaemia condition triggers the release of parathyroid hormone leading to secondary hyperparathyroidism. If left untreated, this hyperparathyroidism can cause skeletal lesions known as renal osteodystrophy.[6]

Renal osteodystrophy includes several bone disorders such as osteomalacia, adynamic bone disease and osteitis fibrosa cystic of which only the latter is high turnover bone diseases while the others are low turnover variants. Due to the increased PTH levels, there is high bone turnover which results in osteoclast activity and bone breakdown causing easy fractures.[7] These changes gradually cause bone loss in specific areas which are substituted by fibrous matter making it appear as brown tumour.

Browns tumour compromises of fibrous tissue along with the woven bone and the supporting blood vessels but without any matrix. The characteristic deposition of hemosiderin in the osteolytic cysts gives it the brown colour.[1] It is commonly seen in long bones, ribs, pelvis, and spinal column and rarely involves the craniofacial bones.[8] The above case is one such illustration of development of brown tumour in the paranasal sinus in a male with renal insufficiency.

Clinical manifestation depends on the size and location of the tumour, but otherwise these are painless bony masses. It might pose an increased risk of fracture, facial disfigurement; difficulty in mastication and phonation, if in the spinal column may cause compression of the cord.[9-11] These facial deformities may also cause social discomfort for the patient. Even symptoms of headache, visual disturbances, displacement of teeth or nasal bleeding are reported.[9,12] In the above case the swelling was asymptomatic, but the ROD was the probable cause for his stunted growth and bowing of legs.

Few studies suggest that elevated serum phosphate levels (>6.5 mg/dL) have increased mortality of 27% when compared to cases with normal levels.[13] There are no specific markers or test to confirm ROD, other than bone biopsy, but that being an invasive procedure is sparsely considered.[14] The values of serum calcium, phosphorus, PTH and alkaline phosphatase help in determining the diagnosis and planning the treatment for ROD.[13]

Management of ROD chiefly targets at maintaining the serum calcium and phosphorus levels, treating hyperparathyroidism and preventing hyperplasia of the parathyroid gland.[15] Therapy should be started at the earliest to prevent major skeletal abnormalities. The PTH level is maintained with drugs, dialysis or parathyroidectomy.[2] In cases of high turnover bone disease, maintaining the serum phosphate levels is the prime objective as its retention participates in development of hyperparathyroidism. Restriction of dietary phosphate (800-1000 mg/day) is advised if the serum phosphate levels are elevated than the range advised depending on the stage of CKD. Phosphorus binders are administered to minimise the intestinal absorption of phosphorus and cautious administration of vitamin D metabolites is suggestive. In cases of severe forms of hyperparathyroidism including PTH levels >800 pg/ml, increased calcium, phosphorus levels, severe bone pains and osteoporosis pose the need for surgical assessment.[16] Surgical resection of the tumour is advised only if the bony mass is hampering functions such as chewing, talking, breathing or causing severe facial deformity or there is no regression despite treatment for 1-2 years.[17] A similar case of no regression despite treatment has been reported by Can O, et al., in a 30yr old female, with a protruding tumour of mandible progressing since 10 years.[8] Initially total parathyroidectomy was performed but the lesion did not regress, later renal transplantation was performed and due to its location with great difficulty surgery was performed.

In the above case since the calcium and phosphorus levels were preserved (9.4 mg/dL and 4.2 mg/dL, respectively) and the PTH elevation was 616.6, pharmacological treatment was provided. This case presented with paranasal swelling, but the entire workup including his stunted growth, bowing of legs and serum PTH levels along with deranged renal parameters made the diagnosis of renal osteodystrophy with Browns tumour. On his follow up visits the tumour showed no regression, hence he is kept on close observation to decide for surgical intervention.

Conclusion

Mineral and bone metabolism abnormalities in patients with CKD causes complex changes which hampers the quality of life. In cases of CKD physicians should be cautious in monitoring the PTH levels to minimise the prevalence of ROD. In patients with CKD with development of any bony mass, secondary hyperparathyroidism with browns tumour should be considered. The main aim of this case presentation is for the clinicians to be keen while treating patients for stunted growth or any cystic lesion along the paranasal region ion children to assess the renal profile and parathyroid functioning.

Conflict of Interest

The authors declare that there is no conflict of interests

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None.

References

1. Araújo SM, Bruin VM, Nunes AS, Pereira EN, Mota AC, Ribeiro MZ, et al.Multiple brown tumors causing spinal cord compression in association with secondary hyperparathyroidism. *Int Urol Nephrol*. 2013;45(3):913-6.
2. Tarrass F, Benjelloun M, Bensaha T. Severe jaw enlargement associated with uremic hyperparathyroidism. *Hemodial Int*. 2008;12:316-8.
3. Buargub MA, Nabulsi MF, Shafeh TA. Prevalence and pattern of renal osteodystrophy in chronic hemodialysis patients: a cross sectional study of 103 patients. *Saudi J Kidney Dis Transpl*. 2006;17:401-7.
4. Hernandez JD, Wesseling K, Salusky IB. Role of parathyroid hormone and therapy with active vitamin D sterols in renal osteodystrophy. *Semin Dial*. 2005; 18: 290-5.
5. Eaton DC, Pooler JP. Fisiologia Renal de Vander. 6th ed. Porto Alegre: Artmed; 2006.
6. Karsburg RM, Campos KR, Peres MPSM, Bologna SB, Lourenco SV, Franco JB. Facial lesions caused by renal osteodystrophy in a patient with chronic renal insufficiency: a case report. *Rev Odonto Cienc*. 2012;27(2):161-5.
7. Pitt SC, Sippel RS, Chen H. Secondary and tertiary hyperparathyroidism, state of the art surgical management. *Surg Clin North Am*. 2009;89(5):1227-39.
8. Can Ö, Boynueğri B, Gökçe AM. Brown Tumors: A Case Report and Review of the Literature. *Case Rep Nephrol Dial*. 2016;6:46-52.
9. Triantafillidou K, Zouloumis L, Karakinaris G, Kalimeras E, Iordanidis F. Brown tumors of the jaws associated with primary or secondary hyperparathyroidism. A clinical study and review of the literature. *Am J Otolaryngol*. 2006;27(4):281-6.
10. Fernández Sanromán J, Antón Badiola IM, Costas López A. Tumor pardoen la sínfisis mandibular comoprimeramanifestaciónclínica de hiperparatiroidismo: diagnóstico y tratamiento. *Medicina Oral Patología Oral y CirugíaBucal*, 2005;10:169-72.
11. Marcos García M, Pino Rivero V, KeituqwaYáñez T, Alcaraz Fuentes M, Trinidad Ruiz G, Blasco Huelva A et al. Brown bone tumor as the first manifestation of primary hyperparathyroidism. *Acta Otorrinolaringol Esp*. 2003;54:470-73.
12. Karabekmez FE, Duymaz A, Keskin M, Tosun Z. Huge deforming brown tumour of the maxilla and mandible in a patient with secondary hyperparathyroidism. *J Plast Reconstr Aesthet Surg*. 2008;61:1404-5.
13. Rodriguez-Benot A, martin Malo A, Alvarez-Lara MA, Rodriguez M, Aljama P. Mild hyperphosphatemia and mortality in hemodialysis patients. *Am J Kidney Dis*. 2005;46:68-77.
14. Moe S, Drueke T, Cunningham J, Goodman W, Martin K. Definition, evaluation and classification of renal osteodystrophy: a position statement from Kidney Disease: Improving Global Outcomes (KDIGO). *Kidney Int*. 2006;69:1945-53.

15. Lopez-Hilker S, Dusso AS, Rapp NS, Martin KJ, Slatopolsky E. Phosphorus restriction reverses hyperparathyroidism in uremia independent of changes in calcium and calcitriol. *Am J Physiol*. 1990;259:F432-7.
16. Lorenzoni V, Trieste L, Turchetti G. The cost-effectiveness of drug therapies to treat secondary hyperparathyroidism in renal failure: a focus on evidence regarding paricalcitol and cinacalcet. *Expert Rev Pharmacoecon Outcomes Res*. 2015;15(4):611-24.
17. Leal CT, Lacativa PG, Gomes EM, Nunes RC, Costa FL. Surgical approach and clinical outcome of a deforming brown tumor at the maxilla in a patient with secondary hyperparathyroidism due to chronic renal failure. *Arq Bras Endocrinol Metabol.* 2006;50:963-7.

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