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CALCIFIED MAXILLARY FIBROMA IN A PATIENT ON HEMODIALYSIS FOR SEVEN YEARS

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ABSTRACT

Background: The presentation of mineral bone disorder (MBD) in individuals with chronic kidney disease (CKD) may vary. Consequently, physicians should be capable of recognising this condition when there is a suspicion of its existence. This case report will describe a calcified maxilla tumour as a manifestation of CKD-MBD.

Case description: Initially asymptomatic, a 32-year-old female presented with progressive swelling of the upper left jaw. She had a history of routine haemodialysis. Further laboratory, radiological, and histopathological workup revealed the mass was indeed calcified maxillary fibroma arising from the manifestation of CKD-MBD.

Conclusion: This study underscores the significance of clinical comprehension of the broad-spectrum manifestations of CKD-MBD, including an initially asymptomatic mass. In addition, the screening of the patient's biochemical was required to determine the necessity of early intervention and improve the patient's outcome.

KEYWORDS

CKD-MBD, complication, hemodialysis, histopathology, parathyroid

LEARNING POINTS

- The case emphasises the importance of recognising atypical presentations of chronic kidney disease-mineral bone disorder (CKD-MBD), such as a calcified mass, which are rarely reported but critical for timely intervention.
- This report underscores the necessity for routine screening for secondary hyperparathyroidism in CKD patients, as early detection can significantly impact patient outcomes.
- Surgical management of the overlying mass and underlying parathyroid gland hyperplasia should always be considered in the management of the symptomatic CKD-MBD patient.

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INTRODUCTION

Chronic kidney disease (CKD) has emerged as one of the most prevalent conditions worldwide, with an estimated prevalence of over 10%[1]. This condition can lead to serious complications, such as mineral bone disorder (CKD-MBD). CKD-MBD increases the mortality risk, cardiovascular complications, and progression to end-stage renal disease (ESRD)[2].

The manifestation of CKD-MBD varies across individuals presenting with CKD, so clinicians should be able to identify the condition. MBD is associated with worse outcomes in CKD patients[3,4]. The patient usually remains asymptomatic until bone disease appears. In rare cases the patient present with calcification or tumour formation, such as a brown tumour, as reported in the literatures[5-8]. Since this is a rare manifestation clinician are not familiar with it which potentially delays the recognition of worsening of CKD. In this case report, we describe the atypical finding of CKD-MBD manifested as a calcified tumour in the maxilla to improve the recognition of this condition.

CASE DESCRIPTION

A 32-year-old female presented to our internal medicine outpatient department with a progressive swelling in the left upper jaw that developed in the course of six months. The swelling was initially painless but steadily became more painful in the last two weeks, and she developed difficulties with mastication. No ulcer, discolouration, history of head injury or tooth removal was reported. The patient reported an unusual non-painful swelling in the neck for more than six months but no additional swelling in other body areas. The patient also reported mild pain in all of her joints. The patient was on routine twice-a-week haemodialysis, initiated seven years previously, in other nephrology departments. She was hypertensive since the first session of haemodialysis and was also receiving medication for this condition.

Physical examination revealed a non-tender dense osseous non-mobile 2 x 2.5 cm mass on the left upper maxillary body approximately on the level of the middle of the nasal and auricle. The overlying skin was normal and intact, with no induration, ulceration or fluctuation. Intraoral examination was unremarkable. Neck examination revealed a non-tender soft 1 x 1.5 cm mass on the midline that moved upon swallowing. The overlying skin was normal. General assessment of the patient was unremarkable, except for the hypertension.

Parameter	Value	Reference range
Haemoglobin (g/dl)	8.9	12 – 16
Urea (mg/dl)	111	5 – 20
Creatinine (mg/dl)	5.5	0.6 – 1.1
Natrium (mmol/l)	136	135 – 145
Potassium (mmol/l=	5.1	3.5 – 5.1
Calcium (mg/dl)	7.8	8.5 – 10.5
Phosphate (mg/dl)	6.6	2.8 – 4.5
PTH (pg/ml)	355	15 – 65
Vitamin D (ng/ml)	17	30 – 50

Abbreviation: PTH, parathyroid hormone.

Table 1. Laboratory parameters of the patient.

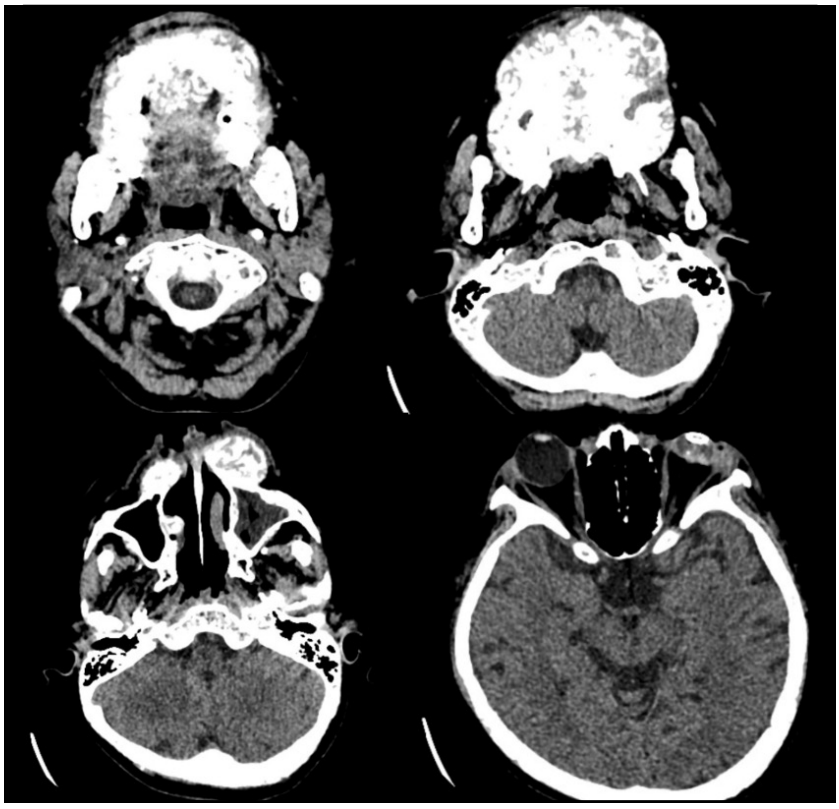

Figure 1. Multiple slices computed tomography scan (MSCT) of the head. MSCT revealed a mixed lytic and sclerotic calcified mass on the anterior part of left maxillary bone sized 3 x 2 x 2 cm, abnormal mixed lytic and sclerotic appearance of the calvaria, maxillofacial, and cervical bone, and calcification of the left eye lens with reduced eye bulb size.

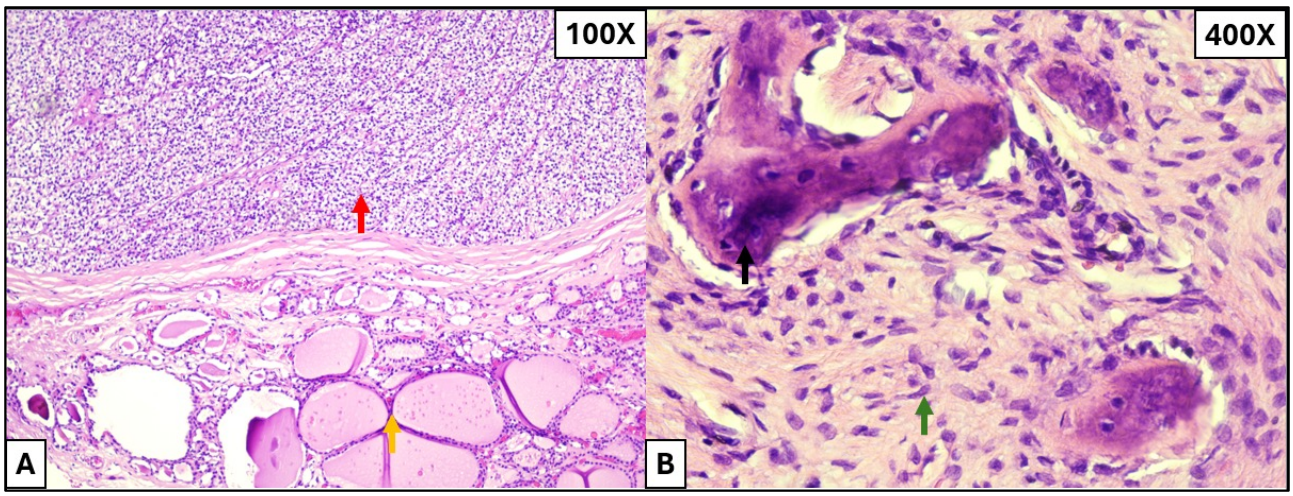

Figure 2. Haematoxylin and eosin histopathological analysis of (A) thyroid-parathyroid tissue and (B) maxillary tumour. (A) Hyperplasia of parathyroid characterised by increased proliferation of parathyroid glandular cells (red arrow) and thyroid hyperplasia characterised by proliferation of thyroid follicular glandular cells (orange arrow) were observed in x100 magnification. (B) Calcified maxillary fibroma characterised by rimming of osteoblast on bone trabeculae (black arrow) with per trabecular proliferation of fibroblasts (green arrow) was observed in x400 magnification of maxillary tumour tissue.

Initially, we hypothesized that the patient had a brown tumour. Further haematological analysis revealed mild anaemia. Biochemical analysis revealed a high level of parathyroid hormone (PTH), mild hypocalcaemia, moderate hyperphosphatemia, and low vitamin D *(Table 1)*. Additional multiple slice non-contrast computed tomography (CT) scan of the head revealed a mass *(Fig. 1)*. Further biopsy of maxillary and neck masses was performed and revealed maxillary calcified fibroma and hyperplasia of the thyroid and parathyroid *(Fig. 2)*. The diagnosis was maxillary fibroma with calcification, secondary to bone and mineral disorder complicating the ESRD.

Surgical excision of the parathyroid and the maxillary tumour was planned for the patient one week after the diagnosis. Unfortunately, the patient's condition deteriorated, and the patient was brought to the emergency room with severe dyspnea. After three days in the intensive care unit the patient's condition deteriorated further, and her family chose palliative care. The patient then died on the following day.

DISCUSSION

In this report, a soft tissue tumour arising in an ESRD patient initially thought to be a brown tumour was diagnosed as maxillary fibroma with calcification. Unfortunately, further planned surgical management of the patient was aborted due to the patient's deteriorating condition. CKD is a prevalent noncommunicable disease that leads to several health issues, such as hypertension, anaemia, hyperkalaemia, acidosis, and MBD caused by a decrease in the glomerular filtration rate. Untreated MBD increases the likelihood of mortality, cardiovascular incidents, and the progression of CKD to ESRD. The current patient was a routine haemodialysis patient for the past seven years, which increased her risk of developing complications related to MBD.

CKD-MBD is defined as one or a combination of the following: (i) abnormalities of calcium, phosphorus, PTH, or vitamin D metabolism; (ii) abnormalities in bone turnover, mineralisation, volume, linear growth, or strength; or (iii) vascular or other soft tissue calcification according to the Kidney Disease-Improving Global Outcomes (KDIGO) Foundation definition. Multiple guidelines have provided different target reference levels for PTH, phosphate, and blood calcium. In our case, the patient presented with a calcified mass, elevated PTH, and low calcium and phosphate deficiency These were sufficient to diagnose CKD-MBD according to current guidelines.

Brown tumour was one of the forms of high-turnover bone disease arising from a high PTH level in CKD patients. A brown tumour is a type of lesion that develops in individuals with hyperparathyroidism (HPT) and represents the latter stages of the bone remodelling process. The term "brown tumour" is used to describe the colouration of the tumour, which is caused by the presence of blood vessels, bleeding, and hemosiderin deposits. All of the bones in the skeletal system, including the bones in the craniomaxillofacial region, can be affected, as suspected in our patient. Only a few case reports from developing countries[6-9] have described this occurrence because routine blood screening for HPT is quite prevalent in developed countries. The CKD patient's previous screening data were unavailable due to a referral to our department. Consequently, the tumour was identified as the primary manifestation of the bone disorder.

As reported in previous cases, only clinical data presupposed the diagnosis of the mass. This is because brown tumours do not exhibit distinctive histologic alterations and cannot be differentiated histologically from other giant cell lesions, including fibrous dysplasia, giant cell tumours, and reparative granuloma. Hence, it is crucial to establish a conclusive diagnosis based on the patient's clinical history and subsequently verify it by biochemical and histopathological examination.

The primary changes in CKD–MBD involve hyperphosphatemia, hypocalcaemia, deficient amounts of vitamin D in the serum, and an excessive release of PTH from the parathyroid glands, usually due to secondary hyperparathyroidism (SHPT). In CKD and ESRD, HPT is mainly caused by SHPT, gastrointestinal malabsorption disorders, severe vitamin D deficiency, or insufficient calcium intake in the diet. Advanced CKD reduces Klotho concentration, which blunts the binding of fibroblast growth factor (FGF)-23 to the FGF receptor-Klotho complex in the parathyroid glands, which usually inhibits the synthesis of PTH[10]. The initial phase of SHPT is marked by widespread hyperplasia and the proliferation of parathyroid cells[11]. In our case, the parathyroid tissue biopsy revealed an adenoma of the thyroid, consistent with the development of HPT in the patient. During the early stage, pharmaceutical treatments such as phosphorus-lowering drugs, vitamin D analogues, and calcimimetics efficiently decreases the elevated levels of PTH in the bloodstream. Failure to correctly treat SHPT can lead to the development of nodular hyperplasia with adenomatous tissue, which can become resistant to treatment[6]. The adenoma morphology prompts the surgical removal of the parathyroid gland to prevent further HPT in the patient; thus, excision was scheduled.

SHPT, alongside other mineral imbalances, could result in adynamic bone disease (low bone turnover), osteomalacia (inadequate bone mineralisation), and osteitis fibrosis (high bone turnover). The calcified lesion in the maxillary fibroma in our patient was thought to be the intermediary phase of the remodelling process due to high bone turnover, similar to the pathogenesis of brown tumours. Classically, the high bone turnover has been described as rapid osteoclastic activity and per trabecular fibrosis leading to erosive bone lesions, as seen in our histopathological analysis, which results from HPT and represents a sort of local destructive phenomenon. This condition necessitates further surgery to minimise damage to the surrounding tissue. In our patient's case, excision was promptly planned once the diagnosis was made.

The unfortunate demise of the patient in this report limits our reporting of the outcome. As with CKD and its complications, the prognosis of the patient was already worse. In addition to the higher risk of bone fractures in this specific patient group compared to the general population, bone abnormalities also have essential implications for mortality and cardiovascular disease[3].

CONCLUSION

This study underscores the significance of clinical comprehension of the broad-spectrum manifestations of CKD-MBD, including an initially asymptomatic mass. In addition, screening of the patient's biochemical and bone mineral parameters was required to determine the necessity of early intervention and improve the patient's outcome.

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