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When parathyroid adenoma meets osteopathia striata, rarest amalgam of bony dysplasia with brown tumors: a case report

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Abstract

Background Osteopathia is a benign entity characterized by linear densities along the diaphysis and metaphysis of the longitudinal axes of the long bone. Parathyroid adenoma is a non-malignant parathyroid gland tumor characterized by increased parathormone levels, causing defecting calcium levels leading to multiple brown tumors, renal calculi, bone pains, polyuria, and arrhythmias in the patient. The coexistence of parathyroid adenoma with osteopathia striata has not been reported yet in the literature. To our knowledge, this is the first case of its kind.

Case presentation An 18-year-old male patient presented to our institution with the complaint of genu valgum deformity in his right knee and was admitted for corrective surgery. While undergoing a preoperative evaluation, we came across multiple focal, lytic lesions in his radiographs and an atypical finding of osteopathia striata in his long bones. Further examination led us to the findings of cranial sclerosis and acro-osteolysis of distal phalanges. The tumor, whose catalyst was unclear, directed us toward an ultrasonographic analysis of the patient's neck, leading us to a solitary parathyroid adenoma.

Conclusions Radiological examinations played a very crucial role in the diagnosis of this condition. Though the reason behind osteopathia and the parathyroid lesion in this patient remains unknown, this presentation remains an exceptional and never-reported case of parathyroid adenoma and associated brown tumors in a patient with osteopathia striata.

Keywords Parathyroid adenoma, Brown tumor, Osteitis fibrosa cystica, Osteopathia striata, Genu valgum, Osteochondrodysplasias

Background

A parathyroid adenoma involves a group of disorders affecting the parathyroid glands, mainly including gland hyperplasia or carcinomas [1]. The parathyroids are located in the neck, near or attached to the backside of the thyroid gland. These produce parathyroid hormone, which maintains the blood's calcium, phosphorus, and vitamin D levels [2]. The etiology for parathyroid adenoma can be radiation therapy, lithium drug consumption, chronic kidney disease, and the most common cause being genetic mutation, i.e., over-expression of cyclin D1 gene, all leading to the overactive parathyroid gland and altered parathyroid hormone (PTH) secretion [1]. Patients may be asymptomatic or manifest symptoms of hyperparathyroidism, which may be nephrolithiasis, polyuria, constipation, fatigue, bone pains, and sometimes neuropsychiatric disturbances. Severe findings include brown tumors, cardiac arrhythmias, coma, and death [1, 3].

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A brown tumor known as osteitis fibrosa cystica is a focal, benign, and erosive bony lesion caused by localized and rapid osteoclastic turnover due to hyperparathyroidism [3, 4]. Being one of the most pathognomonic signs, they are still a rare finding. The inappropriately increased secretion of PTH affects bone metabolism, leading to hypercalcemia and hypophosphatemia [3]. They appear as localized regions of bone loss with hemorrhage, reparative granulation, and proliferating fibrous tissue replacing the regular marrow content [3]. They often cause bone pain, swellings, and pathological fractures, usually localized to the mandible and maxilla regions [4].

Osteopathia striata is a rare sclerosing bone dysplasia with characteristic radiographic features of longitudinal sclerotic striations along the metaphysis and the diaphysis of long bones [5]. It represents a disruption in the process of the endochondral bone formation pathway [6]. There may be associated internal organ defects and nerve deficits [7]. To the best of our literature review, there are cases reported of osteopathia with osteosarcomas and cranial sclerosis, as well as parathyroid adenoma with other sclerosing bony dysplasia [6], but not of osteopathia with parathyroid adenoma.

Osteopathia, when not severe, is asymptomatic and not a life-threatening condition [6, 7]. It has been mainly diagnosed incidentally in patients with different complaints and pathologies [8], as noted in our case. Parathyroid adenomas, when asymptomatic, can go unnoticed for an extended period and cause severe deformities in patients if not diagnosed on time [1, 9]. The radiological modalities are crucial in diagnosing such conditions and deciding on further management courses.

Case presentation

An 18-year-old male patient presented to the orthopedic department of our institution for pain and associated deformity in the right knee, which was affecting his gait and daily routine. On clinical examination, he had genu valgum deformity, which was progressive for the past 2 years (Fig. 1). The patient was suggested a correction surgery for the same and thus came with radiographs of the right knee joint. Consequently, we came across a focal bony lesion in the left proximal tibia and linear striations

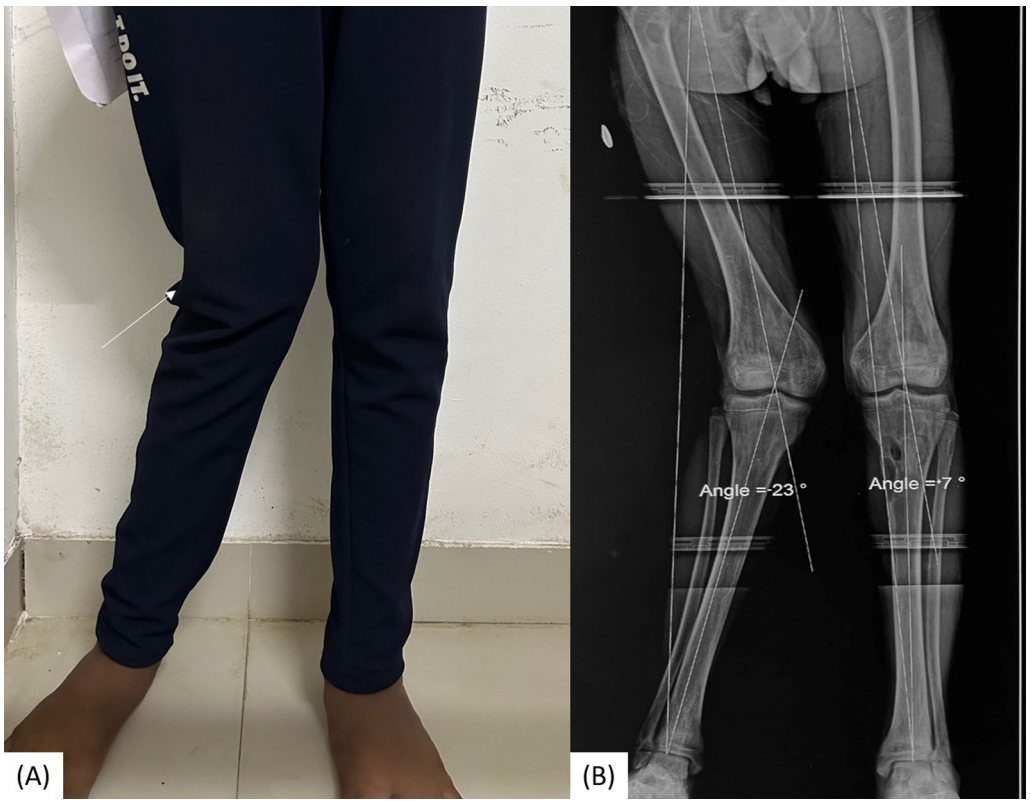

Fig. 1 An 18-year-old male patient came with complaints of pain in the unilateral lower limb and a significant genu valgum deformity in the right knee. He had no significant past or family history and was suggested a corrective surgery because of the progressive nature of the deformity affecting his daily routine. **A** Clinical images of the patient's lower limbs show a visible genu valgum deformity in the right lower limb (white arrow). **B** A scanogram of both lower limbs shows a hip knee ankle angle of 23 in the right limb

along the long axis of the distal femur and proximal tibia. We asked the patient to undergo a skeletal survey to confirm our suspicions. The radiologists reviewed the images, each with more than five years of experience.

On subsequent radiographs, the left proximal tibia revealed a well-defined multiseptated, lytic, eccentric lesion in the meta-diaphyseal region with a narrow transition zone; a similar lesion was discovered in the right proximal femoral metaphysis (Fig. 2a, b), indicating multiple brown tumors. We also discovered an uncommon finding of multiple vertical striations in the diaphyseal and metaphyseal region of the long bones of bilateral knee joints (Fig. 2a). The dense, linear, uniform striations running parallel to the shaft and fan-shaped striations in the wing of the iliac bone led us to the diagnosis of osteopathia striata (Fig. 2c). Decreased bony mineralization and sclerotic changes in the cranial vault and the vertebral end plates were observed. The vertebral bodies showed prominence of vertical trabeculae and loss of horizontal trabeculae (Fig. 3a–c).

We screened him for renal and parathyroid pathology to rule out genetic etiology. With no abnormal renal findings on abdominal ultrasound, we moved on to the neck ultrasound, where incidentally, we came across a well-defined, hypoechoic mass lesion measuring approx. 3×1.4 cm in the left inferior parathyroid gland with arterial vascularity within it (Fig. 4a). A polar vessel was seen supplying the lesion, pointing our diagnosis toward a solitary parathyroid adenoma (Fig. 4b).

When the child underwent a CT (Computed tomography) evaluation, we came across a well-defined lytic lesion in the ramus of the mandible, with a soft tissue component inside and expansion of the surrounding bony cortex, and a similar lesion was found in 2nd molar on the left side suggestive of brown tumors in the mandible (Fig. 5a,b). Also, calvarial thickening with the expansion of the inner and the outer table with multiple hyperdensities was noted (Fig. 5c). A well-defined hypo-enhancing nodule was seen posteroinferior to the left thyroid lobe with a CT density of 64 HUs, suggesting the parathyroid adenoma, thus confirming the ultrasonography findings. On contrast-enhanced CT imaging, a homogenously enhancing lesion was seen just posterior to the left sternoclavicular joint with an arterial supply through a branch from the left inferior thyroid artery (Fig. 6).

We concluded that the child had parathyroid adenoma, causing primary hyperparathyroidism and multiple brown tumors. The child was referred for PTH hormone and calcium levels, which were 224 pg/ml (reference range: 10–55 pg/ml) and 12 mg/dl (reference range: 8.5–10.5 mg/dl), respectively. He was earlier operated on for the parathyroid adenoma and would later be taken for the surgical correction of genu valgum deformity.

Discussion

Osteopathia striata, or Voorhoeve's disease, is an asymptomatic and incidental finding [8, 10]. This rare syndrome, to date, has been reported in only one hundred patients and is thought to be inherited as an autosomal dominant or X-linked dominant disease [6, 10]. It is occasionally associated with other syndromes and with sclerosis of the skull vault. Although striations in the long bones and cranial sclerosis are hallmarks of this

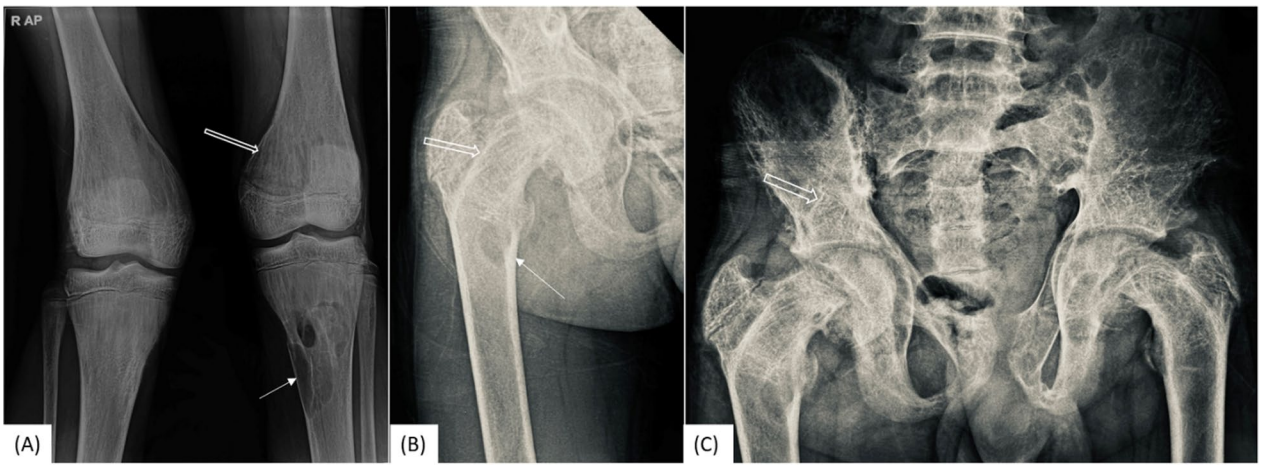

Fig. 2 AP X-ray of the bilateral knee and the right proximal femur of the same patient **A**, **B** depicting multiple vertical striations (white outline arrow) in the meta-diaphyseal region suggestive of osteopathia striata with a multiseptated, lytic eccentric lesion (white arrow) in the proximal region of shaft of the femur and proximal shaft of left tibia with thin transition zone indicating brown tumors. **C** Pelvis shows various fan-shaped striations (white outline arrow) along the wings of bilateral iliac bones, as seen in osteopathia and bilateral protrusio acetabuli

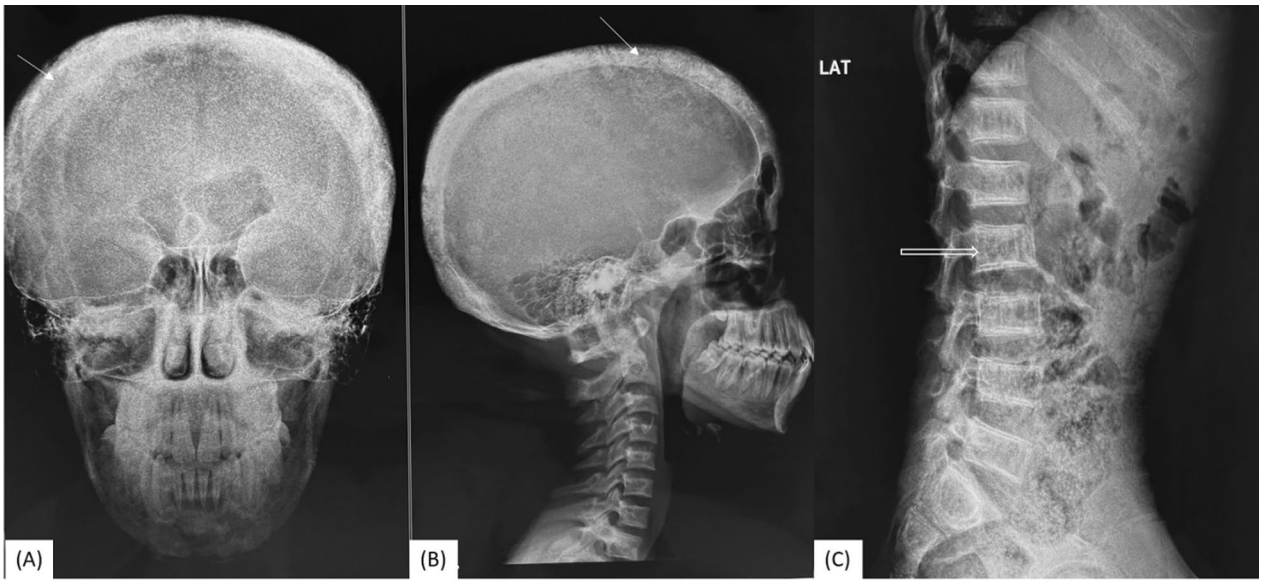

Fig. 3 X-ray of skull. **A** AP view, **B** lateral view, depicting diffuse sclerotic changes in the cranial vault, also known as "salt-and-pepper" appearance (white arrow), which is a typical finding of hyperparathyroidism. **C** Lateral view of the lumbo-sacral spine illustrating increased vertical trabeculae (white outline arrow)

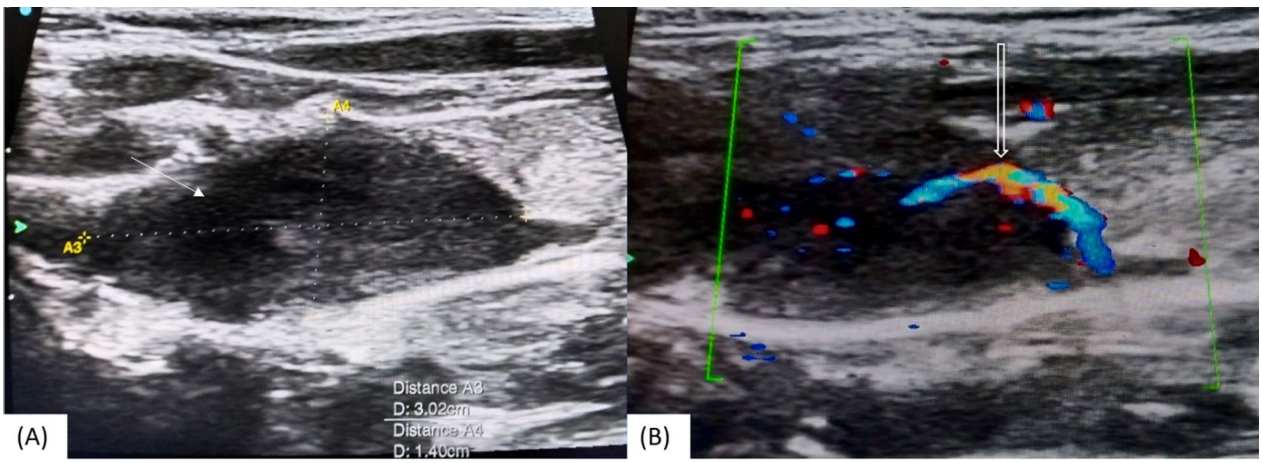

Fig. 4 Neck ultrasonography images in the same patient. **A** Depicts a well-defined, erosive, hypoechoic lesion of 3.02 × 1.4 cm in the left inferior parathyroid gland suggestive of a solitary parathyroid adenoma (white arrow). **B** A single polar vessel is seen supplying the lesion (white outline arrow), representing its arterial vascularity

condition, these radiographic findings are not always seen during the early stages, which impedes early diagnosis [11]. The radiological findings in osteopathia include fan-like striations in the pelvis, facial dysmorphism (macrocephaly, occipital bossing, mandibular prognathism, depressed nasal bridge), vertebral anomalies, club feet, absent or short fibula, various neurological, and internal organ defects [7]. However, in this case, it is not life-threatening and thus requires no treatment. Since there is a marked increase in bone density, ranging from chalky to extremely hard, some surgical instruments have been damaged due to such high bone densities. The changes in bone density can also influence the risk of post-surgical complications (nonunion, infection) and even the time required for surgeries in these patients [5]. Such surgery-related complications should be known and considered while determining the treatment plan.

Most parathyroid adenomas are functional and manifest symptoms of hypercalcemia, causing nephrolithiasis and bone pains. The primary pathophysiology in

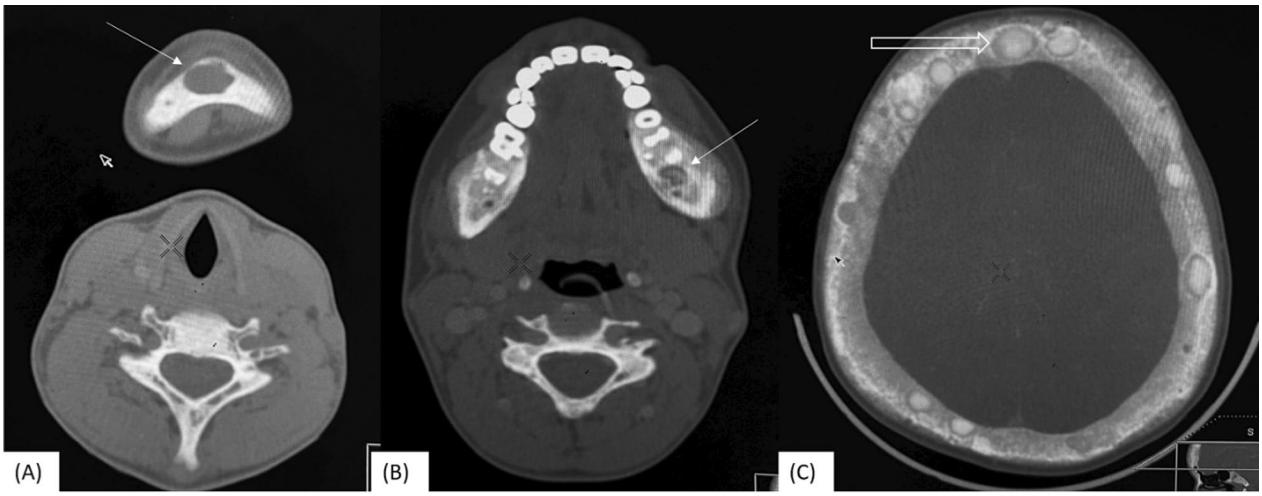

Fig. 5 Non-contrast computed tomography images in axial section. **A** A well-defined, lytic lesion in the ramus of the mandible with subsequent expansion of the bony cortex, suggestive of a brown tumor (white arrow). **B** Another radiologically similar lytic lesion can be seen on the left side, near the base of the 2nd molar (white arrow). **C** Also, increased calvarial thickening with multiple focal areas of hyperdensities within it (white outline arrow)is not to be missed

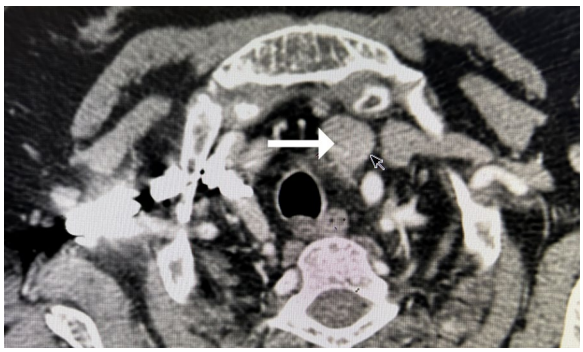

Fig. 6 Contrast-enhanced computed tomography of neck. Axial Section Image of the same patient depicts a solitary, homogenously enhancing nodule arising from the left inferior lobe of the parathyroid gland (white arrow)

such adenomas is the hypersecretion of the parathyroid hormone. The hormone secreted by the gland is inactive and secreted as soon as serum calcium level drops. The hormone is then cleaved from an initial pre-pro-PTH containing 115 amino acids to an active PTH containing 84 [2]. This active form leads to activation of the osteoclastic cells and inhibition of the osteoblasts. The osteoclasts are responsible for forming multiple lytic lesions due to calcium resorption from the bones, as seen in our case. We also detected increased serum calcium levels in the patient but without any severe symptoms of hypercalcemia, except for some episodes of constipation. The ultrasonography of the abdomen could not detect any renal calculi or biliary tract stones.

Hyperparathyroidism can be of three types: primary, secondary, and tertiary. Primary hyperparathyroidism (PHPT), the most common subtype with autonomous secretion of PTH, is usually asymptomatic. Bones are the main target organ of PTH, causing metabolic bone disorders and manifestations comprising subperiosteal bone resorption—acro-osteolysis, salt-and-pepper skull, subchondral bone resorption, brown tumors, and osteopenia due to osteoclastic action. A brown tumor corresponds to localized bone loss, creating a lucent area in the bone, which is replaced by richly vascularised, reparative connective tissue secondary to osteoclastic action. The brown color is characteristic of the hemosiderin deposition associated with the hemorrhages within this newly vascularised tissue. Such lytic lesions can be single or multiple and located at any site. The most common sites include the pelvis, ribs, long bones, mandibles, and sometimes vertebrae. A characteristic brown tumor on the radiograph is seen as a solitary, expansile, or multifocal well-defined soap-bubble imitating lesion with cortical thinning, a narrow transition zone, and without any periosteal reaction [9]. Similar lytic lesions were seen in our case, also noted on CT as circumscribed lytic lesions of soft tissue density. Another radiographic evidence of PHPT in our patient was the granular, salt-and-pepper appearance of the skull, suggestive of the focal area of patchy sclerosis.

On ultrasound, the parathyroid adenomas are mostly homogenously hypoechoic to the overlying thyroid gland due to the compact cellularity of the lesion [12]. The adenoma in the current case displayed a uniform

hypo-echogenicity with arterial vascularity in one of the left parathyroid glands. The remaining glands on the right side were normal-sized, leading to a parathyroid adenoma diagnosis rather than hyperplasia. There have been reports of similar lesions without any clinical signs of hyperparathyroidism [13]. The sensitivity and specificity of polar vessel sign in the prediction of parathyroid adenoma is 79.0% and 55.6%, respectively [14], and similar polar vessel could also be identified in our case. This case is rare because the patient showed no other clinical symptoms besides the brown tumors. There were no signs of bone pains, constipation, renal stones, or weakness. A radiologist should know about such rare conditions to provide timely diagnosis and encourage appropriate management. Surgical management is the most common and suitable treatment for such adenomas. And when operated on, the prognosis is good.

The patient was initially in pain due to surgery but, later on, was satisfied because the real cause of his condition was identified and treated before it led to severe disabilities and affected his daily routine. The serum calcium and PTH levels decreased significantly after the surgery; the calcium level was 10.4 mg/dl, and the PTH level was 92.53 pg/ml. The pain has gradually subsided, and he has decided to go further with genu valgum deformity correction soon.

Conclusions

Adenomas are the most common cause of primary hyperparathyroidism and may not always show systemic symptoms like renal stones or pathological fractures. A radiograph was the most crucial modality in diagnosing brown tumors. In such patients, apart from the biochemical investigations, USG and CECT of the parathyroid glands should be done to determine the cause of such lesions and differentiate between benign and malignant causes. Brown tumors are rare bony lesions in a patient with hyperparathyroidism and can be found at various locations with variable presentation. In addition to plain radiographs, CT can also be used to differentiate them from other bony lesions.

As osteopathia is a rare and asymptomatic finding in benign cases and was incidentally discovered in our patient, it has been reported in very few patients and is indeed a radiological curiosity. Our case was a rare occurrence of concurrent osteopathia striata and parathyroid adenoma in a male patient with multiple brown tumors and significant unilateral genu valgum deformity. When osteopathia striata is radiologically diagnosed, other internal anomalies and cranial osteosclerosis should also be ruled out to prevent any long-term and intraoperative complications. Evaluation and confirmation through three different modalities helped deduce the origin of brown tumors and avoid aggressive treatment.

Abbreviations

PTH	Parathyroid hormone
CT	Computed tomography
PHPT	Primary hyperparathyroidism
USG	Ultrasonography
CECT	Contrast-enhanced computed tomography

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Author contributions

Dr. MS and Dr. AS analyzed and interpreted the patient data regarding his condition and gave the diagnosis and were involved in the patient care. Dr. RC was the major contributor in drafting the manuscript and responsible for patient follow up. Dr. PB reviewed and approved the final version of the manuscript.

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Availability of data and materials

Some or all the data used to support the findings of the case report shall be made available by the corresponding author on reasonable request.

Declarations

Ethics approval and consent to participate

Not applicable.

Consent for publication

Consent was obtained from the patient and his guardian.

Competing interests

The authors declare that they have no competing interests.

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