ORAL MANIFESTATION OF RENAL OSTHEODISTROPHY IN CHILDREN

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ABSTRACT

Very rare in children, the brown tumor, or osteoclastoma, is an understudied ectopic entity that causes severe debilitation in patients with chronic renal insufficiency. We report two cases who developed particular forms with different evolutions of severe renal osteodystrophy.

Key words: *brown tumor, severe renal osteodystrophy, children, end-stage renal disease, secondary hyperparathyroidism.*

INTRODUCTION

Brown tumors are lesions that occur due to abnormal bone metabolism in secondary hyperparathyroidism. They represent the terminal stage of the bone remodeling process. The bones compromised by these lesions may be: the bones of the face, the clavicles, the ribs, the bones of the pelvis and the femur. From the histological point of view, the brown tumors are made up of fibrous tissue, bone trabeculae and vascular support, but without matrix. Brown tumors are not encapsulated. Lymphoma, plasmocytoma, and giant cell tumors should be ruled out within the differential diagnosis of brown tumor (1). The correction of secondary hyperparathyroidism usually results in regression of the tumors. Secondary hyperparathyroidism is described to occur in 92% of the patients undergoing dialysis, but incidence of brown tumors is just 1,5% in adult. in children the occurrence of brown

tumor is quite rare, as long as prolonged hemodialysis (HD) is not common (2). We report two cases of brown tumor in children, first in a 11-year-old boy, with severe renal osteodystrophy, that was not controlled by medications, and another in a 16-year-old girl with a good evolution.

CASES REPORT

The management of chronic renal failure (CRF) must prevents the development of serious forms of renal osteodystrophy. Treatment noncompliance may give rise to severe manifestations of bone involvement in uremic children with active osseus metabolism (3).

Patient 1 is a 13-year-old boy with end-stage renal disease (ESRD) due to hemolytic uremic syndrome, was on continuous ambulatory peritoneal dialysis (CAPD) for 5 years, and then on HD (because of recurrence of peritonitis) for another 2 years. Since the first presentation we showed short stature in context of ESRD, and severe renal osteodystrophy, clinically manifested by important bones deformities, with a typical radiological appearance. The clinical

examination at presentation reveals the presence of severe changes in context of severe renal osteodystrophy (dissociation with superior jaw protrusion, nasal pyramid collapse, costal mantle, metaphysis bracelet, genus valgum, 1/3 left femur left fracture). The biological parameters of the child confirm the renal osteodystrophy: high intact parathormone (iPTH) of 1602 pg/mL, serum calcium of 9.7 mg/dL and phosphorus of 8,5 mg/dL, high serum concentrations of alkaline phosphatase (2644 IU/l), low level of vitamin D 23 ng/mL. The recomended treatment for this complications of ESRD was poor diet in phosphorus, oral administration of calcium carbonate, like phosphorus binder therapy, calcitriol, corrections of metabolic acidosis, multivitamin supplements. The biochemical control of hyperparathyroidism was poor, because of the family's noncompliance at the diet and therapy.

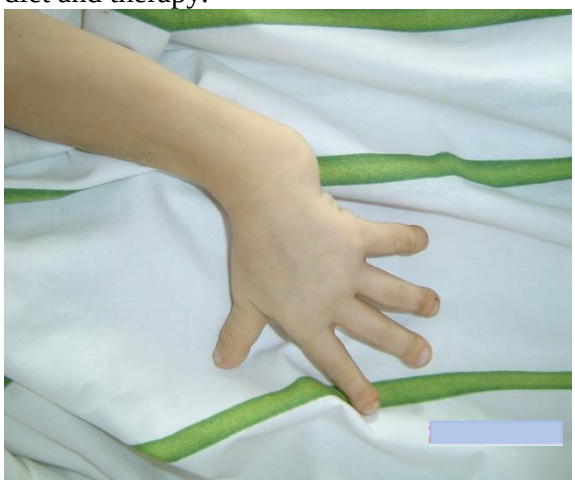


Fig. 1 Bones deformities at patient 1

The patient developed a swelling of the hard palate that increased in size gradually. In time this tumoral formation resulted in tooth loss, swallowing and phonation disorders. The swelling was soft, painless, but tender at palpation, with elastic in consistency (Fig.2)

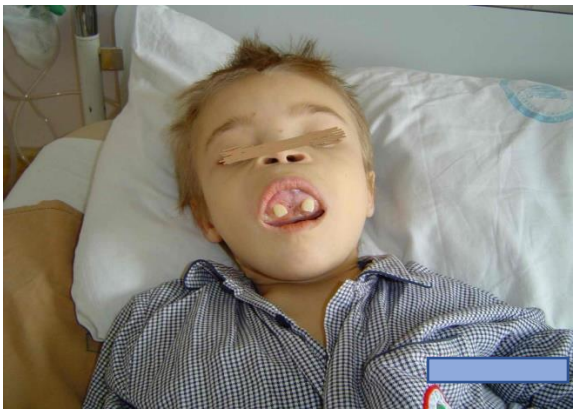


Fig. 2 Patient 1 – brown tumor of hard palate

A diagnosis of brown tumor of the hard palate was made. CT performed to exclude a possible expansive process concluded: fibrous dysplasia with the interest of the mandible, jaw and anterior floor of the lip, skull and calotte, without any intracranial expansive process. The tumor biopsy demonstrated lesions with giant cells that were fibroblasts with a rich vascular network, highly suggestive of a brown tumor. In the same time we discussed with the surgical staff, because the neck ultrasound showed the adenomatous development of parathyroids glands. After three months of parenteral therapy with activate vitamine D the result wasn't quite good, iPTH was reduced to 1852 pg/mL, but the oral mass was increasing. We proposed parathiroidectomy for control of severe ostheodistrophy, but the family refused. Unfortunately the boy died suddenly few weeks later, at home, because of odontoid fracture in context of severe osteodistrophy.

Patient 2 is a girl, 16-yers-old at diagnosis, in our clinic evidence since December 2000 (6 years of age) with chronic end-stage renal failure, caused by a chronic glomerulonephritis in the context of an immune vasculitis. She required hemodialysis in an emergency, then, from April 2001 she has been introduced into a chronic hemodialysis program on arteriovenous fistula. The clinical examination at presentation reveals the presence of severe changes in context of severe renal osteodystrophy (sinistroconvex scoliosis, walked licked, shortening inferior right

member) (Fig.3,4).

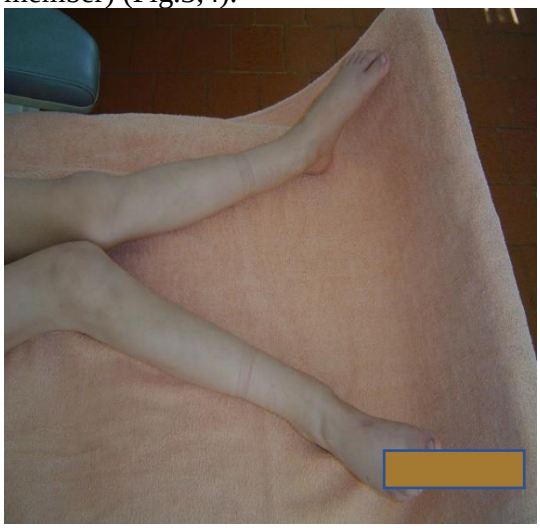


Fig. 3 Patient 2 – legs deformities – clinical aspect

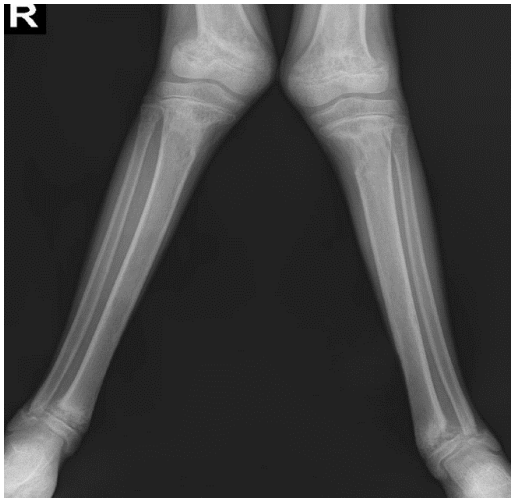


Fig. 4 Patient 2 – legs deformities - radiological aspect

She had a history of aseptic necrosis of the right femoral head at the age of 13 in the context of the brown tumour developed at that level (Fig.5). She benefited from orthopaedic therapy through osteosynthesis with brooches, then right femoral head prosthesis.

Fig. 5 Patient 2 – aseptic necrosis of the right femoral head

For several weeks, she has been accusing major headache, headquartered at the level of the tall central palate, and vertex irradiation, no influenced by the usual antalgic medication. A tumor was protruding through her oral cavity, in the right side of hard palate (Fig. 6), it had appeared insidiously 3 years ago and had increased gradually in size. The lesion was tender to palpate, painless. Some bleeding areas were observed on the fragile mucosa. The girl had functional problems with swallowing, chewing and speech. Serum chemistry revealed an elevated intacte parathyroid hormone (iPTH) level of 3753 pg/dl, serum calcium 9.08 mg/dl, phosphorus 5.2 mg/dl, and alkaline phosphatase 2740 IU/l.

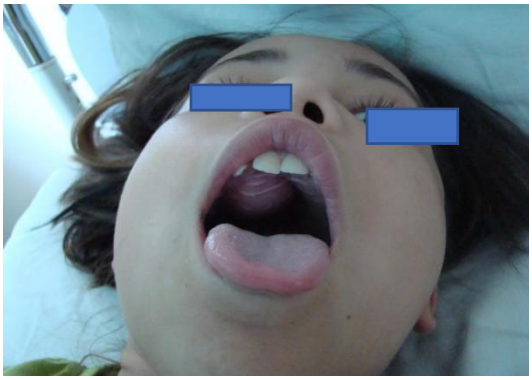
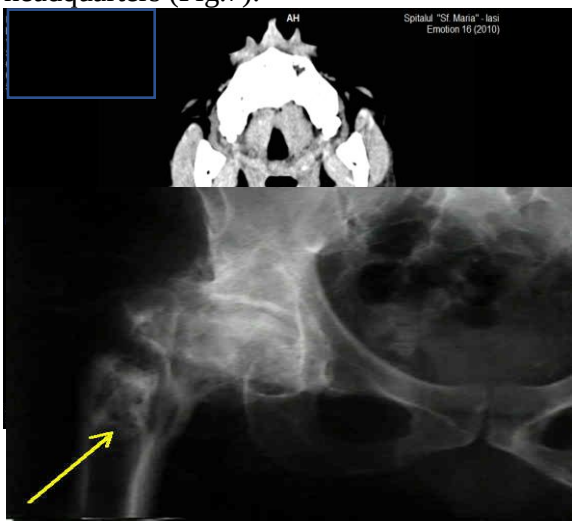


Fig. 6 Patient 2 – brown tumor of hard palate

Cranio-cerebral CT has been performed for excluded possible supra and subtentoral expansion processes. There were highlighted areas of hyperostosis suggestive of brown tumors with parieto-temporo-frontal headquarters (Fig.7).



palate

After two months of activate vitamine D therapy palatine tumor continued to increase and headaches persisted, so we proposed subtotal parathiroidectomy. Six months after surgery we constated complet remision of the brown tumora, but with residual hyperostosis. One year later the girl received a cadaveric renal transplant.

DISCUSSIONS

Brown tumors that occur in patients with ESRD represent an extreme form of osteodystrophy. They occur most often in the long bones, ribs and pelvis, but can be found in any bone (2). Clinically significant lesions in the pediatric population are rare, especially due to the short time spent in chronic dialysis, pending a kidney transplant. The diagnosis must to be as earlier as possible, and the treatment with activated vitamin D and/or surgergy should be administered rapidly. Orejas described in 1993 the smallest case of brown tumor (a 3-year-old girl with peritoneal dialysis) in which family noncompliance with therapy and diet determines the death of the patient (3). The similar aspect was in our first case. In order to develop an accurate management plan and to achieve a good prognosis we need to consider early diagnosis, effective communication with stakeholders, as well as reflection upon the moral values (4). Decision to determine what is best or not for the child can be very difficult, the respect for the child's interest supposes sometimes ignoring the child's disagreement when for example, a surgical act is essential for saving his life (5). Clinical manifestations of abnormal bone metabolism in secondary hyperparathyroidism in ESRD are nonspecific, with majority being asymptomatic for long time. The definitive treatment of brown tumor is parathyroidectomy as studies have shown that 80% of brown tumors result from parathyroid adenoma while parathyroid hyperplasia and co-existence of hyperplasia and adenoma account for the rest (6, 7, 8). In our second case the rapid surgical intervention with subtotal excision of parathyroid glands saved the child. Despite the parathyroidectomy and optimal control of calcium/phosphate metabolism, some brown tumor did not decrease in size during follow-up (9). In our second patient residual hyperostosis was described in CT scan after 6 months after parathyroid surgery. In this case the renal transplant remain the unique chance for bones rehabilitation. Finally the whole patient approach should be part of the internal ethics audit, meaning the process aimed to improve the organization's performance (10).

CONCLUSIONS

1. The brown tumor remains a striking complication of osteodystrophy in the ESRD.

2. The diagnosis should be suspected when we have uncontrolled hyper-parathyroidism, and must to confirmed by CT scan, biopsy and specific serological changes.

3. The parathyroidectomy seems to be the most appropriate treatment for this pathological entity.

4. The renal transplant must to be the final decision making in cases resistant to parathyroidectomy and optimal control of calcium/phosphate metabolism.

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