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Localized Jaw Enlargement due to Fibrous Dysplasia in a Hemodialysis Patient

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Abstract

Osteitis fibrosa is a well documented complication of hyperparathyroidism secondary to chronic renal failure. It preferentially affects the long bones, the spine and the ribs. Maxillofacial bones are rarely affected and leads to facial deformities. We report the case of a 15–year–old female patient who developed hyperparathyroidism secondary to end-stage renal disease that manifested as unilateral jaw enlargement. Histological appearance is that of a fibrous dysplasia.

Keywords: Child, Fibrous Dysplasia, Renal Osteodystrophy, Secondary Hyperparathyroidism

Introduction

Renal osteodystrophy (ROD) is a well-recognized long-term complication of end-stage renal disease (ESRD) in children (1). However, extreme forms such as facial fibrous dysplasia are very uncommon (2). A review of the English-language literature revealed few cases in dialysis patients during the first two decades of life. In the present paper we report the case of a 15–year–old female patient who developed hyperparathyroidism secondary to end-stage renal disease which manifested as unilateral jaw enlargement. To the author's knowledge, this is the youngest reported patient.

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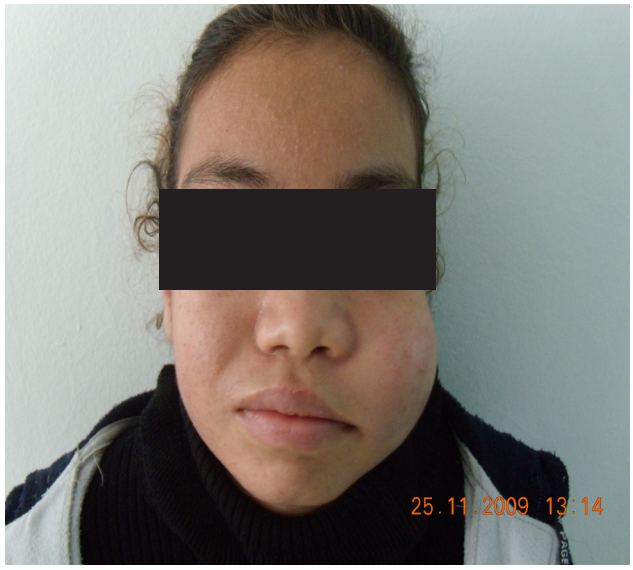
Case report

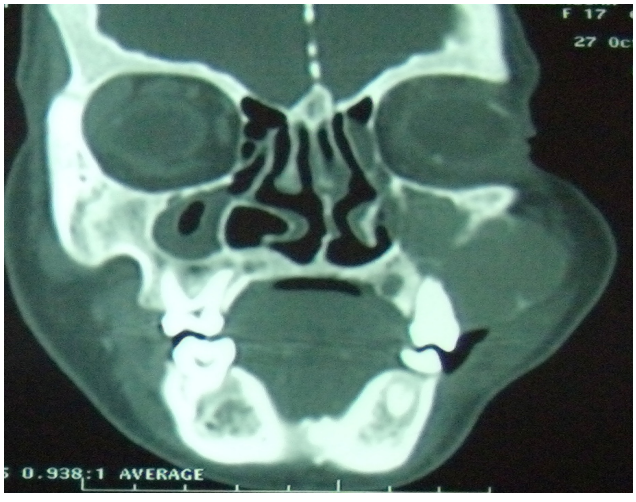
A 15–year–old female on a regular haemodialysis programme had been followed for chronic renal failure secondary to Nephronophtisis type I since the age of 5 years. She was undisciplined, and did not take regularly her treatment. Therefore, she soon developed secondary hyperparathyroidism. Her parathyroid hormone (PTH) was measured as 1600 pg/ml (normal level 15-68pg/ml). The alkaline phosphatase level was more than 1200 IU / l. Serum calcium and phosphorus concentrations were 1.81 mmol/l and 2.5mmol/l, respectively. In January 2009, a hard localized asymptomatic enlargement of the left maxilla appeared. Its progressive growth caused a marked deformity of the face (figure 1). Radiography of the face showed an osteolytic lesion of the left maxilla. A first computed tomographic scan of the skull showed lytic lesion of the left maxilla measuring 42x39 mm surround by a peripheral condensation. It had intense enhancement after injection of contrast. Moreover, there were abnormalities suggestive of osteodystrophy in the mandible and the base of the skull. The diagnosis was that of a brown tumor. A cervical ultrasound revealed a normal thyroid and a nodule of the parathyroid. Parathyroidectomy was performed in April 2009. The postoperative evolution was marked by a normal level of PTH, serum calcium and phosphate, but the swelling did not decrease until after 6 months. The patient had a renal transplantation from cadaveric donor in May 2009 with good outcome.

Figure 1: Facial view reveals prominence of left jaw



A second CT scan was performed due to the persistence of the swelling despite the normalization of phosphorus-calcium balance. It revealed the presence of an ossified mass filling the left maxillary sinus reaching the floor of the orbit and down to the maxillary alveolar (figure 2). Due to the unusual evolution of a brown tumor and the changing of the images in the scan, a biopsy of the maxilla was performed. It revealed a fibro-osseous lesion composed of irregular curved individual spicules of bone within fibrous tissue. The diagnosis of fibrous dysplasia was therefore retained.

Figure 2: CT of maxilla and cranium shows a mass of ossified filling and distorting the left maxillary sinus measuring 5 x 4 cm



Discussion

Renal osteodystrophy is still a major problem in children with progressive chronic renal failure (3). It combines features of secondary hyperparathyroidism, rickets, osteomalacia, and osteoporosis. Several disorders have been retrieved including abnormalities in mineral metabolism and development of bone disease. These alterations occur early in the course of chronic renal failure and are accompanied with subsequent bone deformities and growth failure (4). The deformities often affect the long bones, ribs and pelvis. Clinically significant lesions in the jaws are uncommon especially in children (2). A review of the English-language literature revealed only four cases in dialysis patients less than twenty years of age. The clinical characteristics of all cases, including the present are summarized in table 1.

The impairments of facial skeletal are usually unapparent. When clinically evident, they pose diagnostic difficulties as regards the nature of the underlying histological lesion (9). The osteitis fibrosa cystica is the most common form which is characterized by a combination of increased bone cell activity, peritrabecular fibrosis, and cystic brown tumors (10). The second form is termed fibrous dysplasia in which the radiological diagnosis is relatively difficult (11). The latter form is exceptional and caracterized by significant hypertrophy of the jaws called leontiais ossea (6). In our patient, the difficulty was clinical and radiological diagnosis as a brown tumor has been discussed due to the history of ESRF and the radiographic finding in the first CT scan. However, after parathyroidectomy, the volume of maxillary swelling didn't improve after parathyroidectomy, although some authors have reported a progressive decrease in the size after parathyroidectomy (12). Secondly, the clinical and radiological appearance was not in favor of leontiasis ossea since deformation was not bilaterally symmetric. A maxillary biopsy was indicated to solve the problem of the differential diagnosis. The

Table 1: Age, sex, jaw involvement, histopathologic features, surgical intervention and outcome of jaw enlargement in five patients aged less than 20 years

References	Age (years)	Sex	Location	Histopathologic features	Parathyroidectomy	Surgical excision	Out- come
Damm et al (5)	19	Male	Mandible + Maxilla	Not done	None	None	Lost to follow-up
Aggunlu et al (6)	19	Male	Mandible + Maxilla	Osteitis fibrosae	+	None	?
Benjelloun et al (7)	17	Female	Mandible + Maxilla	Brown tumor	+	None	Progressive decrease in the size of tumor
Tarrass et al (8)	18	Male	Mandible	Brown tumor	+	None	Progressive decrease in the size of tumor
Present case	15	Female	Mandible + Maxilla	Osteitis fibrosae	+	None	No change

unilateral facial deformity was attributed to fibrous dysplasia. It therefore appears that the histological study, although it is not a common assessment, is very sensitive to understand the pathophysiology of renal bone disease (13).

Conclusions

Renal osteodystrophy must be considered in the diagnosis of bilateral or unilateral jaw enlargement. Bone biopsy although is not routinely performed, it is strongly recommended in this situation.

Conflict of interest

None declared.

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