Severe Hyperparathyroidism Associated with Fibrous Dysplasia; A Case Report

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Dear Sir,

Hyperparathyroidism (HPT) as a main etiologic factor of uremic bone disease may cause serious morbidity in uremic patients. On the other hand, fibrous dysplasia (FD) of bone is a disease of unknown etiology in which fibrous tissue replaces normal bone elements and its association with primary HPT has been rarely reported before [1]. In this report, we present a hemodialysis patient with severe HPT concomitant with craniofacial FD causing abnormal fascial deformity.

A 26-year-old woman was admitted to our hospital with a history of progressive difficulty in walking, widespread bone pain and abnormal enlargement of her face. Her history also revealed that she had been on regular hemodialysis therapy for 6 years because of renal failure of unknown etiology. Physical examination showed abnormal bilateral enlargement of the maxilla and mandible as well as multiple nodules palpable in the thy-roidal region (fig. 1). Laboratory studies were as follows; serum calcium 7.6 mg/dl, serum phosphorus 6.3 mg/dl, alkaline phos-phatase 1,560 U/l (normal 98-279), and parathyroid hormone (intact PTH) 1,749 ng/ml (normal 12-72). Bone roentgenograms disclosed extensive typical signs of HPT including widespread Brown tumor formation. Computerized tomography showed significant enlargement of all parathyroid glands as well as a well-trabeculated tumoral pattern in the maxilla and mandible. A whole-body bone scan was obtained following intravenous injection of 20 mCi ^{99m}Tc methylenedi-phosphonate. All scintigraphic features of renal osteostrophy including multiple nodules

Fig. 1. Bilateral maxillary and mandibular enlargement.

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Fig. 2. The whole-body bone scan shows extensive skeletal involvement of severe hyperparathyroidism including Brown tumor formation in the ribs (arrows).

due to brown tumors in thoracic ribs, besides intense tracer uptake in maxilla and mandible, are shown in figure 2. Mandibular biopsy showed FD.

A history of long-term hemodialysis, the presence of hypocalcemia despite severe HPT and hyperplasia of all parathyroidal glands in radiologic investigations may be regarded as the significant diagnostic clues of secondary HPT in this case. But one cannot differentiate primary HPT due to hyperplasia of all parathyroidal glands from secondary HPT due to renal failure.

The well-known example of the association of FD and primary HPT is the McCune-Albright syndrome. This syndrome is characterized by precocious sexual development, fibrous dysplasia and many endocrinological abnormalities such as goiter, hyperthyroidism, HPT, acromegaly, Cushing's syndrome and hyperprolactinemia [2]. In the present case we could not detect any endocrinological abnormality except for HPT. As far as we know, except for the McCune-Albright syndrome, concomitance of HPT and FD had been reported in 6 patients before with no acceptable theory to explain the association of these two clinical conditions and 1 of these patients had had the diagnosis of chronic renal failure [3].

FD and HPT are two main clinical conditions that should be considered in the differential diagnosis of facial skeletal abnormalities. The cystic form of FD may be confused with the radiologic signs of the facial involvement of HPT [2]. In the present case, the radiologic appearance does not show any cystic component, instead it forms a well-tra-beculated pattern rather typical for FD. Besides, in view of the grotesque facial architecture commonly associated with FD, and the pathological diagnosis of FD, we thought FD as a possible diagnosis rather than HPT in this subject. But it must not be forgotten that both diseases may cause similar radiological and histological appearances in facial bones [4]. So, follow-up of the changes in her facial bone disease after parathyroidectomy is going to lead us to reach a clear-cut diagnosis.

References

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