Letter to the Editor

A Lethal Complication, Acute Necrotizing Pancreatitis, of Turner's Syndrome with Primary Hyperparathyroidism

To the Editor;

A 46-year-old female with Turner's syndrome was referred to our hospital regarding coexisting hyperparathyroidism. The patient complained of systemic bone pain but no physical signs were found on her abdominal examination when admitted. She showed extremely short stature (113 cm tall and 20.5 kg weight) with severe kyphosis, and mental retardation and poor development of secondary sexual characteristics since her childhood. Blood examination exhibited normochromic anemia, metabolic acidosis, severe hypercalcemia (14.3 mg/dl) with hypophosphatemia (1.8 mg/dl) and remarkable increase in PTH secretion (intact PTH, 1029 pg/ml; normal 14–66) but no renal or hepatic dysfunction was detected. Her karyotype was 45X. Bone roentgenogram revealed multiple bone fractures including severe lumbar compression and extended bone damage due to osteitis fibrosa was also found in all her limbs and skull. Cervical ultrasonography and CT scan exhibited a cystic micro-nodule in her right thyroid region, in which ^{99m}Tc -sestamibi were clearly accumulated. Immediately af-

ter admission, administration of pamidronate disodium (15 mg/day, three times for four weeks) and elcatonin (80 IU/day) was performed with continuous infusion of physiologic saline and diuretics in order to improve her severe hypercalcemia (Fig. 1). One week after admission, she suddenly complained of abdominal discomfort with remarkable rise in serum pancreatic enzymes including amylase (1330 IU/L) and lipase (270 IU/L). Abdominal CT defined acute necrotizing pancreatitis with massive bloody ascites. She was intensively treated with continuous infusion of gabexate mesilate (300 mg/day), ulinastatin (25000 IU/day), and antibiotics with plasma transfusion. However, the acute necrotizing pancreatitis was progressed irreversibly, leading to the subsequent complication of disseminated intravascular coagulation, which unfortunately resulted in her death.

There has been no report regarding Turner's syndrome with serious pancreatitis related to concomitant hyperparathyroidism, since only a few Turner's cases complicated with primary hyperparathyroidism have ever been documented to date [1]. In the present case, hypercalcemia was resistant and coexisting necrotizing pancreatitis abruptly progressed even during treatment. Regrettably, we could not obtain further information regarding her calcium metabolism. However, given that bone formation and mineralization by endogenous factors including estrogen and vitamin D are impaired in Turner's syndrome [2], the weakness of systemic bone remodeling could be well implicated particularly in aged Turner's cases like ours [3]. This might have

Fig. 1. Clinical course and serial change of serum levels of calcium and amylase.

in part lead to resistant hypercalcemia due to excessive bone resorption induced by PTH hypersecretion, resulting in fulminant pancreatitis through activating intrinsic trypsinogen and/or disrupting intracellular homeostasis in the pancreatic acinar cells [4]. As shown in the reported Turner's cases that developed chronic cholestasis or sclerosing cholangitis [5], the acute-progressive pancreatitis in our case led us to consider possible malformation in the pancreatic/choleangial ducts. Thus, aged Turner's patients with primary hyperparathyroidism must be carefully monitored and intensively treated while paying attention the possibility that this rare but lethal complication of acute necrotizing pancreatitis may suddenly develop. Both early diagnosis and intensive therapy from the outset of acute pancreatitis are absolutely necessary in order to prevent fatal complications in Turner's cases having underlying fragility in bone metabolism.

References

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