**Introduction:** Tyrosinemia type I presents with hepatic and renal failure, growth retardation, and rickets in infants and young children. Death in the untreated child is usually due to liver failure, neurological crisis, or hepatocellular carcinoma. 42% of the children with tyrosinemia type I may have recurrent neurological crises. Typical manifestations of these crises are altered mental status, abdominal pain, peripheral neuropathy, and/or respiratory failure requiring mechanical ventilation. 10% of deaths in untreated children occur during a neurological crisis.

Vitamin C is the cofactor in biosynthesis of collagen, catecholamines and iron metabolism. Copper containing dopamine, which catalyzes dopamine conversion from noradrenalin is a cofactor for beta hydroxylase. The reason for irritability and changes in behavior and consciousness in cases of vitamin C deficiency is the disruption of this tract. Scurvy can be defined by bone pains and aversion to physical touch. The most important reason for this is subperiosteal hemorrhage and significant osteoporosis. In scurvy, bone radiology has idiosyncratic characteristics, and typical images include significant osteoporosis, cortex as if drawn by a very fine-tipped pencil; central osteoporosis in metatarsus and metacarpus, circle-like image in the periphery with the fine cortex (Figure 3).

Ascorbic acid supports reduction activation of hepatic p-hydroxyphenylpyruvic acid (p-HPPA) oxidase, the enzyme that converts p-HPPA to homogentisic acid in the catabolism of tyrosine. Thus, when tissue ascorbate concentrations are diminished, the activity of p-HPPA oxidase is impaired. Hypertyrosinemia and tyrosyluria are likely to occur, particularly in the newborn, when the activity of p-HPPA oxidase is insufficient.
Case report: The patient, who has been followed up with a diagnosis of tyrosinemia type 1 for 8 months, was admitted to the emergency department for 1 month due to constipation and the need to use laxatives. Globe vesicale was also detected in the emergency room (Figure 4,5). It was reported that the patient used his diet and NTBC treatment regularly. It was observed that he also had gait disturbance in his control in our outpatient clinic.

Neurological, oncological, traumatological, infectious, hemtological, orthopedic etiological causes were eliminated with examinations. In further analysis, increased tyrosine and low NTBC levels were detected. (Tyrosine level:237 mcmol/L N: 31-106, dry blood succinylacetone: <1 mcmol/L , n: <1, nitisinone level 17 mcmol/L , n: 30-60, organic acid analyses in urine by GCMS 4hydroxyphenyllactic acid: 994 mmol/mol kreatinin n: <3.6, 4 hydroxyphenylpyruvic acid : 291, n: < 0,3). It was learned that NTBC was added to his Formula and the patient did not finish his food. Although the application errors were corrected and high tyrosine and low NTBC levels were corrected. (Control tyrosine:113 mcmol/L). It was inspected that the patient who constantly cried and did not touch him. We was learned that he did not consume fruit (Figure 1). Vitamin C was found in the lower limit in the blood. (Vitamin C level: 0,3 ng/dl, control 1,3 ng/L N: 0,5-1,5). Neurological complaints improved in 1 month with diet, NTBC and Vitamin C treatment (Figure 2).
Both tyrosinemia and vitamin C deficiency can cause osteoporosis and restlessness. In cases where the two are together, bones and neuropsychiatric findings may occur with more prominent and mixed findings. In our patient, it was thought that constipation and globe vesicale were due to misuse of nitisinone, impaired gait was associated with tyrosinemia, restlessness and not touching oneself were due to vitamin C deficiency. The patient's nutritional history provided guidance in interpreting the data and requesting examinations. Laboratory examinations revealed low levels of NTBC, vitamin C, and high tyrosine levels in the blood and high levels of tyrosine breakdown products in the urine. Clinical findings improved rapidly with the correction of nutritional habits and replacement of NTBC and Vitamin C in the follow-up.
Severe osteoporosis
Figure 3. Typical vitamin C deficiency findings in the radiological imaging of the long bones of our patient's legs are shown in the figure.

Figure 4, 5. The abdominal MR imaging of the patient's neurogenic bladder and constipation signs, which we defined as tyrosinemia neurologic crisis signs, are shown in the figure.
Conclusion: This case report was made to draw attention to vitamin C deficiency and application errors in the neuronopathic attack of tyrosinemia. Suspicion of the presence of scurvy may contribute positively to the diagnosis and treatment in patients who deteriorate rapidly and/or do not achieve the desired result in clinical improvement despite appropriate NTBC and diet therapy.

References:

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