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Letter to editor

Celiac Disease Associated with Pycnodysostosis

Erradi Mariam[1,3*], Abourazzak Sana[2,3], Hida Moustapha[2,3]

[1]Department of Neonatology and Neonatal Intensive Care Unit, Mother-Child Hospital, HASSAN II University Hospital of Fez, Morocco; [2]Department of Pediatric, Mother - Child Hospital - HASSAN II CHU of Fez, Morocco ; [3]Department of Medicine and Pharmacy, Mother-Child Hospital,HASSAN II University Hospital of Fez, Sidi Mohamed Ben Abdellah University, Sidi Harazem, Fes 30070, Morocco

LETTER TO EDITOR

To the editor

The celiac disease manifests itself by several symptoms, among them the staturoponderal delay [1]. However, any sort of relationship between celiac disease and pycnodysostosis has not been described in the medical literature until now.

A 7-year-old girl, who consults for staturoponderal delay with abdominal pain. The anamnesis highlighted the existence of alternating diarrhea constipation with abdominal pain. The clinical examination revealed a statural and weight delay at >-2 decistere, a cranial perimeter at +2 decistere, frontal bumps, short limbs, atrophic extremities and irregular and cracked nails, a discreet abdominal distension without hepato-splenomegaly, a teething disorder (dental malposition and carries). The standard radiological examinations revealed an absence of welding of the anterior fontanel, abnormally wide sutures and hypoplasia of the lower jaw, a lysis of the distal phalanges. Biological examinations: Did not objectify anemia, on the other hand antibodies against transglutaminase (Ig A) very positive. The esophageal-duodenal fibroscopy objectified an atrophy of the duodenal and gastric mucosa, gastric and duodenal biopsies were made. The histological examination is in favor of celiac disease. Thus, the diagnosis of celiac disease associated with pycnodysostosis was retained. The patient benefited from a gluten-free diet, with regular clinical and biological follow-up.

After a review of the literature, no reference has been found of the association between celiac disease and pycnodysostosis. Pycnodysostosis is a very rare hereditary condensing osteopathy, affecting both sexes. Its prevalence is approximately 200 affected individuals have been reported in the medical literature. Pycnodysostosis is estimated to affect about 1-17 individuals per million [2]. Autosomal recessive inheritance, parental consanguinity is described in 30% of cases [3]. Formal diagnostic criteria for pycnodysostosis have not been established, however the radiographic features of acroosteolysis, osteosclerosis, and loss of the normal angle of the jaw are almost pathognomonic [2]. The arguments allowing to make the diagnosis are: Clinical, with predominant statural delay in the limbs which are short and massive, fontanelle open or close late, characteristic facial dysmorphia [4]. The treatment of pycnodysostosis is personalized according to the type of lesions and surgical and non-surgical complications. Celiac disease is an autoimmune enteropathy caused by gluten gliadin in genetically predisposed people whose extra intestinal manifestations are increasing [5]. Celiac disease is often accompanied by autoimmune diseases as well as various associated pathologies. At the first line of the autoimmune diseases, we classify type 1 diabetes, but also Hashimoto's type thyroiditis, autoimmune hepatitis, lupus and many other autoimmune pathologies. Statural delay is the most frequent extra intestinal manifestation in children found in 10 to 40% of cases. The pathogenesis is multifactorial, based essentially on the malabsorption syndrome and the lack of action of growth hormone, the level of which is relatively low [5]. The clinical suspicion must be confirmed by serological tests and the diagnosis requires the performance of an esophageal-duodenal fibroscopy and a histopathological analysis confirming the latter. Currently, the treatment remains exclusively dietary which is the strict gluten-free diet for life. Hence the interest of systematic screening in order to preserve the prognosis of patients as much as possible.

Kind regards,

Erradi Mariam

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Correspondence to: Erradi Mariam, Department of Neonatology and Neonatal Intensive Care Unit, Mother-Child Hospital, HASSAN II University Hospital of Fez, Morocco; Department of Medicine and Pharmacy, Mother-Child Hospital, HASSAN II University Hospital of Fez, Sidi Mohamed Ben Abdellah University, Sidi Harazem, Fes 30070, Morocco, E-mail: erradi.mariam@gmail.com

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