CO-EXISTENCE OF KARTAGENER SYNDROME WITH CELIAC DISEASE: A CASE REPORT

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Case Report

Abstract:
Kartagener's syndrome and celiac disease, also referred to as gluten enteropathy, are two separate conditions with dissimilar clinical manifestations. The combination of celiac disease and Kartagener's syndrome in a 14-year-old girl presents a fascinating medical case. The patient shows stunted growth and has a history of situs inversus and dextrocardia. She also suffers from chronic diarrhea. Tests revealed a lack of growth hormone and vitamin D as well as anemia. High levels of anti-tissue transglutaminase antibodies were also found. These findings support the diagnosis of Kartagener's syndrome. Imaging studies confirmed sinusitis and bronchiectasis considering the dextrocardia. Doctors treated her with supplements and low-dose steroid shots, along with a gluten-free diet. This approach helped to ease her symptoms. While no clear link exists between Kartagener's syndrome and celiac disease, this case suggests a possible connection. It highlights the need to study these unusual conditions further to uncover any potential relationships.

INTRODUCTION

A chronic inflammatory disorder called celiac disease or gluten enteropathy arises when the immune system reacts to gluten, a protein present in wheat and other cereals. The inflammation of mucosa cells in the prox-imal small intestine leads to malabsorption of nutrients, a common indicator of this condition.[1-3] Celiac disease presents with recurrent diarrhea, weight loss, and even develop-mental issues with children; steatorrhea is also likely noted due to fat malabsorption. Health practitioners often recommend tests such as anti-tissue transglutaminase (TTG) IgA test for celiac disease detection since it boasts high 98% specificity and sensitivity rates. An endoscopic duodenal mucosal biopsy stands as the gold standard for confirming diagnosis, a practice deeply rooted within diagnostic norms.[3] The most important treatment for celiac disease is a gluten-free diet and nutritional supplements forever, in some instances doctors can also prescribe steroids orally or through injections to ease inflammation and pain which might be necessary if symptoms are severe.[1,2,4-6] Kartagener's syndrome is an uncommon genetic disorder that runs in families. It is characterized by long-term sinus inflammation, lung damage, and organs that sit in mirror positions to where they normally would. This disorder is also known as "immotile cilia syndrome" because to alterations in genes such as DNAI[1] and DNAH[5], which have a significant influence on how cilia move.[7,8] Because of these genetic alterations, cilia are unable to filter mucus and other foreign particles from the lungs and airways. This causes more frequent sinus and lung infections, makes it difficult to bear children, and causes body components to be positioned differently than usual.[9,10] In this case study, a peculiar and complicated medical condition involving a 14-year-old girl with both celiac disease and Kartagener's syndrome is examined. These two disorders together demonstrate how difficult it may be to manage several health issues at once. It also emphasizes the

significance of carefully designing a customized treatment strategy for every patient.

Case Report:

A 14-year-old girl presented with complaints of six years of terrible diarrhea and stunted develop-ment. She had three to four episodes of watery diarrhea with foul-smelling, partially digested food particles after consuming wheat and its derivatives. She experienced relief from her issues after cutting off gluten. Without a full examination, the patient was only sometimes prescribed medicine by community clinics. The patient had recurrent chest infections, nasal congestion, situs inversus, and dextrocardia from birth. She still developed normally cognitively and physically despite these restrictions, and she did well academically. She was the only sick sibling out of her family of five. Upon doing a physical assess-ment, it was discovered that she weighed 26 kg and was 4 feet tall. Her vital signs were stable, and her blood glucose level was 107 mg/dL. The general physical examination revealed low stature, typical proportions, and mild pallor. The peak beat could be felt in the fifth intercostal gap on the right side. A chest examination indicated coarse crepitations at the lung bases on both sides. The abdomen was swollen, but soft and not tender. There was no visceromegaly, although bowel noises were accentuated. The evaluation of the central nervous system was unremarkable.

Laboratory investigations revealed anaemia, with a haemoglobin level of 9.1g/dl (normal range: 11.5-16.5g/dl), MCV of 60fl (normal range: 75-95 fl), and HCT of 32% (normal range: 34-47%). The serum biochemistry was mainly normal, with the exception of 226 U/l of alkaline phosphatase (normal range: 0-187 U/L). The ESR was 55 mm/hour. The patient showed significantly high serum anti-tissue transglutaminase IgG (>200AU/ml, normal range <10AU/ml) and IgA (>130.8AU/ml, normal range <10AU/ml). Her vitamin D levels were severely low at 5.99ng/ml (normal range: 20-100ng/ml), as were her growth hormone levels of 0.425mg/ml (normal range: 2-5mg/ml).

Situs inversus was discovered with ultrasound imaging of the abdomen and pelvis, while X-ray chest and HRCT (high-resolution computed tomography) scans revealed dextrocardia and early bronchiectatic alterations in the lower lobes. A CT scan of the paranasal sinuses (CT PNS) indicated symptoms of sinusitis in the left frontal, bilateral maxillary, ethmoid, and sphenoid sinuses.

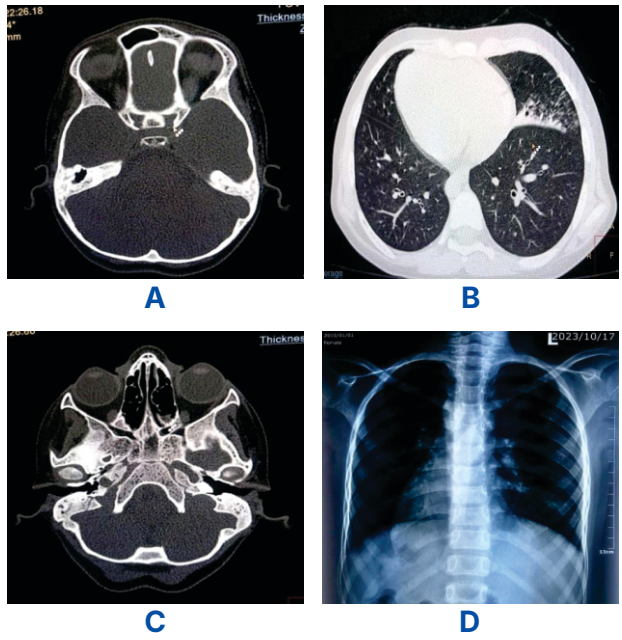

Figure A. CT PNS showing left Frontal sinusitis.
Figure B. CT PNS showing Ethmoidal sinusitis.
Figure C. HRCT showing early Bronchiectasis.
Figure D. X-ray Chest showing dextrocardia.

A diagnosis of celiac disease was made based on the patient's clinical history, examination, and laboratory results, as the patient's parents refused to have an endoscopy and jejunal biopsy. Furthermore, the presence of the characteristic triad of bronchiectasis, dextrocardia, and situs inversus led to the diagnosis of Kartagener syndrome. The patient was instructed to eat gluten-free and to take low-dose steroids, as well as calcium and vitamin D supplements. Fortunately, the patient's symptoms improved noticeably within 3-4 days. To address the growth hormone shortage, the child was sent to an Endocrinologist and will be continuously watched for the duration of his care.

Discussion

Celiac disease is an autoimmune disorder caused by a lack of tolerance to gluten. It is also classified as malabsorption disease as nutrients are not absorbed by the body. Celiac disease is a hereditary disorder due to sensitivity to gluten. The body's immune system damages the small intestine when individuals with gluten intolerance eat foods containing gluten.[11] The small intestine is damaged by the immune system, resulting in the loss of tiny finger-shaped protrusions called villi on the lining of small intestine. Diagnosis of celiac disease is based on symptoms and signs, laboratory and X-ray studies, confirmed by biopsy showing flat mucosa, and followed by clinical and histological improvement on a gluten free diet.

The incidence of celiac disease within the Pakistani population is not documented speci-fically although it effects a vast majority of both pediatric and adult population.[2],[11]Usually presents with diarrhea, steatorrhea and weight loss, it can also be asymptomatic or present with minimal symptoms like anemia, abdominal pain or bloating which can lead to misdiagnosis [6]A study was done in Pakistan which reported that CD was often misdiagnosed as IBS. (10) CD treatment is lifelong Gluten free diet with regular follow ups.[4] GFD is expensive and not affordable for most patients in Pakistantherefore, often leading to non-compliance and loss of follow up. [10] Kartagener syndrome (KS) is a subset of a larger group of ciliary movement disorders called primary ciliary dyskinesis. It is an inherited autosomal recessive disease that causes disorders of ciliary movement and consists of triad of sinusitis, situs inversus, and bronchiectasis. [12] Kartagener syndrome incidence lies between 1 in 20,000 to 1 in 30,000 births. [12] Most cases are diagnosed during early childhood due to recurrent pulmonary infections. Early diagnosis helps in prognosis and better management. Since there is no definitive cure for this, an early diagnosis and regular follow ups help us monitor the disease progression and prevent complications to some extent. [8] A late diagnosis of developed bronchiectasis worsens the prognosis and the complications associated can greatly affect quality of life of the patient.

While there is no proven connection between Kartagener syndrome and celiac disease, their co-occurrence in this patient is unusual. Due to limited resources, additional study into their link is not possible, leaving this topic open for exploration. This case study intends to investigate how these variables may interact, offering insight on a previously unknown relationship.

Conclusion:

In conclusion, the confluence of celiac disease and Kartagener's syndrome is a unique and intriguing scenario. Inflammatory reaction on gluten characterizes immune-mediated celiac disease, whereas Kartagener's syndrome is an autosomal recessive disorder that affects ciliary motility. These two cases appearing simultaneously may be suggestive of potential connections and shared underlying mechanisms. To understand their relationship well, research should be done further on whether there are potential associations between celiac disease and Kartagener's syndrome.

Author's Contributions
AI: Study conception and design
HN: Data collection and write up
TA: Data collection, literature review
FK: Drafting, critical review, and write up

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