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Received: 22.02.2019
Accepted: 07.03.2019
Published: 31.12.2019

Chronic perianal lesions in a normally developing 17-year-old boy

Przewlekłe zmiany okołodbytowe u prawidłowo rozwijającego się 17-letniego chłopca

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Abstract

We present a case of a 17-year-old so far healthy and normally developing boy, who was admitted to our Department due to perianal fistulas persisting for three years. Previous diagnostic procedures, including colonoscopy, failed to provide the diagnosis. No improvement was observed after repeated surgical interventions and oral antibiotic therapy. A suspicion of Crohn's disease was raised. Full diagnosis was performed: gastroscopy and colonoscopy with biopsy, magnetic resonance enterography, and pelvic magnetic resonance. Crohn's disease was diagnosed based on the overall clinical picture and laboratory findings. The aim of this paper was to emphasise the importance of early targeted diagnosis to avoid diagnostic delay and thus reduce the risk of complications. A general practitioner, who provides primary care and refers his patients for additional tests and consultations, plays a very important role in this process. Therefore, by describing the case of this young patient, we aimed to draw attention to some typical symptoms that may be the very first manifestations of chronic inflammatory bowel disease. The chronic or recurrent perianal lesions in an otherwise normally developing adolescent showing no other gastrointestinal symptoms should be a motivation for performing full diagnosis for Crohn's disease.

Keywords: Crohn's disease, children, perianal lesions

Streszczenie

W pracy przedstawiono opis przypadku 17-letniego, dotychczas zdrowego i prawidłowo rozwijającego się chłopca, który został przyjęty do Kliniki z powodu utrzymujących się od trzech lat przetok okolicy odbytu. Wykonane dotychczas badania diagnostyczne, w tym kolonoskopia, nie pozwoliły ustalić rozpoznania. Wielokrotne leczenie chirurgiczne i antybiotykoterapia doustna nie przyniosły poprawy. Postawiono podejrzenie choroby Leśniowskiego–Crohna. Wykonano pełną diagnostykę: gastroskopię i kolonoskopię z pobraniem wycinków, enterografię rezonansu magnetycznego oraz badanie rezonansu magnetycznego miednicy mniejszej. Na podstawie całości obrazu klinicznego oraz wyników badań dodatkowych zdiagnozowano chorobę Leśniowskiego–Crohna. Celem niniejszej pracy jest podkreślenie wagi wczesnej, ukierunkowanej diagnostyki, która pozwoli uniknąć opóźnień w rozpoznaniu i dzięki temu zmniejszyć ryzyko wystąpienia powikłań. Niezmiernie istotna jest w tym procesie rola lekarza sprawującego podstawową opiekę nad pacjentem, kierującego go na badania dodatkowe i konsultacje specjalistyczne. Dlatego też, opisując historię choroby młodego pacjenta, autorzy chcieli zwrócić uwagę na pewne charakterystyczne objawy, które mogą być pierwszymi symptomami przewlekłej zapalnej choroby jelit. Należy podkreślić, że występowanie przewlekłych czy nawracających zmian okołodbytowych u prawidłowo rozwijającego się nastolatka, przy braku innych dolegliwości ze strony przewodu pokarmowego, powinno skłaniać do wykonania pełnej diagnostyki w kierunku choroby Leśniowskiego–Crohna.

Słowa kluczowe: choroba Leśniowskiego–Crohna, dzieci, zmiany okołodbytowe

INTRODUCTION

We present a case of a 17-year-old boy with chronic, recurrent perianal lesions, who was diagnosed with Crohn's disease 3 years after the onset of first symptoms. Crohn's disease (CD) belongs to a group of inflammatory bowel diseases (IBD). Although the lesions are most often located in the final section of the small intestine and the initial section of the large intestine, they may occur at any location in the gastrointestinal (GI) tract. Paediatric patients account for about 25% of all diagnosed cases⁽¹⁾. Only 25% of the youngest patients present with the classic triad of symptoms (abdominal pain, weight loss and chronic diarrhoea), which is typically seen in this disease⁽²⁾. Others develop untypical, extra-intestinal or poorly pronounced symptoms. Isolated perianal lesions may develop as the only initial manifestation of the disease in up to 50% of cases⁽³⁾. This poses a significant challenge for doctors as the risk of severe disease and complications is higher in children than in adults. Undiagnosed and untreated inflammation may lead to, among other things, systemic disorders affecting growth and maturation⁽⁴⁾.

The diagnosis is based on the Porto criteria, including: clinical symptoms with anthropometric measurements, endoscopy (upper and lower GI tract with biopsy and histopathological assessment performed by two independent pathologists) as well as GI imaging (GI passage, enteroclysis, intestinal ultrasonography, computed tomography, and magnetic resonance)⁽⁵⁾.

The activity of Crohn's disease is assessed based on the Pediatric Crohn's Disease Activity Index (PCDAI), which includes the severity of abdominal pain, diarrhoea and bleeding, limitation of daily activity, abdominal tenderness on palpation, and the presence of perianal lesions⁽⁶⁾.

CASE REPORT

A 17-year-old boy was admitted to the Department of Paediatrics, Gastroenterology and Nutrition due to perianal fistulas persisting for three years.

The boy showed proper development (fifth pregnancy, fifth delivery, an Apgar score of 10), had no history of diseases, vomiting, abdominal pain or unexplained articular pain. He was physically active (regular training) and had good appetite. Body weight: at the 50th percentile, height: at the 10th–25th percentile, body mass index BMI: at the 50th–75th percentile.

Over the past three years, the boy was repeatedly hospitalised in the Department of Paediatric Surgery and Department of Paediatrics in the district hospital. He underwent multiple surgical interventions in connection with the lesions located in the left buttock region, with subsequent diagnoses of an abscess, a pilonidal cyst and a fistula of this area. No improvement was achieved despite repeated incisions of the lesions, drainage and oral antibiotic therapies. The buttock lesion did not heal properly and exudation

occurred periodically. Furthermore, the boy developed a furuncle on his left thigh, which was successfully treated with oral antibiotic therapy (Amoksyklav). Physical examination showed no abnormalities other than the left buttock lesion. Laboratory tests showed slightly increased erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) as well as minor anaemia and thrombocytosis. Colonoscopy was performed under general anaesthesia and showed no abnormalities. Abdominal and inguinal lymph node ultrasonography was normal.

Due to unsuccessful treatment of the fistulas located in the region of the left buttock a decision was made to admit the boy to the Department of Gastroenterology for further diagnosis. The patient was in good overall condition on admission. Physical examination showed isolated acne lesions on the trunk; anal sphincter region with no inflammatory changes; a visible seton placed on the left buttock; minor oedema and a palpable thickening at the incision site, tender on compression; and a trace of purulent drainage; other than that no abnormalities were found. Laboratory findings were as follows: slightly increased CRP, calprotectin 437.7 µg/g. Abdominal ultrasonography showed large bowel wall thickening in the caecal region.

Due to the reported pain in the region of fistula, soft tissue ultrasonography was performed and showed a developing abscess. The boy was transferred to the Department of Surgery for an urgent intervention.

Further diagnostic investigations were planned: magnetic resonance (MR) enterography (a contrast-enhanced thickening up to 0.5 cm of the final 5-cm section of the ileum wall likely to correspond to IBD lesions, and blurred fat signal in the region adjacent to these lesions, probably corresponding to small infiltrative IBD lesions were found); pelvic MR (a complex anal fistula with an internal opening 3 cm from the anal canal and an external opening in the region of the left buttock; infiltrative lesions of perineal soft tissues); gastroscopy (multiple haemorrhagic erosions in the region of gastric cardia and gastric body as well as in the prepyloric region); and colonoscopy (rectal and sigmoid mucosa with isolated aphthous erosions; other than that, normal colonic mucosa; ileocaecal valve oedema, probably stenotic; an unsuccessful attempt to pass through the valve). Specimens for histopathological examination were collected during colonoscopy (mild-to-moderate chronic inflammation of the large bowel; no characteristic granulation tissue typical for Crohn's disease).

The patient was diagnosed with Crohn's disease based on the overall clinical picture.

DISCUSSION

It is worth noting that symptoms in the form of perianal lesions in an otherwise healthy, normally developing, well-nourished and physically active boy do not suggest Crohn's disease in the first place. However, chronically persistent lesions in this region and the lack of proper healing despite

surgical interventions and oral antibiotic therapy should be a motivation to extend the diagnosis to include IBD. Properly conducted diagnostic process is also important. Correctly performed colonoscopy showing no mucosal lesions does not exclude Crohn's disease and further diagnostics should be done. Endoscopy of the upper GI tract is necessary to exclude mucosal lesions. It should be noted that Crohn's disease can affect any part of the GI tract, therefore lesions may be endoscopically detected in both its upper and lower sections. MR enterography and pelvic MR to assess perianal lesions, which in the case of our patient allowed for establishing the diagnosis despite scarce clinical symptoms, are another important diagnostic stage.

Once the diagnosis was made, treatment was initiated by including mesalazine. Due to the persistent active fistula a decision was made to intensify the therapy by including azathioprine⁽⁷⁾; however, no improvement was observed. Perianal lesions not responding to surgical treatment, antibiotic therapy and immunosuppressants may be treated with biological agents regardless of the clinical severity of the disease based on the PCDAI score⁽⁸⁾. After receiving the second dose of infliximab, the patient developed a late adverse reaction: eyelid oedema, generalised urticaria and limb tremor. Due to the mentioned symptoms a decision was made to discontinue infliximab and include adalimumab instead. Gradual, proper healing of the anal fistula was observed during subsequent follow-up visits.

CONCLUSIONS

Currently, an increasing number of cases of atypical Crohn's disease are reported. The aim of this paper was to draw attention to the fact that recurrent perianal lesions in the absence of other GI symptoms require full diagnosis for Crohn's disease.

We emphasise the importance of early targeted diagnosis, which will help avoid diagnostic delay and thus reduce the

risk of complications. A general practitioner, who provides primary care and refers his patients for additional tests and consultations, plays a very important role in this process.

Conflict of interest

The authors do not report any financial or personal connections with other persons or organisations, which might negatively affect the contents of this publication and/or claim authorship rights to this publication.

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