



Recurrent Fever and Abdominal Pain: Periodic Fever Syndrome, Inflammatory Bowel Disease or Patent Urachus; A Case Report

Khosro Rahmani ¹, Mohsen Jari ², Reza Shiari ¹, Mehrnoush Hassas Yeganeh ¹ and Vadood Javadi ^{1,*}

¹Department of Pediatric Rheumatology, Shahid Beheshti University of Medical Sciences, Tehran, Iran

²Department of Pediatrics, Imam Hossein Children's Hospital, Isfahan University of Medical Sciences, Isfahan, Iran

*Corresponding author: Mofid Children's Hospital, Shariati Ave, Higher Than Hosseiniyah Ershad, P.O. Box: 1546815514, Tehran, Iran. Tel: +9822227033, Email: vadoodj@gmail.com

Received 2023 July 24; Accepted 2023 October 07.

Abstract

Introduction: The urachus is a duct that connects the allantois to the urinary bladder in the fetus. Partial or total obliteration failure of the urachus leads to various anomalies, which can be discovered in children and adults.

Case Presentation: A 12-year-old boy presented with recurrent fever and abdominal pain two years ago. In the physical examination, he had tenderness in the lumbosacral area and Achilles tendons. Enthesitis-related arthritis (ERA), inflammatory bowel disease (IBD), and familial Mediterranean fever (FMF) were considered differential diagnoses. The patient was treated with a naproxen anti-inflammatory dose. After 15 months, his mother reported a bad odor discharge from his umbilical area. Investigations revealed patent urachus, and he underwent an operation that confirmed the diagnosis and resolved the patient's complaints.

Keywords: Patent Urachus, Abdominal Pain, Recurrent Fever, Children

1. Introduction

Abdominal pain is one of the most common symptoms in children with various etiologies, and the urachal remnant is a rare cause of it. Patent urachus incidence is less than 1 in 1000 live births (1). Urachus is an embryological remnant of the allantois (2). The lumen of the urachus is normally obliterated during embryonic development, transforming the urachus into a solid cord. Patent urachus occurs due to incomplete obliteration of the urachus (3, 4). These children may present at birth with a giant umbilical cord.

On the other hand, it may present in older children as different clinical features such as periumbilical granulation, urine excretion, abdominal pain, purulent discharge, swelling and erythema (as a result of a fistula), sinuses or cysts, an abdominal mass, fever, persistently wet or draining umbilicus, and occasionally a urinary tract infection. Pain, as well as retraction of the umbilicus during micturition, has been described as a sign of a urachal anomaly (5). Surgical therapy, including ligation and transection of the patent urachus or remnant at the level of the bladder, is generally recommended to avoid

infectious complications, skin breakage, and potential malignant degeneration (1). Herein, a teenage boy is reported with presenting features of recurrent fever and abdominal pain who finally was diagnosed and was treated as patent urachus.

2. Case Presentation

A 12-year-old boy with abdominal pain and recurrent fevers (every 3 - 4 weeks lasting 1 - 3 days) was referred to pediatric rheumatology since two years ago. In family history, he had an aunt who had two abortions and was suspicious of having systemic lupus erythematosus (SLE). He had not any history of previous admission. In the physical exam, he had lumbosacral and bilateral Achilles tendon tenderness, but his joints didn't have any tenderness or swelling. He fulfilled the Beighton and Shiari-Javadi criteria for generalized joint hypermobility (6). All laboratory data were in the normal range.

Furthermore, lumbosacral magnetic resonance imaging (MRI) was normal. He was prescribed naproxen 20 mg/kg for 15 months with the primary diagnosis of ERA. The recurrent fever attacks stopped with no

significant relief of recurrent periumbilical abdominal pain. His mother reported a bad odor and a little discharge around his umbilical area during urination. After that, an ultrasonography was performed, and a patent urachus was detected in the abdominal wall. So, the patient underwent surgery, and histopathology showed the remnant of the urachal duct with fibrosis. In 6-month follow-up after surgery, he did not have any complaint of abdominal pain.

3. Discussion

Recurrent abdominal pain is a common problem in children, with a prevalence of 2% - 41%. About 4% to 25% of school-aged children experience recurrent abdominal pain that interferes with their life activities (7, 8). Partial or total obliteration failure of the urachus is a rare cause of abdominal pain in childhood and adolescence.

Herein, we present a boy with recurrent fever and abdominal pain, enthesitis, and generalized hypermobility. The differential diagnoses were hypermobility spectrum disorder (HSD), enthesitis-related arthritis (ERA), inflammatory bowel disease (IBD), and periodic fevers. Despite generalized joint hypermobility, the attacks of fever and abdominal pain cannot be described with this diagnosis, and it seems that hypermobility is an accidental finding. Enthesitis was one of the significant signs in physical examination, but in these situations' recurrent infections due to immunodeficiency, organ malfunctions, and malignancies should be excluded. Furthermore, enthesitis-related arthritis (ERA) is defined according to the inclusion and exclusion criteria of the International League of Associations for Rheumatology (ILAR) classification. Due to the lack of peripheral arthritis, sacroiliitis, uveitis, positive family history of the HLA B27-associated disease, and negativity of HLA B27 in the patient, the ILAR criteria for ERA were not fulfilled. Considering the lack of recurrent diarrhea, blood and mucus in stool, weight loss, anemia, and elevated inflammatory markers, the diagnosis of IBD can be ruled out. Because of finding a definite diagnosis for the origin of the patient's fevers, the diagnosis of periodic fever was ruled out. He had a malformation, which was the etiology of his recurrent fever and abdominal pain. With surgery and removal of the patent urachus, the signs and symptoms of the patient disappeared.

3.1. Conclusions

In approaching children with recurrent fever and abdominal pain, the physician should be alert about

the etiology, which can be a wide range of structural problems, infections, immune deficiencies, malignancies, and periodic fever syndromes. Patent urachus is one of the malformations which can be presented as recurrent fever and abdominal pain.

Acknowledgments

We thank the patient and his parents for consenting to publish this case study.

Footnotes

Authors' Contribution: KR, MJ, RS, and MHY acquired medical records and clinical data; literature research. MJ wrote the manuscript. KR and VJ were the guarantors of the integrity of the entire study manuscript editing. The final manuscript was read and approved by all authors.

Conflict of Interests: The authors declare no conflict of interest.

Funding/Support: This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Informed Consent: Informed consent has been obtained from the parents of the patient.

References

1. Kokoska E, Weber TR. Umbilical and Supraumbilical Disease. In: Ziegler MM, Azizkhan RG, Allmen DV, Weber TR, editors. *Operative Pediatric Surgery*, 2e. 2nd ed. New York, NY: McGraw-Hill Education; 2014. 485 p.
2. Nguyen M, Addicott B, Chu J, Parham D, Kim E. Congenital Cyst of the Umbilical Cord. *Fetal Pediatr Pathol*. 2016;35(5):344-7. [PubMed ID: 27494161]. <https://doi.org/10.1080/15513815.2016.1188871>.
3. Ueno T, Hashimoto H, Yokoyama H, Ito M, Kouda K, Kanamaru H. Urachal anomalies: ultrasonography and management. *J Pediatr Surg*. 2003;38(8):1203-7. [PubMed ID: 12891493]. [https://doi.org/10.1016/s0022-3468\(03\)00268-9](https://doi.org/10.1016/s0022-3468(03)00268-9).
4. Korterink JJ, Diederik K, Benninga MA, Tabbers MM. Epidemiology of pediatric functional abdominal pain disorders: a meta-analysis. *PLoS One*. 2015;10(5). e0126982. [PubMed ID: 25992621]. [PubMed Central ID: PMC4439136]. <https://doi.org/10.1371/journal.pone.0126982>.
5. Cilley RE. Disorders of the Umbilicus. *Pediatric Surgery*. Sixth ed. Elsevier Inc; 2006. p. 1143-56. <https://doi.org/10.1016/b978-0-323-02842-4.50075-9>.
6. Javadi Parvaneh V, Shiari R. Proposed modifications to Beighton criteria for the diagnosis of joint hypermobility in children. *Indian Journal of Rheumatology*. 2016. <https://doi.org/10.1016/j.injr.2016.03.009>.
7. Konijnenberg AY, Uiterwaal CS, Kimpen JL, van der Hoeven J, Buitelaar JK, de Graeff-Meeder ER. Children with unexplained chronic pain: substantial impairment in everyday life. *Arch Dis Child*. 2005;90(7):680-6. [PubMed ID: 15899922]. [PubMed Central ID: PMC1720481]. <https://doi.org/10.1136/adc.2004.056820>.
8. Cuda SP, Vanasupa BP, Sutherland RS. Nonoperative management of a patent urachus. *Urology*. 2005;66(6):1320. [PubMed ID: 16360476]. <https://doi.org/10.1016/j.urology.2005.06.121>.