Cite as: Potyrała PJ, Puła M, Pytrus T, Kukawczyńska-Noczyńska J, Sharma G, Kofla-Dłubacz A: Right atrial myxoma in a paediatric patient with inflammatory bowel disease – a case report. Pediatr Med Rodz 2023; 19 (4): 447–450. DOI: 10.15557/PiMR.2023.0069.

Piotr Jan Potyrała[1], Michał Puła[1], Tomasz Pytrus[2], Joanna Kukawczyńska-Noczyńska[3], Girish Sharma[4], Anna Kofla-Dłubacz[2]

Received: 19.07.2023
Accepted: 12.12.2023
Published: 29.12.2023

Right atrial myxoma in a paediatric patient with inflammatory bowel disease – a case report

Śluzak prawego przedsionka u pacjenta pediatrycznego z nieswoistym zapaleniem jelit – opis przypadku

[1] Thoracic Surgery Department, Lower Silesian Oncology Centre, Wrocław, Poland
[2] 2nd Department of Paediatrics, Gastroenterology and Nutrition, Faculty of Medicine, Wroclaw Medical University, Wrocław, Poland
[3] Department of Paediatrics, Allergology and Cardiology, Wroclaw Medical University, Wrocław, Poland
[4] Department of Paediatric Cardiac Surgery, Lower Silesian Centre for Heart Diseases, Zabrze, Poland
Correspondence: Michał Puła, Thoracic Surgery Department, Lower Silesian Oncology Centre, Hirszfelda 12, 53-413 Wrocław, Poland, e-mail: michal.pula97@gmail.com

https://doi.org/10.15557/PiMR.2023.0069

ORCID iDs
1. Piotr Jan Potyrała https://orcid.org/0009-0006-6304-9778
2. Michał Puła https://orcid.org/0000-0002-3265-7424
3. Tomasz Pytrus https://orcid.org/0000-0001-7609-8479
4. Joanna Kukawczyńska-Noczyńska https://orcid.org/0000-0002-5706-6057
5. Girish Sharma https://orcid.org/0009-0002-2861-5018
6. Anna Kofla-Dłubacz https://orcid.org/0000-0002-8093-1301

Abstract

Background: Cardiac myxoma is the most common primary cancer of the heart in adults. It is characterised by non-specific clinical symptoms and radiological findings. The aim of this study is to present the case of a 3.5-year-old boy diagnosed with right atrial myxoma briefly after the onset of inflammatory bowel disease treatment. **Case report:** The patient was hospitalised because of chronic diarrhoea, dyselectrolytaemia, and severe cachexia. Following multiple examinations, a diagnosis of inflammatory bowel disease was made. Additionally, due to upper limb oedema, a cardiac consultation and echocardiography were conducted, revealing a solid mass in the right atrium. Consequently, a cardiac surgical intervention was carried out due to the patient's condition. **Conclusions:** Right atrial myxomas are extremely rare in paediatric patients, and the coexistence of inflammatory bowel disease and cardiac myxoma has not been previously reported in the literature. Further research is needed to establish the potential correlation between these two diseases.

Keywords: thrombus, right atrium, myxoma, Crohn's disease

Streszczenie

Wstęp: Śluzak serca jest najczęstszym pierwotnym nowotworem serca u dorosłych. Charakteryzuje się nieswoistymi objawami klinicznymi i radiologicznymi. Celem pracy jest przedstawienie przypadku 3,5-letniego chłopca, u którego w krótkim czasie po rozpoczęciu leczenia nieswoistego zapalenia jelit rozpoznano śluzaka prawego przedsionka. **Opis przypadku:** Pacjent hospitalizowany z powodu przewlekłej biegunki, dyselektrolitemii i ciężkiego wyniszczenia. Po przeprowadzeniu wielu badań postawiono diagnozę nieswoistego zapalenia jelit. Dodatkowo z powodu obrzęku kończyny górnej wykonano konsultację kardiologiczną oraz badanie echokardiograficzne, które ujawniło lity guz w prawym przedsionku serca. W związku z tym ze względu na stan chorego przeprowadzono interwencję kardiochirurgiczną. **Wnioski:** Śluzaki prawego przedsionka są niezwykle rzadkie u dzieci, a współwystępowanie nieswoistego zapalenia jelit i śluzaka serca u dziecka nie było dotychczas opisywane w piśmiennictwie. Konieczne są dalsze badania w celu ustalenia korelacji między tymi dwiema jednostkami chorobowymi.

Słowa kluczowe: zakrzep, prawy przedsionek, śluzak, choroba Leśniowskiego–Crohna

INTRODUCTION

The most common malignancies of the heart are metatype lesions, occurring 20 to 40 times more often than primary cardiac tumours (PTCs)[1]. PTCs are extremely rare, diagnosed in 0.0017–0.03% of autopsy studies, of which cardiac myxomas constitute the largest percentage (50–75%) among adults. However, in paediatric patients, myxomas are relatively rare, accounting for 2–4% of PTCs in infants, with considerably higher occurrence rates of rhabdomyomas, teratomas, and fibromas[2,3]. Myxomas are benign tumours composed of multipotent connective tissue cells. They are usually located in the atria, most commonly in the left atrium (75%), originating from the area of the foramen ovale and in the right atrium (20%)[4,5]. Their shape is most often similar to polypoid formations with a thin pedicle. In most cases, they are diagnosed between the ages of 30 and 60[6]. Cases in children have been reported infrequently in the literature. Only 10% of myxomas are hereditary, seen as part of Carney syndrome, while the remaining 90% are the result of *de novo* mutations[7]. In the hereditary form of myxoma of the heart, the occurrence of chromosomal aberrations involving chromosomes 2p, 12, and 17q has been reported[8]. Symptoms associated with PTC can be classified by aetiology into resulting from heart failure and thromboembolic complications. The former group includes shortness of breath, dizziness, fainting, stasis of blood flow, and oedema as well as arrhythmias. Due to the increased susceptibility to thromboembolic events, stroke, hemiplegia, convulsions, blindness, rash, and purpura are often the only initial symptoms[9,10]. Other manifestations associated with PTC include fever, anaemia, fatigue, loss of appetite, and decrease in body weight[11]. The diagnosis of PTC is typically based on the presence of the aforementioned symptoms in conjunction with findings from transthoracic echocardiography (TTE). Computed tomography (CT) and magnetic resonance imaging (MRI) can be used to verify the diagnosis[5,10]. The only method of treatment described so far is cardiac surgery. To ensure preoperative TTE accuracy, transoesophageal echocardiography (TEE) is performed intraoperatively. The procedure is conducted in extracorporeal circulation with sternotomy access. After visualising the right atrium, the operator makes an incision of its wall, followed by complete excision of the mass[1,11,12].

CASE REPORT

A 3.5-year-old patient was admitted to the Paediatric and Gastroenterology Clinic due to fatigue and chronic diarrhoea with electrolyte imbalance. Upon admission, physical examination revealed subcutaneous tissue atrophy, and body weight and body mass index significantly below the third percentile according to the World Health Organization charts, enlarged abdominal circumference, discrete peripheral oedema in the lower limbs, tachycardia on auscultation, and clear heart sounds. The dominant feature in the medical history was chronic diarrhoea of unknown onset. There was no data on the onset of growth arrest due to the patient's difficult social situation. The child's activity was limited, and there was no verbal communication. Laboratory tests revealed anaemia, thrombocytosis, significant hyponatraemia, and hypokalaemia. Inflammatory markers (erythrocyte sedimentation rate – ESR, C-reactive protein – CRP, procalcitonin) were in the normal ranges. The level of calprotectin in the stool was not elevated. Infectious causes of diarrhoea were excluded (*Salmonella*, *Shigella*, toxin-producing strains of *Clostridium difficile*, *Campylobacter jejuni*, pathogenic strains of *Escherichia coli*, viruses).

As part of the diagnostic process, abdominal CT was performed, revealing paralytic intestinal obstruction, with no changes indicative of abdominal tumour. Brain MRI showed no pathological findings. The child was referred for colonoscopy and gastroscopy. The examination of the lower gastrointestinal tract included the rectum-transverse colon segment due to limited preparation caused by the patient's severe condition. No macroscopic changes typical of inflammatory bowel disease (IBD) were visualised in the evaluated sections of the gastrointestinal tract. In the histopathological examination, inflammatory changes of significant intensity were observed, and no epithelioid granulomas or crypt abscesses were found. Simultaneously with diagnostic tests, the patient's condition was stabilised, oral feeding was stopped, and a central catheter was inserted into the right internal jugular vein to initiate parenteral nutrition. The patient was consulted with a cardiologist, and an echocardiographic examination of the heart was performed, revealing no abnormalities in the cardiovascular system. As part of the diagnostic process, autoantibody levels were assessed, and the presence of anti-*Saccharomyces cerevisiae* antibodies (ASCA) was detected at a high titre. In addition, the patient's genome was mapped, showing no pathogenic mutations, including IL-10, IL-10 receptor mutations, or mutations typical of secretory diarrhoea.

The patient's severe condition persisted despite the suspension of oral food intake, with severe diarrhoea associated with electrolyte imbalance and hypoalbuminaemia requiring constant correction. As soon as histopathological specimens taken from the colon were assessed (chronic severe inflammatory infiltrate) and high titre of ASCA antibodies was affirmed, treatment was started according to the standard approach in IBD, introducing systemic steroid therapy, azathioprine, and obtaining minor improvement. Parenteral nutrition was continued through a central catheter. The patient's treatment was interfered by severe acute respiratory syndrome-related coronavirus 2 (SARS-CoV-2) infections (four episodes within three months). In the fourth month of treatment, the patient developed asymmetrical oedema in the upper right limb. Because of the patient's worsening condition, another cardiological consultation was ordered. This time, the echocardiogram revealed a solid mass filling the right atrium and protruding through the tricuspid valve into the right ventricle in systole (Figs. 1 and 2). Considering the patient's central catheter, use of parenteral nutrition, systemic steroid therapy, autoimmune disease, and SARS-CoV-2 infections, a thrombus

DOI: 10.15557/PiMR.2023.0069

on the cannula entering the right atrium was suspected. The patient was treated with low-molecular-weight heparin. Due to the size of the lesion, the patient was deemed eligible for cardiac surgery, with the chest being opened through a sternotomy. The operation was conducted in extracorporeal circulation, through an incision of the right atrium wall. At the procedure, the entire mass, which appeared to have a narrow base attached to the atrial wall, was resected. The course of the procedure was uncomplicated. The material collected during surgical intervention was subjected to histopathological examination which confirmed the presence of a right atrium myxoma. Following the stabilisation of the patient, he was transferred to the paediatric gastroenterology ward for further treatment on the third day post-surgery. Anticoagulant therapy was continued for six months, while the patient was under cardiac monitoring (echocardiography), which did not reveal any abnormalities.

Due to persistent gastrointestinal symptoms including chronic diarrhoea, oral nutrition intolerance, and lack of weight gain despite parenteral feeding, the patient was qualified for anti-TNF-alpha therapy. Adalimumab at the dose of 5 mg/kg was administrated. Upon induction of remission with adalimumab, improvement in stool frequency and electrolyte stabilisation was observed. Parenteral nutrition was discontinued. The patient was subsequently able to switch back to oral feeding. During the 18 months of therapy, the patient gained 5 kg in weight. Follow-up colonoscopy did not reveal any macroscopic changes typical of inflammatory bowel disease (in the large intestine and the terminal ileum). Magnetic resonance enterography was not performed due to the lack of general anaesthesia necessary for the examination, considering the patient's young age.

Based on the patient's overall clinical presentation and the clinical response achieved with adalimumab, the diagnosis

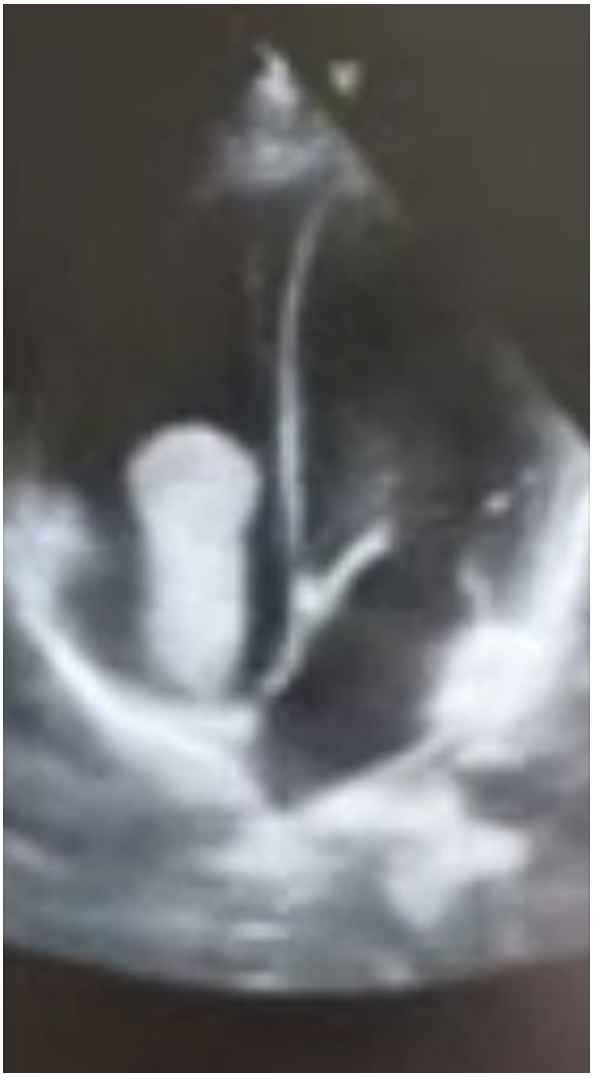

Fig. 1. Echocardiogram (subcostal view) showing a huge echogenic mass located in the right atrium during atrial constriction

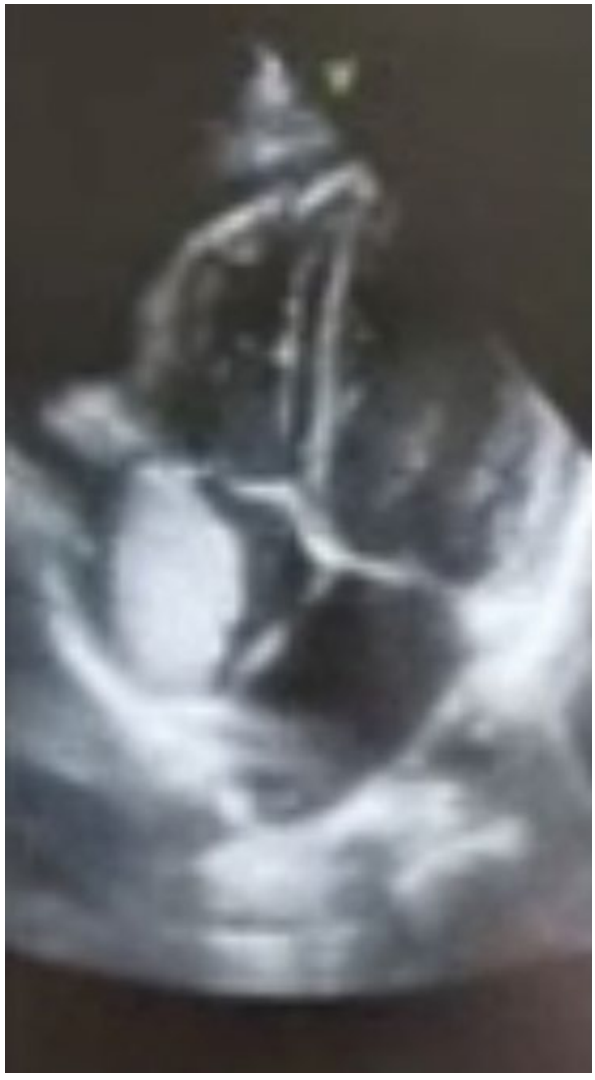

Fig. 2. Echocardiogram (subcostal view) showing a huge echogenic mass located in the right atrium during atrial diastole

 DOI: 10.15557/PiMR.2023.0069

of unspecified inflammatory bowel disease complicated by right atrial myxoma was confirmed.

DISCUSSION

The diagnosis of myxoma located in the right atrium of the heart in a paediatric patient is extremely rare. Cases of the disease described in the literature concern patients aged 30–60 years[6]. The coexistence of PTC with autoimmune diseases has been reported in the literature[13]. As of today, only one case report of concomitant PTC and Crohn's disease is found in the PubMed database[14]. A 26-year-old patient described by Parollo et al. reported to the Hospital Emergency Department presenting symptoms similar (diarrhoea and abdominal pain) to the patient described in our study. In both cases, diagnostic imaging assessment revealed pathological masses in the heart cavities, raising the suspicion of a thrombus. Both patients underwent anticoagulant therapy and open-heart surgery under general anaesthesia. The procedure was performed via a sternotomy. Currently, according to our study and the available literature, the correlation between IBD and cardiac myxomas cannot be unequivocally stated. However, the reappearance of such a correlation suggests the need to extend research in this direction. Myxoma of the heart often manifests as heart failure or the effect of embolic complications, most often affecting the cerebral vessels, but also the pulmonary, renal, coronary, and lower limb vessels[15,16]. The symptoms described in the medical literature include shortness of breath, hemiparesis, arrhythmias, fainting, nausea, weight loss, and even sudden death[1,11]. In the case of the reported patient, only loose stools, vomiting, and cachexia were recorded. The basic diagnostic tools include imaging tests such as echocardiography, both transthoracic and transoesophageal, magnetic resonance imaging, and computed tomography. The morphological similarity of this tumour to a thrombus may pose a diagnostic difficulty in radiological imaging. A factor delaying the correct diagnosis may be the presence of laboratory test abnormalities, such as elevated levels of CRP, IL-6, and growth factors, e.g. vascular endothelial growth factor (VEGF). During hospitalisation, the presented patient was diagnosed on the basis of histopathological examination, TTE, and IL-6 level. Regardless of the symptoms, the suspicion of myxoma is always an indication for surgical treatment, which is the cornerstone of therapy[1]. Treatment should be initiated immediately due to the high risk of embolic complications and cardiac blood flow disorders. In a small number of cases, the disease recurs after treatment (more commonly in patients with hereditary tumour type). Although myxoma is considered a benign tumour, cases of local malignancy and distant metastasis have been described in the literature[5,8].

Conflict of interest

The authors have no competing interests to declare relevant to this article's content.

Author contributions

Original concept of study: PJP, MP, AKD. Collection, recording and/or compilation of data: PJP, MP, AKD. Analysis and interpretation of data: PJP, MP, TP, JKN, GS, AKD. Writing of manuscript: PJP, MP, AKD. Critical review of manuscript: TP, JKN, GS. Final approval of manuscript: TP, AKD.

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DOI: 10.15557/PiMR.2023.0069