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The Mediastinal Tumor Diagnosed in 15-Year-Old Patient with Renal Colic Syndromes - A Case Report

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Abstract

Background: Urolithiasis in the pediatric population is about 10 times less common than among adults. Adolescents present with similar symptoms to adult patients, among them renal colic (acute, severe flank pain, which radiates to the groin). This case report shows an atypical course of retromediastinal tumor, which mimicked symptoms of renal colic.

Case presentation: A 15-year-old boy was admitted to Pediatric, Nephrology and Allergology Department, Military Institute of Medicine due to suspicion of urolithiasis. Patient presented left flank pain treated as renal colic. Before admission the boy was hospitalized in three centers during March 2016, July 2016 and September 2016. During those hospitalizations X-ray examination revealed a small calcification in the left kidney, however no renal calculi were detected in urography and repeated ultrasound examinations.

Discussion: Patient was treated twice with antibiotics (cefuroxime, amikacin) due to increased inflammatory parameters. Analgesics and antispasmodic drugs were administered almost continuously, without any significant effect on reported symptoms. At admission no stones were visible in ultrasound examination, therefore abdominal computed tomography (CT) was performed. No signs of urolithiasis were observed in the scan, however abnormal mass on the left-side of the chest wall was found. The study was extended to thoracic CT scan, which revealed a mediastinal tumor. Patient was referred to an oncology department where Ewing sarcoma/ primitive neuroectodermal tumor (PNET) was diagnosed in pathologic analysis.

Conclusion: Symptoms which mimic renal colic may not always be caused by urolithiasis. Even though posterior mediastinal tumors among children are significantly less common than renal stones, doctors should always consider all potential causes of symptoms that are reported by our patients. Ultrasonography remains the study of first choice, however CT should be recommended in patients with persistent symptoms of urolithiasis or nondiagnostic ultrasound examination.

Keywords: Urolithiasis; Renal colic; Renal calculi; Mediastinum tumor

Abbreviations: CT: Computed Tomography; PET-CT: Positron Emission Tomography- Computed Tomography; PNET: Primitive Neuro-ectodermal Tumor; NHL: Non-Hodgkin Lymphoma; AP: Anterior-Posterior; VIDE: Vincristine, Ifosfamide, Doxorubicin, Etoposide

Introduction

Urolithiasis is a civilisation disease that affects about 1,5% of the world population. Its incidence in the Polish population is estimated to be 2% [1]. Occurrence of urolithiasis among children is much less studied. In American records the condition is diagnosed among 1:1000-1:7600 hospitalised children [2,3]. In the pediatric population, urolithiasis is about 10 times less common than among adults, but its incidence in recent years is rising [4,5].

One of the most serious complications of urolithiasis is the complete or partial obstruction of the ureter caused by the displacement of a stone from renal calyx or pelvis. This leads to a sudden increase in hydrostatic pressure in the renal pelvis, calyces and ureter's section proximal to obstruction, dilatation of their wall and contractions of their muscular layer, ischaemia and inflammation [6].

Urolithiasis can present with renal colic: acute and severe pain in the lumbar area, which radiates to sacral bone, anterior abdominal wall, urethra and into the groin. Symptoms usually have an acute character and appear early in the morning or at night, waking the patient up. As with other visceral pain, patients exhibit significant agitation and seek a more comfortable position.

Pain may be accompanied by nausea, vomiting, hematuria, dysuria and urinary tract infection. However, in many cases, especially among younger children, urolithiasis may be olygosymptomatic and may be incidentally identified during routine ultrasound examination. Symptoms that suggest urolithiasis at a younger age include abdominal pain, dysuria, recurrent urinary tract infections, hematuria, but also diurnal and nocturnal enuresis [5].

Pain in the lumbar region may be caused by non-urinary disorders such as gastrointestinal and genital diseases, as well as due to orthopedic and neurological causes. Less common causes of pain in this region among the pediatric population include cancer, abdominal aortic aneurysm, retroperitoneal bleeding, osteopenia or osteoporosis [7,8].

Mediastinal tumors are rare among pediatric population. They represent 15-20% of all neoplasms in this age group. Considering the anatomical division of the mediastinum, neoplasms may be divided into three groups: anterior mediastinal tumors (59%), middle mediastinal tumors (29%) and posterior mediastinal tumors (16%) [9-13].

The cancers' location may indicate its origin and character. Causes of anterior mediastinum tumors include: lipoma, angioma, mature teratoma, retrosternal goitre, enlarged thymus in infants, thyroid tumor, lymphoma, thymoma [14].

Causes of middle mediastinum tumors may be as following: bronchiogenic cyst, pericardial cyst, inflammatory lymphadenopathy, hiatus hernia, Hodgkin and non-Hodgkin lymphoma (NHL), and lymph node metastases [15]. Causes of posterior mediastinum tumors include: bronchogenic or enterogenous cyst, meningeal hernia, NHL, cancers originating from the nervous (sympathetic) tissue: Neuroblastoma, ganglioneuroblastoma, ganglioneuroma, neurofibroma/ neurofibrosarcoma, carcinomas originating from bone-tissue: Ewing sarcoma, PNET (primitive neuroectodermal tumor) [16]. Clinical presentation depends on the tumors' location.

Cough, dyspnea, superior caval vein syndrome are associated with anterior and middle mediastinal tumors, whereas posterior mediastinal tumors may be asymptomatic for a long time [17,18].

Non-Hodgkin lymphoma is the most serious tumor among the above mentioned due to its fast development and rapid growth. Its symptoms are often life-threatening (dyspnea, superior caval vein syndrome, tumor lysis syndrome) [12].

Most common tumors in the posterior mediastinum are neuroblastoma and ganglioneuroblastoma, which originate from sympathetic ganglia. These tumors usually occur among younger children, may penetrate the spinal canal and cause no bone destruction. Symptoms include pain, limb paralysis, dyschezia and dysuria. Calcifications within the mass are characteristic for these tumor types [15].

Ewing sarcoma in the mediastinum originates either from bone or soft tissue of the thorax (bone and extraosseous Ewing's sarcoma) [11]. Adjacent ribs may be destructed (diagnosed in thorax CT or skeletal scintigraphy), but penetration to spinal canal is rare. Disease is more common among older children and adolescents. Symptoms include pain (often misinterpreted as spinal or renal colic pain) and chest wall bulging.

Diagnosis of Ewing sarcoma may be delayed as the disease often mimics inflammatory process. Neurological symptoms and nerve root pain may occur in case of vertebrae originating tumors that penetrate the spinal canal and infiltrate spinal nerves [19].

This case report presents the diagnostic difficulties of persistent pain in the lumbar region, which suggested renal colic in an adolescent patient.

Case Presentation

A 15-year-old boy was admitted to the Pediatrics, Nephrology and Allergology Department of the Military Institute of Medicine in Warsaw due to lumbar pain, which suggested renal colic. Additionally, slight dilatation of renal calyceal-pelvic complex was observed.

In March 2016 patient was hospitalized in a district hospital due to pain in the left lumbar region. Minor dilatation of the left renal calyceal-pelvic complex was observed, no renal calculi in the urinary tract were found.

Urinary tract infection was ruled out. Considering the clinical picture and positive family history of urolithiasis (father), renal colic was diagnosed. Symptomatic treatment resulted in a slight reduction of pain, with no obvious improvement of the child's well-being.

Symptoms persisted and patient was hospitalized twice in an urgent manner. In August 2016, ultrasound examination in a regional hospital revealed a similar dilatation of the left renal calyceal-pelvic complex.

Therefore, urography was performed, which showed no calcifications in the urinary tract. After contrast administration dilatation of the left renal calyceal-pelvic complex was observed (pelvis 24 mm, calyx 12 mm in the anterior posterior projection- AP). The renal pelvis was modelled on the psoas major muscle.

In September 2016 patient was hospitalized in a clinical hospital due to left lumbar pain, fever and dysuria. Clinical examination revealed tenderness on palpation beneath the ribcage on the left side, accompanied by a positive Goldflam sign on the left side. Ultrasound examination was similar to previous findings, no obvious urinary tract deposits were found. Abdominal X-ray revealed a 5 mm shadow at the S1 vertebral body level. Due to clinical symptoms and markedly elevated inflammatory markers (CRP 14 mg/dl), urinary tract infection was diagnosed. Parenteral antibiotic therapy (cefuroxime) and symptomatic treatment was administered. After two days no improvement was observed, inflammatory markers remained elevated and pain in the lumbar region persisted. Therefore, patient was transferred to a pediatric surgery clinic specializing in endoscopic treatment of urolithiasis.

In the surgery department, constant pain the left lumbar region was observed, whereas dysuric symptoms resolved. Urine test and culture revealed no signs of urinary tract inflammation, yet inflammatory markers remained elevated.

Therefore, empirical antibiotics therapy was extended (amikacin was started) and pain gradually diminished. As no obvious deposits were found in the urinary tract (despite repeated imaging) and due to signs of ongoing infection, no endoscopic procedure was performed. Further diagnostics (including CT-scan) was planned in a pediatric nephrology clinical hospital.

Patient was admitted to the Pediatrics, Nephrology and Allergology Department of the Military Institute of Medicine in Warsaw. Ultrasound examination revealed no calculi in the urinary tract. CT urography scan was performed and revealed a heterogenous mass on the left side of the thorax. The decision to extend the scan was immediately made.

Thorax CT showed the mass of approximately 80 × 60 × 60 mm in the posterior mediastinum at the level of 7^{th}-10^{th} rib (**Figures 1-3**). Tumor infiltrated the chest wall and caused substantial osteolysis of the 9th rib. Furthermore, periosteal reaction of the 8^{th} and 10^{th} rib and no destruction of the vertebrae body were found. No visible calcifications were observed. Peripheral wide network of pathologic vessels within the tumor, atelectasis, and a trace of pleural effusion were observed.

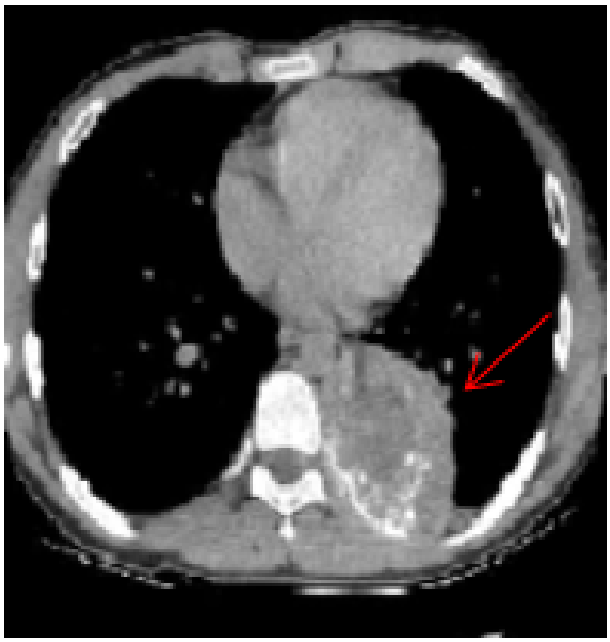

Figure 1 Chest CT: The mass in the posterior mediastinum (arrow).

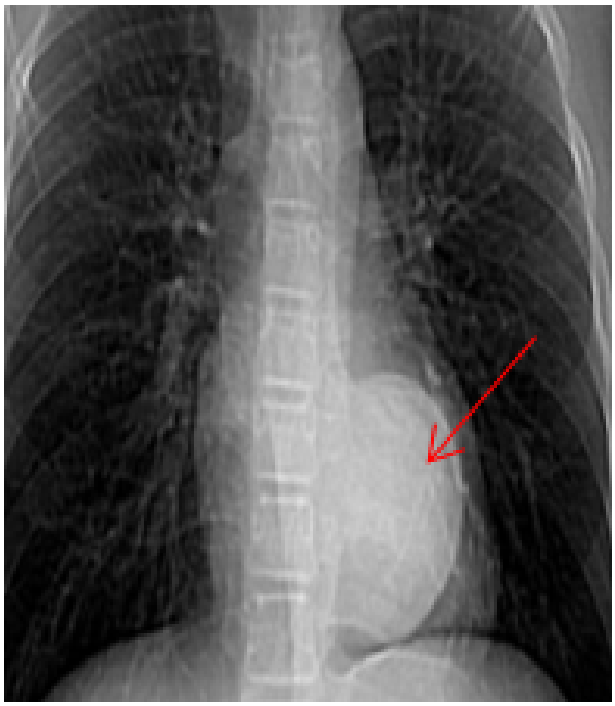

Figure 2 Chest CT: Tumor projection over the cardiac silhouette (arrow).

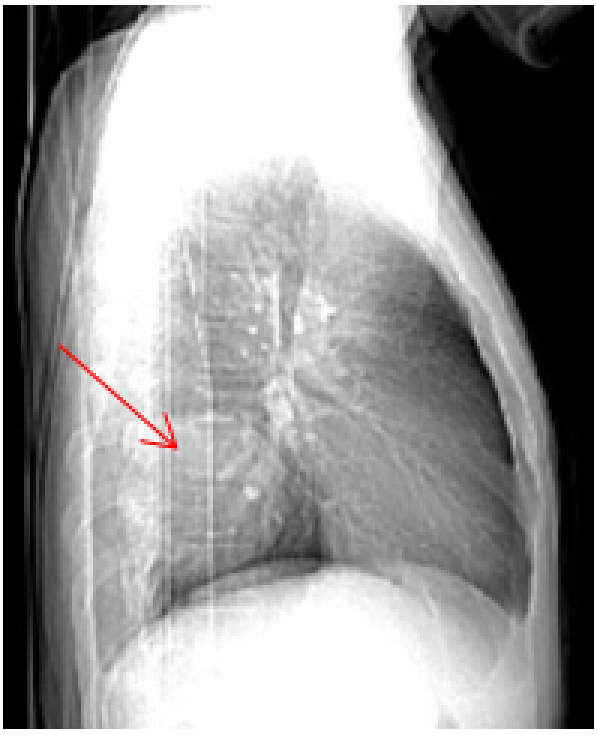

Figure 3 Chest CT: Tumor in the posterior mediastinum (arrow).

Patient was transferred to an oncology centre for further diagnosis and treatment. Clinical examination in the Oncology Clinic revealed a slight chest asymmetry – inferior angle of the

left scapula was slightly higher. Additionally, bulging of the chest wall was observed on a distance of 7cm in the paravertebral line on the left side, which corresponded to the findings of the CT scan. In the tumor biopsy histopathological features of the Ewing/PNET sarcoma were identified. No pathological changes in the aspiration and bone marrow biopsy were found. PET-CT confirmed the presence of a metabolically active tumor, which caused the destruction of the 9th rib, with no signs of metastases. Patient was treated with the VIDE chemotherapy protocol (vincristine, ifosfamide, doxorubicin, etoposide), with good tolerance. Partial regression of the tumor was observed in the thorax CT scan and the renal colic symptoms resolved. Patients' treatment is continued.

Discussion

Diagnosis of urolithiasis is based on clinical examination and the visualization of urinary tract deposits in imaging studies. Ultrasonography is characterized by high sensitivity and specificity in urolithiasis diagnosis [9], therefore it is the method of first choice. Calculi in the kidney and ureteropelvic junction are usually visible in ultrasound examination. Deposits in the ureter may also be seen after appropriate patient preparation and with careful examination. Calculi are seen as hyperechogenic structures with posterior acoustic artifacts: twinkle artifact in color Doppler ultrasound and acoustic shadow upon larger deposits (signal void behind the calculi) [7]. At times, the only indirect signs of urolithiasis may be the dilatation of the renal calyceal-pelvic complex and of the ureter proximally to the deposit.

Abdominal X-ray or urography may be also a valuable method, especially due to their widespread availability. However, they are additional tests, and they are more frequently replaced by non-contrast CT scanning, which in many centres has become a method of choice for diagnosing urolithiasis. The sensitivity and specificity of CT is 100%, furthermore CT gives the opportunity to differentiate the chemical composition of the deposit [4]. New protocols with reduced radiation dose (3.7 mSv vs. 0.5-0.7 mSv) may be used for testing among pediatric population [7].

It is estimated that around 15% of lumbosacral pain is caused by inflammatory, infectious or neoplastic disease. Red flag symptoms include patients' age below 20 years old, fever not associated with infection, weight loss, and pain intensification at night, progressive fatigue, dysuria, dyschezia and abnormal gait [8].

The most common cause of colic pain is urolithiasis. Renal colic presents with characteristic symptoms, which often suggests the diagnosis. Quite often patients with diagnosis of urolithiasis are referred to the Pediatric Nephrology Department. However, the diagnosis is often based solely on patients' medical history and physical examination, with no visualization of any calculi in diagnostic imaging.

This was also the case in the presented report, where ultrasound examination was performed several times at different centres and none confirmed urolithiasis. The small calcification seen in the projection of the left kidney during abdominal X-ray was not confirmed in the urography. Urolithiasis diagnosis was not supported by imaging studies and no calculi were excreted during the entire observation period. The diagnosis was based on indirect signs: dilatation of the renal calyceal-pelvic complex and urinary tract infections. Careful examination of patient's medical history revealed that patients' pain was not typical (persistent, not responding to analgesic and antispasmodic drugs). Furthermore, asymmetry of the thoracic-lumbar spinal region was observed, which at first assessment suggested a postural defect.

Conclusion

Because of the untypical nature of pain, diagnostic imaging should have been considered earlier. Therefore, CT scan should be recommended in patients with persistent symptoms of urolithiasis or non-diagnostic ultrasound examination. In this case, the posterior mediastinal tumor was diagnosed incidentally during an abdominal CT scan. Tumor's low localization reaching 11th rib caused episodes of renal colic and persistent dilatation of the renal-calyceal complex. Patient was referred to an oncological center where Ewing/PNET sarcoma with no metastases was diagnosed. Ewing/PNET sarcoma occurs in 90% of cases at an age younger than 20 years, usually about 13 years. Tumors are more common among boys, as observed in this case. Disease progression at time of diagnosis is advanced among the majority of patients (presence of metastases), therefore prognosis is poor-5-year survival is about 40%. However, 5 years survival for localized disease (with no metastases) is 60-70% [19]. No metastases were diagnosed in the described case report, which may improve the patients' prognosis.

Lumbar pain remains one of the common reasons for referrals to the Emergency Room. In questionable cases, where the cause of pain cannot be diagnosed despite repeated ultrasound examinations, diagnostic imaging should be extended. It is vital that pediatricians and family physicians take into account all potential causes of symptoms. Such practice may result in an earlier diagnosis and treatment, which may improve the overall prognosis.

Author's Contributions

Marianna Lichosik took the lead in writing the manuscript with the support from JK. KJ supervised the nephrological part of the paper. Aneta Czajńska-Deptuła wrote the oncological part of the manuscript with supervision from Bożenna Dembowska-Bagińska. Aleksandra Paturej translated article into English. Joanna Kacik and Aleksandra Paturej edited the article according to the guidelines. Bolesław Kalicki made critical revision of the article and gave final approval. All authors read and approved the final manuscript.

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