

CLINICAL CASE

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Splenic Giant Epithelial Cyst – the Rare Reason of Proteinuria

Olbrzymia cysta śledziony – rzadka przyczyna białkomoczu

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Abstract

The case of a 12-year-old girl suffering from splenic giant epithelial cyst and proteinuria is presented. The authors would like to underline the extremely rare coincidence of a childhood splenic giant epithelial cyst and proteinuria resulting from left renal vein compression by the cyst and its disappearance after elective cystectomy (*Adv Clin Exp Med* 2005, 14, 3, 621–624).

Key words: children, splenic cyst, proteinuria.

Streszczenie

Opisano przypadek 12-letniej dziewczynki z olbrzymią cystą śledziony i białkomoczem. Autorzy chcieli zwrócić uwagę na wyjątkowo rzadkie występowanie białkomoczu spowodowanego uciskiem żyły nerkowej przez olbrzymią cystę śledziony oraz całkowite jego ustąpienie po chirurgicznym usunięciu cysty (*Adv Clin Exp Med* 2005, 14, 3, 621–624).

Słowa kluczowe: dzieci, cysta śledziony, białkomocz.

Splenic cysts are a relatively rare entities, but the increasing use of ultrasound makes these diagnoses more frequent. The traditional treatment of splenic cysts has been splenectomy. It is now well established that the spleen is involved in several functions including regulation of circulating blood volume, haemopoiesis, immunity and protection against infections and malignancies. The recognition that splenectomy has the potential for short and long-term complications, especially in children, has led to emphasis on conservation of splenic tissue [1, 2]. Elective splenectomies are currently performed only in children with absolute indications [2–4]. Due to the increased risk of severe infection, children qualified for the splenectomy, regardless of the cause, are scheduled for certain vaccinations in the pre-operative period [5]. Reports on proteinuria caused by compression of kidney vessels are relatively infrequent, however, to

our best knowledge there is only one previous report describing the splenic cyst causing compression of kidney vessels manifested by proteinuria.

The aim of our report is to point out the extremely rare coincidence of splenic giant epithelial cyst and proteinuria caused by constriction of left renal vein by this cyst in children.

Case Report

A 12-year-old girl was admitted to the Pediatric Department due to proteinuria and giant tumor in the left upper quadrant of the abdominal cavity, which had been accidentally found on routine physical examination. The history revealed periodic left upper quadrant discomfort or even pain, occurring for last 3–4 years. Parents denied abdominal injury in a child. The palpable tumor in the central

abdomen and its left upper quadrant extending 10 cm below the costal margin was found on physical examination. With the exception of decreased platelet count (100–132 G/l) other laboratory tests: ESR, RBC, WBC, reticulocyte count, liver tests, BUN and blood ions were normal. Urinalysis revealed proteinuria (120–250 mg/dl); 24-hour proteinuria was 11–34 mg/kg/24 h (nighttime and daytime proteinuria was comparable), whereas serum protein and cholesterol levels were normal. Abdominal ultrasound revealed the presence of giant cyst (16.3 × 14.7 × 13.6 cm) that compressed renal vessels causing distension of the left renal vein (LRV) and pushed down the left kidney (Fig 1). Abdominal CT scan revealed the single splenic cyst measuring 11 × 17 cm in transverse section and 13 cm in length (Fig. 2). The pediatric surgeon initially scheduled the girl for the partial splenectomy; however, due to the size of the cyst, total

splenectomy could not be excluded at that moment. The child was vaccinated with the following vaccines: Act-HIB, Pneumo 23 and meningococcal polysaccharide vaccine A+C. One week later the girl underwent partial splenectomy (Fig. 3). Histopathological specimens showed features typical of congenital epidermoid (epithelial) cysts with no signs of atypia. Following surgery, proteinuria and thrombocytopenia subsided on the second and seventh day, respectively. Control abdominal ultrasound did not show distended LRV seen on previous examination.

Serum levels of CA125 and CA19-9 were within normal range six months after the surgery. However, this parameters had not been determined before the surgery.

Discussion

Splenic cysts are classified as true (primary) cysts and pseudocysts (secondary) irrespectively of whether they are lined with epithelium or not [3, 6]. Posttraumatic pseudocysts result from nonresorption of haematoma or infarction of the spleen, represent 75% of all splenic cysts. Epithelial cysts are thought to arise from mesothelial inclusions of the visceral peritoneum during fetal spleen development and undergo squamous metaplasia later in life [3]. Differential diagnosis includes neoplasms and abscesses of the spleen. Congenital cysts eventually become symptomatic as space-occupying lesions and are manifested by non-specific complains or as a palpable abdominal mass [3]. This picture is consistent with our observations. In the presented case, which is the only one seen in our Department, the girl complained of occasional, moderate pain in the left upper quadrant. The ab-

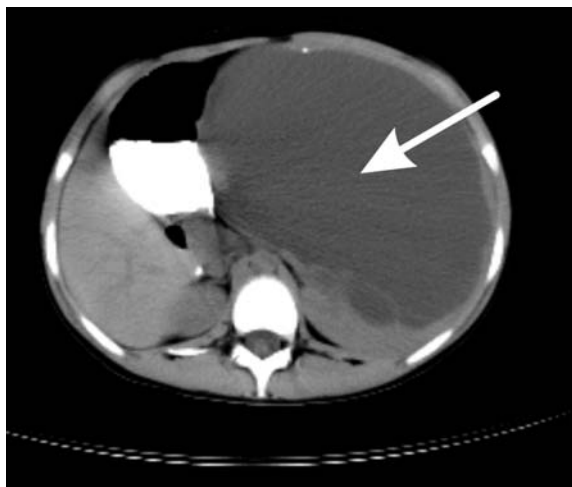


Fig. 1. CT scan. Giant cyst of the spleen (arrow)

Ryc. 2. TK. Olbrzymia cysta śledziony (strzałka)

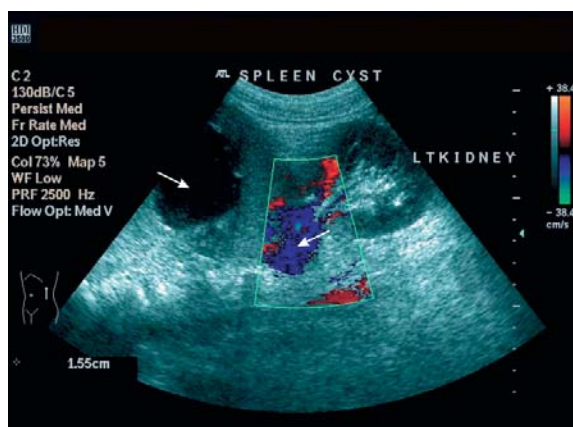


Fig. 2. USG. Giant cyst of the spleen (left arrow) with distended left renal vein (right arrow)

Ryc. 2. USG. Olbrzymia cysta śledziony (lewa strzałka) z poszerzoną lewą żyłą nerkową (prawa strzałka)



Fig. 3. Intraoperative view. Giant cyst of the spleen (arrows)

Ryc. 3. Widok śródoperacyjny. Olbrzymia cysta śledziony (strzałki)

dominal tumor was diagnosed accidentally on routine physical examination. In addition, the girl was presented with permanent, moderate proteinuria (maximal 34 mg/kg/24 h) caused by cyst compression on LRV. Total subsidence of proteinuria was seen on the second day following surgery.

We believe this is the second report on the splenic cyst causing compression of LRV and subsequent proteinuria. A similar case was reported in 2002 by Faizan et al. [10]. Cases of proteinuria caused by compression of kidney vessels are rarely described in the literature. Proteinuria of variable degree has been observed in some patients with nutcracker phenomenon (syndrome). The nutcracker phenomenon (entrapment of the LRV) refers to the compression of the LRV between the aorta and superior mesenteric artery, resulting in elevation of pressure in the LRV and usually leading to gross haematuria and orthostatic proteinuria [7, 8]. Experimental partial obstruction of the renal vein in rats can also lead to proteinuria mediated by angiotensin II [9].

Due to increased risk of complication e.g. potential splenic infection, rupture following even trivial trauma, haemorrhage or abscess formation elective surgical removal of the cysts is the recommended treatment [1–4, 6]. The technique of the operation must be individualised to the etiology encountered. An effort is made in each instance to preserve a minimum of one third of the normal amount of functional splenic tissue [1]. In our patient the single-cavity cyst containing approximately 2 liters of fluid was excised in total without extensive resection of the spleen.

The distinct decrease in platelet count due to the hypersplenism, before the surgery, has been noted in many patients with huge splenic cysts. In the presented case the platelet count was also decreased. According to observations reported by Brown et al. and Kimber et al., almost every patient treated with partial splenectomy had significant elevation of their early postoperative platelet count [1, 2]. This is also consistent with our observations. In our patient the recovery of the platelet count was seen on the 7th day following surgery.

The giant epithelial cyst of spleen can be associated with elevated levels of serum and intracystic tumor markers, CA125 and CA19-9. After the cyst removal both of the markers return to normal levels [11]. In the present case, serum levels of CA125 and CA19-9 were within normal range six months after the surgery (the preoperative levels had not been measured).

Because of the increased risk of infections, routine management should include vaccination before elective splenectomy [5]. The dimension of the cyst in our patient did not allow to plan the extent of the elective surgery before the operation (we considered even total splenectomy) thus the girl, before the surgery was vaccinated.

The girl is currently under supervision of the Outpatient Department of Hematology for one year. No abnormalities in laboratory tests are seen. The authors would like to underline the extremely rare coincidence of a childhood splenic giant epithelial cyst and proteinuria resulting from left renal vein compression by the cyst and its disappearance after elective cystectomy.

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