Accessory spleen is present in approximately 10% of the general population [1]. It is usually small in size (< 6 cm) and located near the splenic hilum, the tail of the pancreas, or the greater omentum [1]. In atypical cases, the diagnosis is difficult preoperatively. Pelvic accessory spleen is rare and only a few cases have been reported [2, 3]. Here, we describe a large pelvic accessory spleen in a teenage girl and discuss the differential diagnosis of this adnexal solid mass.

**Case Report**

A 17-year-old girl, gravida 0, visited our hospital on February 5, 1999, because of intermittent dull pain in the lower abdomen for 1 month. Her menarche had occurred at 13 years old. She had a regular menstrual cycle and had her last menstrual period on January 25, 1999. There was no history of dysmenorrhea or hypermenorrhea. However, intermittent and dull pain in the lower abdomen had developed 1 month prior to her visit and was refractory to medical treatment from local clinics. There was no recent body weight loss, poor appetite, rectal tenesmus, or urinary frequency.

At admission, the patient’s vital signs were stable. Physical examination showed mild tenderness over the lower abdomen. Vaginal examination was not performed because of no coital experience. Abdominal ultrasound showed a large, well-defined, heteroechoic, solid mass in the right side of the pelvis (Fig. 1). No ascites was found. Color Doppler ultrasound demonstrated prominent blood flow in the mass with low resistance index (RI; 0.49–0.39) and pulsatility index (0.70–0.48) (Fig. 2). The left ovary was not remarkable. Therefore, a right adnexal tumor was diagnosed, and malignancy with intermittent torsion was suspected. Laboratory evaluations demonstrated normal white blood cell count and hemoglobin levels but a low platelet count (136,000/µL). Tumor markers including CA-125, carcinoembryonic antigen, human chorionic gonadotropin, and alpha-fetoprotein were all within normal limits.

Laparotomy was performed on February 9, 1999, because of the progressing abdominal pain. The operative findings were as follows: a wandering congestive mass (13 x 8 x 7 cm) with a tortuous vascular pedicle that originated along the dorsal aspect of the greater omentum situated in the cul-de-sac; unremarkable uterus and ovaries; and a normal-sized spleen palpated in the left upper quadrant by an experienced surgeon. Pelvic accessory spleen was diagnosed and the mass...
was completely excised from the vascular pedicle. The mass weighed 174 g and the histopathologic study confirmed an accessory spleen with congestive splenomegaly. The patient recovered well and was discharged on the third postoperative day. The follow-up platelet count at 9 months had increased to 374,000/µL.

**Discussion**

Accessory spleen is the result of failure of fusion of separate masses originating from the left side of the dorsal mesogastrium. Accessory spleens may cause clinical symptoms by compression of the adjacent organs, rupture, torsion, or infarction [4, 5]. Sonography and computerized tomography (CT) scan of an accessory spleen often reveal a small round or ovoid hypoechoic mass with similar echogenicity below the splenic hilum [6]. An accessory spleen may have homogenous appearance, a normal vascular branching pattern, low resistance flow, and similar RI to that of the native spleen [3]. In typical cases, accessory spleen can be diagnosed sonographically by demonstrating blood supplies from the splenic vessels [7]. Scintigraphy, splenic angiography, or magnetic resonance (MR) imaging may be helpful in the diagnosis of atypical cases [8]. The presence of an accessory spleen is generally asymptomatic and of no clinical importance. Surgical removal can be performed in patients with acute abdomen caused by torsion or rupture of the accessory spleen.

Accessory spleen plays an important role in thrombocytopenia, especially in idiopathic thrombocytopenic purpura (ITP). ITP is an immune disorder mediated by autoantibodies to platelet membrane antigens, in which platelet destruction by the reticuloendothelial system occurs, largely in the spleen [9]. Patients with chronic ITP should be investigated for the presence of accessory spleen. This abnormality may be found in about 10% of patients with recurrent ITP after primary splenectomy and is suggested by the absence of Howell-Jolly bodies in the peripheral blood smear [9, 10]. Facon et al reported that accessory splenectomy was beneficial in cases of severe thrombocytopenia after splenectomy and might be delayed until worsening of the clinical course in patients with moderate thrombocytopenia with a small accessory spleen [10]. In our patient, accessory splenectomy resulted in complete remission of thrombocytopenia and lower abdominal pain.

Why the accessory spleen of our patient became so large is unclear. Holloway et al described a case of portal hypertension resulting in massive enlargement of an accessory spleen [11]. Marked engorgement of the vessels may result from intermittent torsion or compression of the long vascular pedicle, which may lead to venous congestion and then enlargement of the accessory spleen. This phenomenon may explain how accessory splenomegaly and lower abdominal pain developed in our patient.

Most solid pelvic masses in women are of uterine or ovarian origin. Rarely, solid pelvic tumors are caused by conditions including metastasis, tubal carcinoma, lymphadenopathy, or ectopic pelvic kidney. Ovarian echogenic cysts and uterine leiomyomas can be differentiated from other ovarian solid tumors because of the presence of main blood flows within these tumors, instead of at the periphery [12]. Fast-growing tumors contain many newly formed vessels and often have low resistant blood flow. Timor-Tritsch et al reported that the combination of a high morphologic score and a low RI on color Doppler sonography predicted all of a series of 14 ovarian malignancies. It is worth noting that benign ovarian solid masses such as fibromas and thecomas in their study had high scores but high RIs [13].

Generally, the diagnosis of ovarian cancer requires an exploratory laparotomy; abdominal and pelvic CT or MR imaging scans are of no value for patients with a definite
pelvic mass [14]. About one-third of ovarian tumors in children and adolescents are malignancies, including germ cell tumors, sex cord-stromal tumors, and epithelial carcinomas [15], with the CA-125 concentration elevated in only 30% to 50% of stage I ovarian carcinomas [12]. Besides, some germ cell tumors such as immature teratomas and pure germinomas do not secrete tumor markers [14]. Therefore, ovarian malignancy, such as germ cell tumor or sex cord-stromal tumor, was suspected preoperatively in our patient.

In conclusion, an accessory spleen should be included in the differential diagnosis of a solid adnexal tumor with low resistant blood flow, especially in patients with thrombocytopenia.

References