Hypersplenism Caused by an Accessory Spleen Mimicking an Intra-Abdominal Neoplasm: Report of a Case

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Abstract
Accessory spleens are usually asymptomatic, although they may cause hematological disorders associated with hypersplenism, usually after splenectomy. Moreover, cases of hypersplenism occurring secondary to enlargement of an accessory spleen with an unaltered normal spleen have been reported. An accessory spleen can also mimic an intra-abdominal neoplasm. We report a case of hypersplenism that occurred secondary to an increase in the size of the accessory spleen, which was located in the mesentery close to the cecum, mimicking recurrence of previously resected renal carcinoma.

Key words Accessory spleen · Intra-abdominal mass · Intra-abdominal neoplasm · Hypersplenism · Hemolytic anemia · Thrombocytopenia · Renal carcinoma

Case Report
A 75-year-old woman, whose medical history was notable only for asthma and osteoporosis, presented to our Emergency Service, pale and sweaty in appearance. Physical examination revealed no remarkable findings. Laboratory data revealed hemoglobin 7g/dl with 5.4% reticulocytes and a platelet count of 2000/mm3. The serum tumor markers were normal and serological tests for hepatitis B and C as well as human immunodeficiency virus were negative. A peripheral blood smear showed a leukoerythroblastic process without dysplasia or blasts, and a bone marrow tap revealed no remarkable findings. Coombs test was positive. Computed tomography (CT) showed a right renal mass, 13 cm in maximum diameter, with a 3-cm mesenteric solid node. After corticoid therapy and two transfusions of packed red blood cells, the patient recovered from her bicytopenia and the Urology Department performed a right radical nephrectomy. Pathological examination of the mass revealed a chromophobe cell renal carcinoma (Fuhrman grade 2, limited to the kidney). The patient recovered uneventfully, and her hemoglobin and platelet counts 7 days after surgery were normal. She did not require further corticoid therapy. A diagnosis of autoimmune hemolytic anemia and thrombocytopenia caused by a paraneoplastic syndrome secondary to chromophobe cell renal carcinoma was made.

A control CT scan done 9 months later showed a 5-cm mesenteric mass, with a normal spleen size (Figs. 1 and 2). Fine-needle aspiration cytology (FNAC) of the lesion was inconclusive. A repeat CT scan done 2 months later showed no evidence of mass growth. Another 2 months later the patient presented again to the hospital with a hemoglobin level of 6.6 g/dl and a total serum bilirubin of 4.77 mg/dl, which was mainly indirect bilirubin. A Coombs test at this time was negative. A CT scan showed no growth and FNAC was again inconclusive. We decided to perform an exploratory laparotomy because of the chance that the mass was a recurrence of the renal tumor. Intraoperatively, a 5-cm mass was found in the mesentery close to the cecum. It was well circumscribed, encapsulated, and with nutrient vessels of a considerable size (Fig. 3). Bonds were placed around the nutrient vessels and these were cut; then the mass was excised, without any need for bowel resection. The spleen was normal in size and features. Pathological examination revealed that the mass was an accessory spleen. The patient recovered uneventfully and was discharged 7 days after surgery, with normal hemoglobin levels and no increase in the platelet count.

Retrospectively, the 3-cm mesenteric node described in the first CT scan report was in the same location as
the mesenteric mass and corresponded with the accessory spleen that later increased in size. Finally, this episode was diagnosed as hemolytic anemia caused by hypersplenism secondary to accessory spleen enlargement. At follow-up 6 months after surgery, the patient was asymptomatic with normal hemoglobin and platelet levels, and not receiving any treatment.

Discussion

An accessory or ectopic spleen is present in an estimated 25%–40% of the general population. Its main locations are the splenic hilum, around the splenic artery, splenocolic ligament, inside the pancreas, major omentum, mesenterium, adnexial region, or scrotum, but always in the left side of the abdominal cavity.1 Accessory spleens are usually asymptomatic, but hematological disorders can be caused by associated hypersplenism, usually after splenectomy,2 although there have been cases of patients with hypersplenism secondary to accessory spleen enlargement with an unaltered normal spleen. Once the accessory spleen is resected, the hypersplenism disappears.3,4

In some patients, the accessory spleen may cause abdominal pain due to cysts or abscesses formation, spontaneous rupture, or infarction caused by vascular pedicle torsion, clinically evident as acute abdomen.5 Cases of accessory spleens mimicking neoplasms of other organs, such as the liver, pancreas, kidney, ovarium, stomach, or retroperitoneum, have been described.6,7

Our patient probably suffered two different episodes of hemolytic anemia: the first was probably autoimmune hemolytic anemia and thrombocytopenia, possibly related to the renal cell tumor (Coombs test positive); and the second was a hemolytic process secondary to an accessory splenic sequestration with enlargement of the intra-abdominal mass, corresponding with the accessory spleen (Coombs test negative).

References