

Splenic pseudocyst: a rare association with splenoptosis and vertebral segmentation anomalies

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ABSTRACT

Splenoptosis (wandering or ectopic spleen) is a congenital fusion anomaly of the dorsal mesogastrium in which the spleen is abnormally mobile due to its attachment by a long vascular pedicle. This abnormal mobility predisposes the spleen to complications such as torsion, infarction, gangrene and pancreatic necrosis. Pseudocyst formation is one of the rarely reported complications of splenoptosis. Few cases of splenoptosis associated with vertebral segmentation anomalies have been reported in the past. Here, we present the case of a young man with kyphoscoliosis, vertebral segmentation anomalies and splenoptosis complicated by pseudocyst formation.

Keywords: kyphoscoliosis, pseudocyst, spleen, vertebra

Singapore Med J 2011;52(7):e141-e142

INTRODUCTION

Splenoptosis, a congenital fusion anomaly of the dorsal mesogastrium, is a rare entity with a reported incidence of 0.2%.⁽¹⁾ Pseudocyst formation is an unusual complication of a wandering spleen. This case highlights the association of wandering spleen, splenic pseudocyst and vertebral segmentation anomalies.

CASE REPORT

A 21-year-old man presented with intermittent abdominal pain of three months' duration. On physical examination, there was gross kyphoscoliosis and a large mobile lump in the epigastrum and umbilical region (Fig. 1). Routine laboratory tests were unremarkable. Computed tomography (CT) examination revealed an enlarged ectopic spleen with a well-defined, thin-walled, non-enhancing, high-density cystic lesion measuring 15 cm × 10.4 cm × 15.2 cm (Fig. 2). Skeletal anomalies, including scoliosis of the thoracolumbosacral spine, multiple vertebral segmentation anomalies (including spina bifida



Fig. 1 Photograph shows kyphoscoliosis with fullness in the epigastrum and umbilical region in our patient.

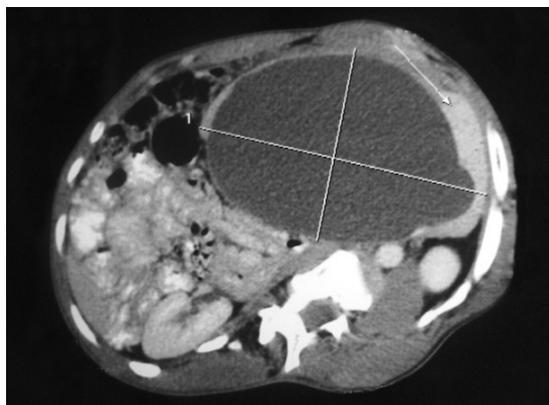


Fig. 2 Contrast-enhanced CT image shows an ectopic spleen with a pseudocyst displacing the bowel loops. Note the vertebral anomaly.

at upper sacral level) and agenesis of the lower sacrum and coccyx, were observed.

At laparotomy, the spleen was found to be enlarged and freely mobile, with a cystic lesion compressing the splenic tissue. Splenic vascular pedicles were abnormally long, and the hilar vessels were splayed out. Ligamentous attachments of the spleen to diaphragm were lax. We proceeded with splenectomy. Histopathology was suggestive of a splenic pseudocyst. Culture of the cystic fluid was sterile. The postoperative period was uneventful, and the patient was administered pneumococcal vaccination.

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DISCUSSION

Pseudocysts of the spleen lack an epithelial lining, and the cyst wall is composed of fibrous tissue. They result from trauma or infarction of the spleen and comprise around 80% of non-parasitic splenic cysts.^(2,3) Splenoptosis (wandering or ectopic spleen) results from an absence or abnormal laxity of splenic supports such as the gastrosplenic and lienorenal ligaments. This causes a long splenic vascular pedicle and a myriad of complications like torsion, splenic abscess and pancreatic tail necrosis.⁽³⁾ It has a bimodal age distribution, with children less than ten years of age and women aged 20–40 years being the most commonly affected.⁽⁴⁾ A wandering spleen can be asymptomatic, or it can present as acute abdomen or an abdominal lump.

Few cases of wandering spleen in association with split notochord syndrome have been described previously in infants.⁽⁵⁾ One case has been reported in association with Marfanoid joint hypermobility.⁽¹⁾ In our patient, a large pseudocyst was present in association with kyphoscoliosis and vertebral segmentation anomalies, which is an unexplained feature. In our review of literature, we could find only a few reported cases of traumatic pseudocysts in a wandering spleen.⁽⁶⁾ Our patient denied any history of trauma with regard to the associated skeletal anomaly. This is the first case reported in an adolescent.

Ultrasonography is a useful screening tool, but CT remains the investigation of choice in suspected cases.^(3,4,7) Mobility of the spleen has to be demonstrated

by examining the patient in different positions in order to confirm a wandering spleen.⁽³⁾ Expectant management is unwise, and splenopexy is now regarded as the treatment of choice in patients with a wandering spleen so as to prevent any future complications.^(4,7) Indications for splenectomy include an infarcted spleen and huge splenomegaly precluding splenopexy.⁽⁴⁾

In conclusion, splenoptosis poses a diagnostic challenge to clinicians due to its rarity and paucity of symptoms. To the best of our knowledge, this is the first case that reports the association of wandering spleen, splenic pseudocyst and vertebral segmentation anomalies.

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