Left Diaphragmatic Eventration with a Suprapubic Spleen: Report of a Case

Simmi K. Ratan¹, Shabnam Bhandari Grover², Rajiv Kulsreshtha¹, Vandana Puri², and Archana Puri¹

¹Department of Paediatric Surgery and ²Department of Radiology, Safdarjung Hospital, West Kidwai Nagar, New Delhi, India

Abstract Diaphragmatic defects such as eventration and hernia are known to be associated with a high-sited, sometimes intrathoracic spleen. We report here a unique case of an 8-year-old boy found to have a left congenital diaphragmatic eventration and a suprapubic wandering spleen after presenting with symptoms of an “acute abdomen” due to torsion of the splenic pedicle. To our knowledge only one other case of a similar paradoxical association of these anomalies has been reported before; interestingly, in this patient splenic infarction had also resulted secondary to splenic torsion. We recommend careful evaluation of the location and vascular status of the spleen in all patients with congenital diaphragmatic defects due to the common occurrence of splenic vascular insult.

Key words Diaphragmatic eventration · Wandering spleen · Splenic torsion

Introduction

The usual location of the spleen is in the left hypochondrium just under the left leaf of the diaphragm. Thus, in patients with left diaphragmatic defects such as eventration and hernia, a high-sited spleen and even an intrathoracic spleen may be observed.¹ There are case reports describing patients with diaphragmatic defects who presented with torsion and infarction of a herniated intrathoracic spleen.²,³ Therefore, the occurrence of a low-lying spleen with a diaphragmatic defect sounds paradoxical. We recently observed this association in an 8-year-old boy with left diaphragmatic eventration. This case is presented because of its rarity: A review of the literature revealed it to be only the second such case reported.

Case Report

An 8-year-old boy presented with acute pain in the hypogastrium, high-grade pyrexia, painful and frequent micturition, and a tender suprapubic lump of 2 weeks’ duration. There was no history of pyuria, hematuria, or local trauma, although he reported having experienced episodes of intermittent suprapubic pain during the past year.

The general physical examination revealed a febrile child with a temperature of 38.5°C. On local examination there was a tender, firm, fixed, 8 × 5 cm mass, with palpable lower limits in the suprapubic region. The overlying skin was normal. Initially, the clinical diagnosis of an inflamed urachal cyst was considered.

The laboratory parameters were hemoglobin 10.2 g/dl, total leukocyte count 11 000/mm³, polymorphonuclear leukocytes 80%, lymphocytes 18%, eosinophils 1%, and monocytes 1%. Routine and microscopic examination of the urine proved normal. A chest radiograph revealed left diaphragmatic eventration (Fig. 1). Ultrasonography demonstrated that the spleen was absent from the splenic fossa, and that the echotexture of the mass in the suprapubic region was normal. Initially, the clinical diagnosis of an inflamed urachal cyst was considered.

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A laparotomy was performed through a left subcostal approach, and a medial supraumbilical extension of the incision revealed a congested, slightly enlarged spleen overlying the urinary bladder and fixed to it with mul-
multiple adhesions. An infarcted area of about 3 × 3 cm was also found near the lower pole of the spleen, which correlated with the sonographic findings. The spleen was devoid of its ligamentous support, and its pedicle was found twisted by 180°. The left kidney was normally placed. A splenectomy was performed, and the left dome of the diaphragm was plicated using multiple rows of nonabsorbable sutures. The patient had an uneventful recovery and was discharged on the seventh postoperative day. The child remains well after 1 year of follow-up.

Discussion

Development of the diaphragm is a highly complex process that takes place during early embryonic life. It develops from the septum transversum, the dorsal mesentry of the esophagus, and the pleuroperitoneal membranes; the body wall contributes to its muscular portion. It is the incomplete development of this muscular portion that results in diaphragmatic eventration. The spleen develops in the left upper quadrant of the abdomen from the mesenchymal cells of the dorsal mesogastrium. The splenorenal ligament, which forms by fusion of the posterior leaf of the dorsal mesogastrium with the parietal peritoneum in front of the kidney, contains the splenic pedicle and is considered to be the primary support of the spleen. The gastrosplenic, phrenicocolic, and splenophrenic ligaments are considered to be secondary support structures of the spleen. The phrenicocolic ligament is part of the greater omentum that extends between the splenic flexure of the colon and diaphragm and forms a sling in which the spleen rests. Abdominal pressure and support from the surrounding viscera and abdominal musculature also hold the spleen in position.

Eventration of the diaphragm is known to be associated with a constellation of congenital anomalies, the most commonly described being pulmonary hypoplasia, malrotation of the gut, and various cardiac anomalies. On the other hand, the congenital anomalies predisposing to a wandering spleen are positional abnormalities of the colon and an absent or ectopic kidney. Because diaphragmatic defects are usually associated with a high-sited or intrathoracic spleen, to find a suprapubic spleen with left diaphragmatic eventration, as seen in our patient, is not only surprising but paradoxical. To the best of our knowledge, there has been only one other such case, described by Melikoglu et al. in 1995, noting a similar association. Those authors hypothesized that this association is due to abdominal wall laxity. We believe that because there was congenital maldevelopment of the diaphragm the resulting absence of the phrenicocolic ligament, which forms a sling for the spleen, also may have contributed to the ectopic location of the spleen.

Ultrasonography is the current modality of choice for detecting both the diaphragmatic defects and the ectopic spleen. As spleen-preserving procedures are