Journal of Pediatric Surgery (2013) **48**, 449–453

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Journal of Pediatric Surgery

www.elsevier.com/locate/jpedsurg

Infantile myofibromas obstructing opposite ends of the gastrointestinal tract

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Received 6 July 2012; revised 26 November 2012; accepted 27 November 2012

Key words:
Myofibroma;
Congenital;
Visceral;
Intestinal;
Obstruction;
Neonatal

Abstract Myofibromas are benign congenital tumors of soft tissue that can present at birth or during infancy in solitary or multicentric forms. Visceral myofibromas are rarely reported, but are typically symptomatic due to involvement of vital structures. We present two cases of congenital myofibromas, one obstructing the proximal esophagus and the other obstructing the distal rectum. Lessons learned from the treatment of these two patients are shared and the pertinent literature is reviewed.

Myofibromas are benign neoplasms of myofibroblasts. Although rare, they represent the most common fibrous tumor of infancy and early childhood [1]. Presentation is highly variable and can lead to a broad spectrum of clinical symptoms, depending on location. Treatment options differ, based on the location of the lesion and the rate of growth. Here, we present two cases of visceral myofibromas resulting in neonatal obstruction of the upper and lower gastrointestinal tract.

1. Case 1

A baby girl born at 36 weeks gestation, with a history of maternal polyhydramnios and absent gastric bubble, was presumptively diagnosed with Type A esophageal atresia due to inability to pass a nasogastric tube, combined with a gasless abdomen on x-ray. On the second day of life, the patient developed respiratory distress. Evaluation by the otolaryngology service identified bilateral vocal cord paralysis, as well as a mass at the level of the right arytenoid. The patient required intubation. An MRI suggested very proximal esophageal atresia with the esophagus terminating at the third cervical vertebra and non-visualization of the distal esophagus (Fig. 1). A solid mass measuring 1.9 cm × 0.4 cm × 0.8 cm was seen lateral to the right vocal cord, descending in the tracheoesophageal groove, extending inferiorly to the cranial aspect of the right thyroid lobe.

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http://dx.doi.org/10.1016/j.jpedsurg.2012.11.041

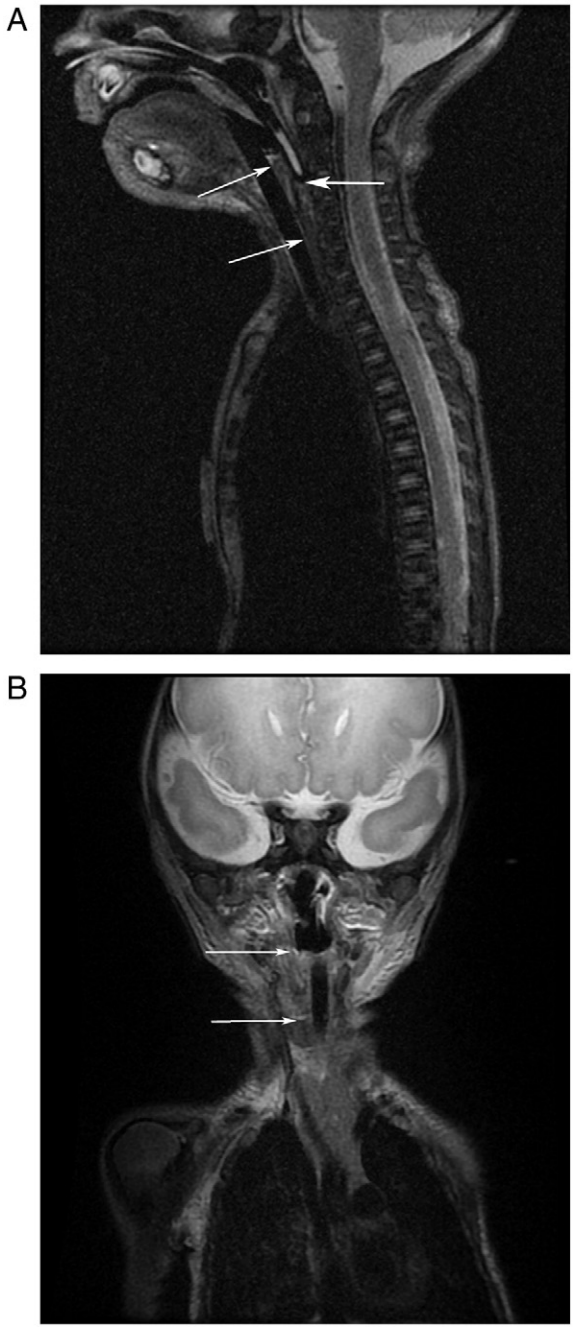

Fig. 1 T2-weighted sagittal (A) and coronal (B) MRI of Case 1. On sagittal view, the tip of the blind ending esophagus with nasogastric tube in place is indicated by the right arrow, while the mass borders are indicated by the left arrows. The coronal image better demonstrates the mass with its proximal and distal extents (arrows).

The patient underwent a biopsy of the mass and insertion of a gastrostomy. Contrast instilled through the gastrostomy tube refluxed into the distal esophagus up to the fifth cervical vertebra, establishing a gap of two vertebral bodies. Biopsies of the 'submucosal' mass yielded normal mucosa. A work-up for VACTERL association revealed no other congenital anomalies.

The patient was fed by gastrostomy to allow the gap to decrease, and to coordinate operative intervention with otolaryngology. At day of life 51, she underwent a joint procedure. At the start of the procedure, a guidewire advanced pre-operatively by interventional radiology through the gastrostomy tube into the distal esophagus to aid in its identification, was found exiting through the mouth without resistance, dispelling the diagnosis of esophageal atresia. The mass was then excised from the right lateral pre-epiglottic space. Pathology revealed the mass to be a myofibroma.

A symptomatic left orbital myofibroma was diagnosed at approximately 3.5 months of age, and resected by ophthalmology.

Complete work-up of the patient revealed multiple congenital anomalies including left thyroid lobe agenesis, uterine didelphys, central sleep apnea, and minor upper limb anomalies, in addition to the previously diagnosed bilateral vocal cord paralysis. A general chromosomal microarray was normal, and no genetic syndrome could be identified.

At 1 year of age, the patient was still dependent on gastrostomy feeding due to severe nasopharyngeal reflux, and vocal cord paralysis leading to a tracheostomy. In addition, beginning at approximately 3 months of age, multiple transient soft tissue masses on the scalp, neck, abdominal wall, and tracheostomy site appeared and resolved within two to three weeks. However, two myofibromas, in the right submandibular area and along the left aspect of the tongue, failed to resolve, and were felt to be potentially life-threatening due to location and rapid growth. A regimen of vinblastine and methotrexate appeared to result in arrest of further tumor growth and gradual regression. A trial off chemotherapy was being considered at the time of this report.

2. Case 2

A male infant born at term presented on the first day of life with a distal bowel obstruction. On rectal exam, a pelvic mass was felt, with severe rectal stenosis. Abdominal CT showed a 3 cm × 2 cm mass in the pelvis abutting the rectum, causing extrinsic rectosigmoid obstruction and compressing the bladder, without discrete borders.

A trans-rectal needle biopsy of the pelvic mass and a diverting sigmoid colostomy were performed on the third day of life. Due to the size and fixation of the mass, as well as the possibility of a malignancy that may respond to neoadjuvant chemotherapy, an attempt at resection was deferred. Differential pathologic diagnoses included fibrosarcoma and myofibroma. Testing for 12;15 translocation was negative. After extensive pathologic review, a diagnosis of low-grade myofibroblastic lesion was made. The patient continued to manifest symptoms of partial bowel obstruction with inability to achieve full oral feeding. MRI of the pelvis (Fig. 2) at 30 days of life revealed no contraindications to resection.