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Management of visceral vascular anomalies

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ABSTRACT

Vascular malformations affect the viscera less commonly than the head and neck, extremities, and extracavitary soft tissues. They present with a wide spectrum of symptoms and findings including pain, respiratory compromise, hemoptysis, chylothorax, ascites, gastrointestinal bleeding, and obstruction. Management options depend upon the subtype of malformation and anatomic extent and may include sclerotherapy, embolization, surgical extirpation, coloanal pull-through, and occasionally more innovative individualized surgical approaches.

Management of visceral vascular malformations

Vascular malformations can affect all regions of the body. Visceral involvement can be extensive and problematic. Presentation, complications, and management options are determined by the type of lesion, the organ systems affected, and extent of involvement. Vascular malformations are anomalies that occur during the morphological development of the vascular system. These lesions are present at birth, generally growing with the child, but often undetected until symptoms occur. Some are associated with or are direct extensions of more visible cutaneous and soft tissue lesions. These malformations can be exacerbated by trauma, infection, or hormonal changes. Herein we present an overview of some of the types of visceral vascular malformations and management options. For more comprehensive descriptions and discussion of more rare entities, the reader is referred to Chapter 25 of Mulliken and Young's Vascular Anomalies, 2nd Edition.[1]

Thoracic vascular malformations

Lymphatic malformations

Most vascular malformations in the thoracic cavities and mediastinum are lymphatic malformations (LMs). Mass lesions can be macrocystic, microcystic, or combined. Microcystic malformations are sponge-like masses with individual cysts that are too small to drain. Macrocystic lesions are almost always multicystic masses similar in appearance to compressed bunches of variably sized grapes. Thoracic LMs are most commonly in the mediastinum or retropleural space. They can be in contiguity with lesions in the thoracic inlet, neck, retroperitoneum, or torso soft tissues. Small asymptomatic lesions can be followed without intervention. Lesions causing or threatening airway compression or that are so large as to impact pulmonary development should be treated. Macrocystic lesions are generally best ameliorated with image-guided intralesional sclerotherapy, as described in detail by Elluru and Padua elsewhere in this monograph. Large predominantly microcystic lesions require resection. These lesions are not malignant, so vital structures should not be sacrificed; however, leaving large amounts of residual LMs will generally lead to re-expansion of the lesion. Reoperation to remove residual scarred mass is very challenging and creates significant increased risk for neurovascular injury. Thus, the surgeon undertaking such a resection should be prepared to meticulously remove nearly the entirety of the lesion in a defined anatomic region regardless of operative duration.

LM masses in the lung parenchyma are extremely rare. Lobectomy cured life-threatening hemoptysis in one case.[2] The most common intrapulmonary lymphatic anomaly is congenital pulmonary lymphangiectasia (CPL). CPL is a rare disorder that is thought to result from a diffuse dysplasia of the lymphatic network. It is characterized by dilation of the pulmonary lymphatics that drain the subpleural and interstitial spaces of the lung. Autopsy results suggest that CPL has an incidence of 0.5–1% in infants who die in the neonatal period.[3] Cases are thought to be sporadic although associations with Noonan's, Downs, and Ullrich–Turner syndromes are known. There are preliminary reports that VEGF-C overexpression can induce pulmonary lymphangiectasia during development.[4] Patients can present in utero as a hydrops fetalis with pleural effusions.[5] It appears at birth with intractable respiratory

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distress in the newborn, often associated with chylous pleural effusions.[6] It may also present as part of generalized lymphangiectasias, as secondary to pulmonary venous hypertension or venous obstruction, or as isolated lesions. Diagnosis is made with a combination of radiologic, pathologic studies in conjunction with the clinical presentation. A chest radiograph has increased interstitial markings, hyperexpansion, and may have pleural effusions. Chest computerized tomography (CT) scans show ground glass infiltrates, interstitial densities, and pleural effusions. Lung biopsy is the most definitive means of diagnosing these patients. Biopsy reveals dilated lymphatics within the interstitium of the lung. Treatment of this disease is supportive with gentle ventilation strategies, extra-corporeal membrane oxygenation (ECMO), and nutritional support, all playing important roles. Localized lesions are sometimes amenable to surgery. The overall prognosis, however, remains poor when patients present in the neonatal period despite advances in neonatal intensive care.[3]

Particularly problematic lymphatic conditions in the chest include those resulting in pleural effusion and/or chylothorax. These include generalized lymphatic anomaly, Gorham–Stout disease,[5] and poorly characterized central conducting lymphatic anomalies. Medical management for chylothorax has included octreotide, interferon,[7,8] and, more recently, sirolimus therapy has been utilized.[9] There is also some preliminary data that radiation therapy may effectively prevent bony disease progression and may also aid in management of resistant chylothoraces, using total doses ranging from 30 to 45 Gy.[10] The long-term outcome of radiation treatment is still unclear in these patients.

Surgical management of chylothorax secondary to lymphatic disease can be quite challenging and is often unsuccessful. Pleurodesis, ligation of the thoracic duct, and pleurectomy have all been reported with some success in the management of effusions. Recently, there have been reports of the use of embolization of the thoracic duct to treat chylothorax.[11] Reconstruction of the thoracic duct by lymphovenous anastomosis has been performed successfully, but its general applicability remains to be established.[12] Often, a combination of both medical and surgical treatment is the best means of management of these difficult problems.[13]

Venous malformations

Endothoracic venous malformations (VMs) of significance are uncommon and are usually extensions of chest wall or neck lesions. Treatment is rarely necessary, and image-guided endovascular techniques are almost always preferred. Although subtotal surgical resection of large chest wall VMs can be performed without exsanguination by closing skin over residual lesion, this is not possible inside the chest. Thus, endothoracic surgical resection of a poorly circumscribed VM is almost never indicated.

Arteriovenous malformations

Arteriovenous malformations (AVMs) in the thorax are also uncommon. Most are best managed, if not cured, with endovascular obliterative techniques, discussed by Richter and Alomari elsewhere in this issue. Intrapulmonary arteriovenous malformations can cause hemoptysis and shunting, causing hypoxia and high-output cardiac failure. They can also permit paradoxical embolism. Some such lesions are due to hereditary hemorrhagic telangiectasia (HHT). HHT is an autosomal dominant disorder with manifestations that include visceral arteriovenous malformations seen in the GI tract, lungs, and liver. Mucosal involvement is common with epistaxis being the most frequent symptom. In a recent study, about 20% of children with HHT were noted to have pulmonary AVMs.[14] These lesions are also often diffuse, and generally, surgical resection is not usually possible or indicated.

Abdominal vascular malformations

Lymphatic malformations

Abdominal lymphatic malformations often arise from the mesentery, omentum, or retroperitoneum. These may remain asymptomatic for long durations and thus sometimes do not become apparent until adulthood. Intra-abdominal LM is more commonly macrocystic than microcystic. Children tend to present acutely with abdominal pain, distention, nausea, diarrhea, or constipation.[15] The cysts are generally anechoic or contain echogenic debris. Imaging studies reveal a uni- or multi-septated mass with thin walls (Figure 1). Initial study should be ultrasonography, and for those in whom sonography does not provide enough anatomic information, MR imaging should be undertaken. Treatment of symptomatic or large intra-abdominal LMs is indicated. Traditionally, surgical resection was the primary treatment of choice; however, this may involve significant risk of bowel injury, persistent chylous ascites, and re-enlargement of residual lesion. Segmental bowel resection is only justified for a limited regional lesion that can be completely excised. Macrocystic lesions can usually be easily substantially reduced with percutaneous image-guided intralesional sclerotherapy. Doxycycline is the most commonly used agent.[16,17] Lymphatic cysts are aspirated under US guidance and the sclerosant is injected into the cyst, under fluoroscopic guidance to ensure proper distribution of the agent. In larger malformations, a closed suction drain may be placed to aspirate ongoing fluid drainage and perform repeated doxycycline instillations. Microcystic lesions are more commonly asymptomatic. Surgical resection is occasionally required for large lesions causing mass effect, such as urinary tract obstruction. Occasionally a LM, either macrocystic or microcystic, can mimic an inguinal hernia, sometimes extending fully into the scrotum (Figure 2). Macrocystic lesions can be sclerosed. Microcystic lesions require meticulous dissection since they are usually intimately associated with the vital structures of the spermatic cord. Partial resection will result in re-expansion of the residual malformation in the

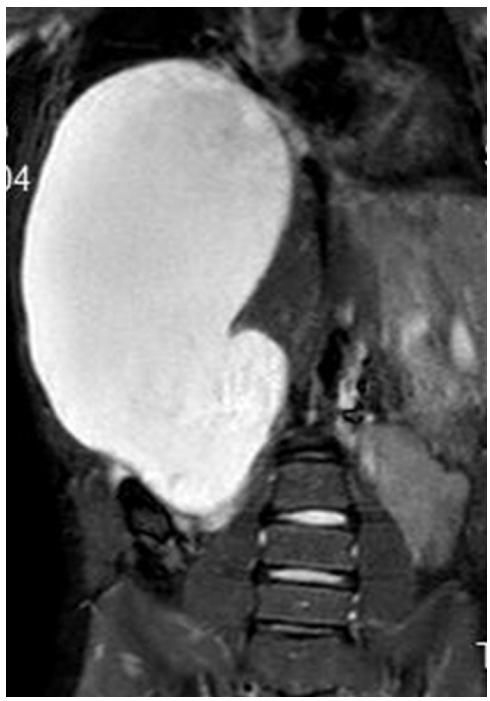

Fig. 1. A T2-weighted MR image demonstrating a large retroperitoneal lymphatic malformation.