composed of smooth muscle fibers bundled with fibrous and neural tissue. They have little cytologic atypia and low mitotic activity. Eosinophilic interlacing spindle cells with elongated cigar-shaped nuclei are observed.

Surgical resection is the treatment of choice and is generally recommended for large or symptomatic tumors. Because of the exceedingly rare incidence of malignancy [2], small asymptomatic leiomyomas may be observed. Resection is most commonly performed by thoracotomy or a video-assisted thoracoscopic approach, although robotic techniques have also been reported, even for complex tumors [5]. Video-assisted thoracoscopic resection offers the advantage of decreased postoperative pain and length of hospital stay [6]. If thoracotomy is indicated, a right-sided approach is used for lesions in the upper two thirds of the esophagus, whereas a left-sided approach is used for more distal lesions. Right thoracotomy was chosen for the patient in this case because of the location, large size, and circumferential nature of the tumor. Enucleation is preferred and is generally feasible; however, mucosal injury is fairly common (12.7%) and is associated with preoperative endoscopic biopsies within 1 month of resection but not with tumor size, location, or operative approach [7]. When injury occurs, the mucosa should be repaired in 2 layers with absorbable suture. Formal esophageal resection is rarely indicated but is more common if the tumor is large (> 8 cm), annular, or densely adherent to surrounding structures [2].

Large series have demonstrated excellent outcomes after resection, with long-term symptomatic relief, low recurrence rates, and low mortality rates [4, 7]. Malignant transformation is exceedingly rare but has been described in large case series [2]. Although the majority of esophageal leiomyomas are smaller homogeneous submucosal tumors, some tumors may become quite large and present as heterogeneous calcified circumferential masses, as in this patient. Careful operative planning and meticulous dissection are critical for optimal patient outcomes.

References

Expansile Kaposiform Hemangioendothelioma Deformed Thoracic Cage in an Adult
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Kaposiform hemangioendothelioma is a vascular tumor that commonly presents as a cutaneous mass, is observed in children, and is associated with Kasabach-Merritt phenomenon. Herein we report a case of kaposiform hemangioendothelioma with chest wall deformity in an adult who did not show the Kasabach-Merritt phenomenon or cutaneous lesions. To our knowledge, this is the first case of asymptomatic kaposiform hemangioendothelioma arising from the pleura and deforming the chest wall. The patient was treated with tumor excision and chest wall reconstruction.

Accepted for publication Feb 25, 2013.

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A 21-year-old man presented with a chest wall tumor in our clinic. Right-sided chest wall protrusion and scoliosis had developed when he was 15 years old, and he started to feel mild pain in his right chest at that time. He had not experienced any inciting trauma but described that the pain had worsened with time. On physical examination, protrusion of the ribs with localized tenderness to palpation was noted over the...
area between the right nipple and the sternum. No palpable masses, overlying skin changes, erythema, or discoloration was observed in that area.

Roentgenography revealed severe scoliosis and deformity of the thoracic cage (Fig 1). Computed tomography of the chest with intravenous contrast medium revealed an enhancing mass in the right fifth costochondral junction invading multiple ribs (Fig 2A). Whole-body bone scintigraphy revealed mildly increased tracer uptake at the right fifth costochondral junction. The patient underwent chest wall tumor biopsy for pathologic diagnosis. Pathologic assessment revealed a vascular lesion. Therefore, magnetic resonance imaging was performed for further examination (Fig 2B and C).

Preoperative angiography (Fig 2D) and transarterial embolization were performed smoothly. The tumor’s supplying arteries, including a branch of the right internal mammary artery and the right intercostal arteries (T4, T5, and T6), were embolized.

Two weeks after preoperative embolization, we performed chest wall resection, removing the tumor and parts of the fourth, fifth, and sixth ribs. During the operation, we found an 8.5 × 7.2 × 5.4 cm tumor at the right anterior chest wall that bled easily. The tumor originated from the pleura and had invaded the ribs. A
chest wall defect, 12 × 10 cm, was created and reconstructed with a titanium mesh. The patient experienced a smooth postoperative recovery; the chest tube was removed on postoperative day 3, and the patient was discharged on postoperative day 6. We followed up the patient 1 week, 1 month, 2 months, and 5 months after the operation. His condition was stable without pain or recurrence.

The pathologic examination revealed a cellular tumor with a multinodular growth pattern and an infiltrative border. The tumor-free margins were approximately 1 cm on the right and 2.5 cm on the left. Rib and skeletal muscle invasion were discernible. Each nodule was composed of spindle cells and numerous interspersed small vessels containing erythrocytes (Fig 3A). Some larger vessels were seen in the peripheral area of the nodules. The tumor cells were immunoreactive for CD31 (Fig 3B) and D2-40. On the basis of these findings, a diagnosis of KHE was made.

Comment

Our case did not present any of the characteristic findings of KHE. KHE rarely occurs in adults older than 21 years. According to published reports, several features of KHE in children are different from those in adults [5]. KHE shows equal sex distribution in children, but 80% of patients with adult-onset KHE are male [3]. The tumor diameter for adult-onset KHE is small: only 4.5 cm on average. By contrast, children usually have tumors larger than 5 cm in diameter. KMP is not often observed in adults with KHE because small lesions are less likely to cause platelet abnormalities.

The reason for the association between thoracic cage deformity and KHE in our case is unclear. Our case may be similar to the case of scoliosis with KHE-related spinal destruction reported in 2011 by Zhu and colleagues [6]. Tumor expansion may have led to the destruction of the ribs, which resulted in progression of thoracic cage deformity. We hypothesize that the tumor initially presented at age 15 and led to the pectus deformity. The scoliosis may have been secondary to the abnormal rib growth, though this remains speculative because the tumor was imaged only after the patient turned 21 and became symptomatic. Although some previous adult cases of KHE manifesting in areas of previous trauma have been reported [5], and various forms of hemangioendothelioma have been observed in damaged tissues [7], there was no evidence that our patient had any history of chest wall trauma.

Because KHE is rare, no treatment guidelines have been established for this disease. Aggressive treatment is usually needed for symptomatic KHE, especially when it is associated with KMP. Complete surgical removal is the primary therapeutic option, and most tumors in adults can be successfully excised. However, masses in the retroperitoneum are typically extensive, and unresectable lesions frequently lead to the patient's death. Such tumors respond best to vincristine, interferon, and corticosteroid treatment. In our case, an operation was necessary because vascular tumors in the chest wall have the potential to cause hemotorax [8]. We suggest 1-cm safe margins for the operation and preoperative transarterial embolization because these strategies reduce both the chance of bleeding and the difficulty of the operation. If the tumor recurs, we shall consider additional medical treatment because KHE responds well to radiation, chemotherapy, and corticosteroid therapy.

This report suggests that physicians should always keep in mind all possible pathologic conditions that can cause bone destruction. We hope that our case prompts future research on the poorly understood disease, KHE.

This study was supported by the National Taiwan University Hospital (NTUH.101-S1793) National Health Research Institutes (NHRI-EX102-10032BI), and National Science Council (NSC101-2314-B002-020-MY3) of the Republic of China.
Unusual Presentation of Lymphoepithelioma-Like Carcinoma of Lung as a Thin-Walled Cavity

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Most thin-walled cavities in the lung are benign lesions, but a few cases of lung cancer can have this unusual pattern. All previously reported cases were adenocarcinomas. Here we report a case of lymphoepithelioma-like carcinoma (LELC) of the lung presenting as a thin-walled cavity with a smooth inner surface. LELC is more commonly seen in Southeast Asia, including Taiwan, and its gross picture is usually a solid mass with a round, circumscribed border, which is indistinguishable from other non-small cell lung cancers. Asymmetric thickness of the cystic wall and lymphadenopathy are important features in the diagnosis and the selection of treatment.

Accepted for publication March 1, 2013.

Comment

The thickness of a cavity wall has already been recognized by radiologists as useful in evaluating solitary cavities in lungs. Woodring and colleagues [1] showed that most cavities whose maximum wall thickness was 4 mm or less were benign and those with maximum wall thickness more than 15 mm were malignant. It becomes indecisive when the maximum wall thickness is between 5 and 15 mm because half are benign and half are malignant [1]. Another useful feature is the contour of the cystic wall, because benign lesions usually have a smooth wall, whereas more than 80% of cases with irregular wall were malignancies in the study by Woodring and colleagues [1].