Case Challenge

Editor's note: Each month, this department features a discussion of an unusual diagnosis. A description and images are presented, followed by the diagnosis and an explanation of how the diagnosis was determined. As always, your comments are welcome via email at pedann@Healio.com.

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A 16-Year-Old Boy with Acute Chest Pain and Shortness of Breath

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A 16-year-old boy with a history of adenoidal hypertrophy, gastroesophageal reflux disease, and pectus carinatum presented to his pediatrician with “unbearable” chest pain and a dry cough after 1 week of mild but constant chest pain. His temperature was 36.6°C, his pulse was 68 beats per minute, his blood pressure was 96/64 mm Hg, his respiratory rate was 13 breaths per minute, and his O₂ saturation was 100% on room air. His pulmonary examination was clear to auscultation bilaterally.

Seven days prior to admission, the patient experienced acute superficial centrally located chest pain rated at 8 of 10 along with shortness of breath while playing a game of catch. As the pain did not remit, the patient presented to the emergency department of a local hospital on the same day. There, he underwent transthoracic echocardiography and chest radiography, both of which were unremarkable. He was diagnosed with pleuritic chest pain and discharged with ibuprofen.

The patient was seen in his pediatrician’s clinic 1 week after his previous visit to the emergency room. After the pediatrician reviewed the chest radiograph that was ordered, the patient was admitted to the inpatient unit of our hospital.

PHYSICAL EXAMINATION

On admission the patient was alert, interactive, and in no apparent distress. His temperature was 36.7°C, his pulse was 60 beats per minute, his blood pressure was 139/73 mm Hg, and his respiratory rate was 20 breaths per minute. His O₂ saturation was 100% on room air. His height was 185.4 cm (95th percentile) and his weight was 67.1 kg (approximately 70th percentile, for a body mass index of 19.5 kg/m²); the patient’s wingspan was 188 cm. His heart had a regular rate and rhythm, with normal S1 and S2 heart sounds and no murmurs, rubs, or gallops. His lungs were clear to auscultation bilaterally, with no wheezes, rales, or rhonchi. No breath sounds were noted at the left lung apex. He demonstrated appropriate strength and tone on musculoskeletal examination; no joint laxity or wrist hypermobility was noted. Lordosis and scoliosis were not demonstrated on spinal examination.

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Diagnosis:

Left-Sided Spontaneous Pneumothorax

The patient underwent chest radiography (posterior-anterior and lateral films) prior to his admission to the pediatric inpatient unit, and the radiographs showed a moderate to large left-sided pneumothorax with minimal air in the mediastinum. There was no pneumothorax on the right side. No signs of tension pneumothorax were present, the lungs were clear bilaterally, and no effusions were noted.

HOSPITAL COURSE
The patient was placed on continuous pulse oximetry without supplemental oxygen, and thoracic surgery was consulted. The decision was made for chest tube drainage with suction, primarily due to the size of the pneumothorax. The patient was initially placed on suction at 20 cm H₂O overnight, and a chest radiograph performed approximately 2 hours after chest tube placement documented a significant interval improvement in the size of the pneumothorax.

On the first day after admission, the pneumothorax increased in size from tiny to small, and subsequently evolved to a moderate-sized pneumothorax the next day; the patient was on overnight suction for both nights. A persistent air leak was also noted on the water seal, and given the increase in the size of the pneumothorax, the decision was made for surgical intervention. Three days after admission, video-assisted thoracoscopic surgery was used to perform an apical bleb resection and apical pleurectomy; pleurodesis was also performed by mechanical abrasion of the remaining parietal, mediastinal, and diaphragmatic pleura. The patient had an unremarkable postoperative course; his chest tube was maintained at -20 cm H₂O suction for 3 days postoperatively. Chest radiography performed on postoperative day 3 showed re-demonstration of a tiny left apical pneumothorax that was stable on water seal, so the decision was made to remove the chest tube and discharge the patient. A radiograph performed on postoperative day 17 showed no residual pneumothorax. The patient has had an otherwise unremarkable clinical course after discharge and was doing well at 8 months of follow-up with no recurrence of pneumothorax to date.

MAKING THE CALL: A NEED FOR SUSPICION

With an incidence of approximately 3 per 100,000 children, spontaneous pneumothorax (SP) is relatively uncommon. However, the set of characteristics found in our patient lends itself to the diagnosis. For example, SP is 4 times more common in boys than in girls, and it should be suspected in teenagers possessing an asthenic body habitus with the appropriate clinical picture. In fact, around 82% of SPs occurring in patients between the ages of 0 and 17 years were noted in those older than age 15 years, with only 7% occurring in those younger than age 10 years.

Knowledge of the pathogenesis of SP may also assist the clinician in suspecting the diagnosis. Acute increases in pressure at the alveolar-pleural interface are known to distend and rupture alveoli. Given the preferential ventilation and greater pressures encountered in the upper part of the lung, it comes as no surprise that apical blebbing is often encountered during SP. The tendency for rupture may be particularly great with the weak visceral pleura encountered in some lung diseases. Therefore, a reasonable amount of suspicion may also be appropriate in patients with conditions such as cystic fibrosis, asthma, necrotizing pneumonia, and foreign body aspiration, or in those who smoke.

Conversely, SP may be indicative of an underlying lung condition or anatomic abnormality. Our patient’s stature and pectus carinatum were suspicious for Marfan syndrome. Although testing can be performed for the FBN1 mutation, diagnosis of Marfan syndrome requires satisfaction of specific clinical criteria, with two of the most important being lens subluxation and aortic root abnormalities. An ophthalmology follow-up at approximately 3 months was only significant for hypermetria, which is not clinically suggestive of the disease. The patient was also recommended for an outpatient echocardiogram.

SPONTANEOUS PNEUMOTHORAX: A TEXTBOOK PRESENTATION
An SP is a collection of air in the pleural space occurring in the absence of trauma due to disruption of the visceral pleura, and it can be further classified as a primary spontaneous pneumothorax (PSP) or secondary spontaneous pneumothorax (SSP). PSP is often a diagnosis of exclusion, as it is defined by the absence of any underlying lung disease. Causes of SSP include connective tissue disorders, interstitial lung disease, and malignancies, but a recent assessment of a national inpatient database noted asthma without status asthmaticus as the most common associated condition.

As seen in our patient, most SPs occur when the individual is at rest or under mild exertion. Our patient also exhibited very classic presenting symptoms: ipsilateral chest pain that may radiate to the shoulder, dyspnea, and a dry cough. Symptoms usually resolve within 24 hours even if the SP does not. If the pneumothorax is large, physical examination results can include decreased chest wall movement, diminished breath
sounds, and tactile fremitus as well as increased resonance with percussion on the ipsilateral side. If the pneumothorax is small, tachycardia may be the only sign. In fact, our patient had only one sign of pneumothorax—absent breath sounds at the left lung apex.

**IMAGING THE LUNGS**

SP can be diagnosed with a chest radiograph, preferably in the upright position, as posterior-anterior and lateral views of the chest highlight the visceral pleura. Associated signs include flattening of the diaphragm, hyperlucency, and a contralateral mediastinal shift in cases of tension pneumothorax. Although chest radiography is the most common method used to detect pneumothorax, ultrasonography (US) may be a superior imaging modality, especially for ruling out the diagnosis. This was highlighted by a 2012 meta-analysis (1,048 total patients) that documented 90.9% versus 50.2% sensitivity and 98.2% versus 99.4% specificity in detecting pneumothorax for ultrasound versus chest radiographs, respectively. However, it should be noted that these data are primarily obtained from cases of traumatic pneumothorax, given that US is routinely used as part of the Extended Focused Assessment with Sonography for Trauma protocol. Findings on US include the “barcode” sign (a pronounced reflection arising at the junction of the parietal pleura and pneumothorax with concomitant disappearance of the normal sliding pleural line) and an absent “lung pulse” (the normal transmission of cardiac movement to the pleura).

**MANAGEMENT OF SPONTANEOUS PNEUMOTHORAX**

Although management of SP in children is extrapolated from adult guidelines, initial management of SP largely depends on the patient’s clinical status, the size of the pneumothorax, and the presence of underlying lung disease. Conservative management often includes 100% high-flow oxygen administration by facemask, not just for maintenance of appropriate O₂ saturation but for more rapid resorption of the pneumothorax. Admission and observation may be all that are required in up to 30% of patients with SP, but the most common interventions for SP include chest tube drainage, bleb excision, and thoracoscopy. Regardless of intervention, a significant concern after resolution of a primary SP is recurrence.

Two studies in the pediatric population, comprising a total of 89 patients, have noted recurrence rates between 51% and 61% after PSP, so identification of patients at high risk for recurrence and appropriate early intervention is necessary. A potential indicator has been the presence of apical blebs and bullae on computed tomography (CT). Some studies have shown that CT itself is poor for bleb identification, and that recurrence rates are similar between those with and without blebs. In addition, an evidence-based management article from 2009 stated that there is no indication for the use of CT for bleb or bullae assessment after the initial occurrence of SP, and this is in concordance with a 2001 consensus statement from the American College of Chest Physicians (ACCP). However, a retrospective review from 2013 of 114 children undergoing conservative management after primary PSP found that the presence of blebs or bullae on high-resolution CT was a significant risk factor for ipsilateral recurrence. Ultimately, studies in children have been inconclusive, but CT may have utility depending on the clinician’s threshold for surgical or preventative intervention after first-time SP.

Currently, guidelines suggest surgery at time of primary PSP for patients with bilateral SP or continued air leak after 5 days of drainage, but there is evidence suggesting decreased length of stay, length of chest tube drainage, and recurrence of SP with surgery at time of PSP when compared with conservative management. In modern practice, patients with persistent air leak after 48 hours are recommended for VATS bleb resection and pleurectomy, as are patients with PSP and documented blebs or bullae. Observation is recommended for patients who do not require chest tube drainage and who do not have documented blebs or bullae. Decreases in morbidity are greatest with VATS when compared with open procedures, whereas rates of recurrence may be higher (5% vs 1%). Intervention prior to the second pneumothorax was recommended by 15% of the ACCP Pneumothorax Consensus group, and seems to be a reasonable option given the significant burden of recurrence.

**CONCLUSION**

Although SP is usually encountered in a target demographic (ie, teenage boys older than age 15 years), it should still be suspected when there is underlying lung disease or acute increases in transpulmonary pressure. The most common presenting symptoms include ipsilateral chest pain, dyspnea, and a dry cough. Although diagnosis is conventionally made through chest radiography, US may come to be the method of choice given its increased sensitivity and equal specificity.

Management criteria for SP are not clearly defined in children, and there is no conclusion regarding the role of surgery for PSP. The most recent adult SP management guidelines instituted by the British Thoracic Society recommend a surgical consultation in cases of a persistent air leak (5-7 days) or bilateral synchronous SP. However, there
are documented advantages to performing surgery at the time of PSP, the most important being eliminating the risk of recurrence. VATS procedures have lower morbidity than open procedures with comparable efficacy, and it seems that shared-decision making will play an increasingly prevalent role in future patient management.

REFERENCES