

PULMONARY LYMPHANGIOLEIOMYOMATOSIS

by Respiratory Care Student Daniel Nordstrom



Lymphangioleiomyomatosis (LAM) is a rare lung disease of the lung parenchyma that usually affects childbearing age women. LAM is pathologically characterized by an unusual type of muscle cell that invades the tissues of the lungs, including the airways, blood and lymph vessels. Over time, these atypical pulmonary interstitial smooth muscle cells obstruct the flow of air, form cysts within the lung, and prevent the lungs from ventilating and oxygenating the body. LAM may be associated with a genetic disorder known as Tuberous Sclerosis (TS). There have been over 1,000 women diagnosed with LAM in the US, but the exact number of people with LAM is unknown. Scientific studies of LAM estimate over 250,000 women have LAM worldwide. Difficulty in diagnosis may be due to the initial presentation of LAM.

"The diagnosis of LAM can be difficult because many of the early symptoms are similar to those of other lung diseases. LAM is most often misdiagnosed as asthma, emphysema, or pulmonary bronchitis. LAM may not appear on an x-ray, and may require a high resolution CT scan. An abdominal CT scan is also recommended, as a benign kidney tumor, known as an angiomyolipoma, is found in 40% of women with LAM. A lung biopsy is recommended only as a last resort for diagnosis".

The classical presentation of LAM is often, young healthy women begin experiencing progressive dyspnoea, suffer recurrent spontaneous pneumothoraces and haemoptysis, culminating in respiratory failure and death. However, some patients may initially present with extrapulmonary symptoms. In this case report the case of a young woman that initially presented with a leiomyoma located on her right kidney, a possible rhabdomyoma of her heart, skin lesions, and possible retinal hamartomas, suggesting a diagnosis of TS. Until the sudden development of pleurisy followed by hemothorax and a collapsed lung, less than 1.5 years post diagnosis, resulting in the discovery of the underlying lung disease LAM.

CASE HISTORY

A 26 year-old Chinese American female social worker visited her family physician in 2002 with a history of back pain for more than one year prior. There was no prior history of drug use, oral contraceptives, trauma, or travel. She had a smoking history of 1-2 cigarettes per day for two years and was raised in a non-smoker household. Alcohol use was described by patient as being "normal college kid consumption" with no continued use post graduation. She had no prior history of illness and her previous health was good. Her doctor noted possible retinal hamartomas with mild skin lesions and arranged a CT scan of her thorax and abdomen. Results of CT showed a leiomyoma on the right kidney, possible rhabdomyoma of the heart suggesting diagnosis of TS. At one and a half years post diagnosis she developed sudden pleurisy

during a white water rafting trip. Less than two months later she presented at emergency room with massive left hemothorax and collapsed left lung. Physical exam findings were unremarkable with the exception of the hemothorax and collapsed lung. CT angiogram was ordered to confirm a pneumothorax. The CT showed cystic lung disease combined with a large left pneumothorax and loculated right pneumothorax. Bilateral chest tubes were placed and she underwent mechanical pleuraldesis with bilateral thoroscopic surgery. During the procedure the patient was mechanically ventilated on assist control with pressure support of 10. Tidal volume was set at 600 cc's and respiratory rate was recorded at 12 breaths per minute. Intubated with a 7.5 endotracheal tube measured to be 21 cm at the teeth, her peak inspiratory pressures were 21-25 mmHg with static pressure noted at 19 mmHg. One hour post surgery she was extubated and placed on 4 LPM nasal cannula and maintained a SaO₂ of 98%. Two hours later nasal cannula was reduced to 2 LPM. The right chest tube was then discontinued while the left chest tube was continued with Heimlich valve and she was discharged to home care with oxygen therapy continued at 2 LPM three days post surgery.

DISCUSSION

This patient has been seen numerous times now since the diagnosis of LAM in 2004. The presentation and treatment of her illness has been constant, the only remarkable change over the last two years is duration of time between episodes of spontaneous pneumothoraces. Beginning with months separating visits to the ICU her illness has now progressed to the point of her developing complications bi-weekly. Treatments of her condition have been continuously the same with the only variables being number of chest tubes placed and length of stay. During the most recent visit to the ICU she had five chest tubes in place for less than two days and was discharged to home care with her usual Heimlich valve with air leak in place one week post exacerbation. When not suffering complications her physical findings have been noted as: Vitals; blood pressure 110/70, pulse 86, SaO₂ 98% on room air, temperature 36.5o, and weight 113.7 pounds. Her general appearance is of a healthy young female in no acute distress who is ambulatory and can walk 6 minutes for 1360' with no desaturation or dyspnea. The HEENT findings are normocephalic and atraumatic with very small benign popular lesions across nose and cheeks. Her neck, abdomen, musculoskeletal and lab results are all normal.

Mr. Daniel Nordstrom is a Respiratory Care Student at Highline Community College in Des Moines, Washington. His paper on Pulmonary Lymphangioleiomyomatosis was chosen from 16 papers on various topics submitted to Focus for this issue. Mr Nordstrom will receive a \$100 gift certificate and a gratis registration to the 2008 Focus Conference. His school's RC Program will also receive a \$100 donation. Students are encouraged to submit their papers for the Sept/Oct issue by Sept 5th. Papers should be between 900 and 1250 words and should be submitted as MS Word files to our Craig Baker at BakerCT78@yahoo.com.

every way possible but must not be lulled into a sense of security that we become complacent. How often do we hear people ask how something could have happened? Our world is not 100% safe yet our technology and systems mislead us into thinking that it is and everything is taken care of.

What can we do?

We can be safer and reduce the risks in our everyday lives if we acknowledge and understand how our minds and their models work. The most important thing we can do is getting off autopilot when it comes to operating in areas or risk. Protocols and checklists do if fact work but we must also slow down the process and examine what you are doing. Does the phrase "take a time out" sound familiar? The Joint Commission looks for this procedure in every operating room to reduce the risk of operating on the wrong patient to cutting off the wrong leg. It is simply a break in the fast routine of everyday operations to reexamine and think. Allow yourself time for a second thought, because first thoughts are sometimes not thoughts at all. Have you heard of the acronym STOP (Stop, Think, Observe, Plan)? In diving we have a similar recipe when faced with problems and nearing a panic situation: STA (Stop, Think, Act). First Stop, get off autopilot, this may be a situation you never before experienced so don't act yet. Think, see all your options and pick the best. Act, implement your plan. I would add to this, take a slow Deep Breath, which actually helps to break the panic cycle. Your world will be safer when you are not operating on autopilot and just think how wonderful the world is when you are actually observing it. Slow down the pace. Just think how many accidents occur when you are rushed and autopilot is in high gear. When you are interrupted during a routine safety check, start over! Yes it takes time but then ask yourself how important is it and what are the consequences if something is missed. In the hyperbaric environment that cost may be extremely high. Before you close the chamber door, take a time out.

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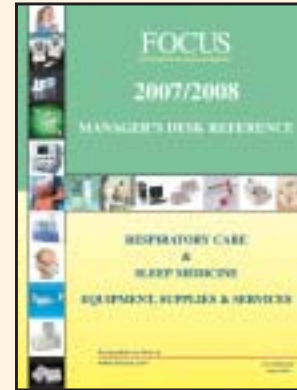
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She is a completely normal young female in appearance except for the findings when examining her chest and chest x-ray. Auscultation reveals clear breath sounds throughout with the presence of plural rub bilaterally and the Heimlich valve with an air leak located on her right chest. The chest x-ray report notes the presence of an FBO located over the medial aspect of the left hemidiaphragm. Normal mediastinum is noted in the radiology reports. Also seen has been the presence of multiple small pulmonary cysts, bilaterally, renal and liver lesions, and multiple sclerotic bone lesions, all consistent with TS and LAM. A stable right pneumothorax with thoracotomy tube is in proper placement for treatment of persistent pneumothorax on the right side.

Treatment options for this patient are limited. "Without lung transplant, there is a 50-80% 5-year survival rate". Since LAM primarily occurs in child bearing aged women it is thought that hormonal stimulus affects the disease process. Treatments with; progesterone, oophorectomy, tamoxifen, gonadotropin-releasing hormone (GnRH) agonists, and androgen therapy have been used to diminish estrogens effects with varying results and unwanted side effects. Treatment with doxycycline may have little side effects and sirolimus is being tested as a treatment for LAM.

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I was not being deliberately foolhardy--I respect my co-workers greatly and made preparations to use bi-PAP temporarily and re-intubate quickly if necessary. To justify my decision to myself and to others, I had to integrate many pieces of unquantifiable data and weigh their influence against that of an accepted numeric index of 'weanability'. I was aware that rapid shallow breathing is inherent to ambulatory patients with restrictive (low compliance) diseases of the chest and to those with reflexes triggered by lung parenchymal disorders. Moreover, the RSBI rises predictably when ventilatory requirements increase--even in healthy exercising normal individuals. The patient had good cardiac reserve, appeared alert, and although working noticeably to breathe, did not appear in marked distress, despite his frighteningly high RSBI. His minute ventilation did not fall when on low level pressure support, and there was noticeable variation in the depth and frequency of breathing. Perhaps most convincingly, he had only a modest secretion load and seemed to exhibit some strength reserve when his cough reflex was stimulated by the suction catheter. Finally, I explained as best I could what adverse events might happen when we removed the tube, and yet the patient remained eager to try. The "gleam in the eye" and breadth of the smile were hard to score on the protocol sheet.

Widespread enthusiasm for protocolized ventilator care reflects its generally positive impact. If thoughtfully designed and implemented, protocols expedite quality care. even Even when unsuccessful identify those patients whose management details need closer attention. Many of the toughest problems, however, seem to yield only to experience and a reasoned analytical approach that integrates clues from a variety of sources. Some of these signals defy facile coding into rules and care directives. In these all-too-frequent instances, the rules must and should be broken.

Dr. Marini, MD, Professor of Medicine at the Univ of Minnesota, is a clinician-scientist whose investigative work has concentrated in the cardiopulmonary physiology and management of acute respiratory failure. In the majority of his research, he has been positioned at the interface between basic physiology and clinical medicine so as to develop insights into advancing clinical practice.