ASTHMA & ZEBRAS

4 Pediatric Case Studies of
Uncommon Pulmonary Conditions

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“ASTHMA AND ZEBRAS”

Speaker Disclosure: Dr. Benjamin Francisco has no relationship with any commercial firm having products related to topics discussed at this conference. Actual disclosure forms are available upon request.

BACKGROUND

- Cases were seen in a pediatric outpatient clinic, pulmonary medicine & allergy, over the last decade, 95% asthma visits
- High volume asthma practice serving a large rural referral area in mid-Missouri
- Collaborative practice – four pulmonologists, two pediatric nurse practitioners
- Full compliment of subspecialists and primary care providers
- Free-standing Women’s and Children Hospital
UNCOMMON (ZEBRA) VS ASTHMA (HORSE)

I THOUGHT IT WAS JUST ASTHMA, IT USED TO BE ASTHMA, THIS IS WAY MORE THAN ASTHMA, THIS DOESN'T ACT LIKE ASTHMA AT ALL

RECOGNIZING THE UNCOMMON PATTERN

HYBRIDS/CROSSES
PERSISTENT COUGH, UNRESPONSIVE PNEUMONIA

- 15 year old, longstanding moderate persistent asthma
- New onset chronic cough (3 months)

HISTORY & PRESENTATION

- Routine follow-up, previously well controlled asthma
- Step 4, medium dose ICS/LABA, not well controlled, ACT= 16
- Increased coughing for 3 months since a winter “cold” with fever
- Not much improvement with albuterol
- Afebrile, itchy macules on legs and feet, HCZ helps

FINDINGS & INITIAL MANAGEMENT

- Chest clear to auscultation
- CXR – pneumonia, superior segment, right lower lobe
- Azythromycin
- Advanced to step 5, asthma therapy
- Scheduled albuterol for a week
TREATMENT RESPONSE, ADDITIONAL FINDINGS

- Phone follow-up: no improvement at 2 weeks
- Started a 10 day course of cefuroxime
- Scheduled follow-up with repeat CXR
- CXR unchanged, lost 5 pounds, left 3 cm axillary lymph node
- Chest CT - Left supravacular, axillary, mediastinal, bilateral hilar and upper abdominal lymphadenopathy. Multiple bilateral pulmonary nodules/masses, largest in the right lung. Hepatosplenomegaly with multiple ill-defined splenic and hepatic hypo dense lesions. Differential diagnosis based on the constellation of findings is highly suspicious for lymphoma/leukemia. Another consideration is atypical TB mycobacterial infection or fungal infection

OVERVIEW OF THE PROBLEM, INCIDENCE

- Hodgkin’s Lymphoma (stage IVA, nodular sclerosing)
- 7% of childhood cancers, most common cancer in teens 15-19
- Initially painless, rapidly growing lymphadenopathy of cervical, supravacular, or axillary chains, mediastinal mass
- Lifetime incidence of 2 per 1000 (2009-2011 data)
- In 2011, 185,793 people in the US were living with Hodgkin’s
- 5 year survival rate 85.3% (2004-2010)
WHAT WERE THE RED FLAGS?

• Unresponsive cough, previously well-controlled asthma
• Unresponsive pneumonia
• Weight loss of >5 pounds over 30 days in an athletic male
• Enlarged axillary lymph node

THE PATTERN

OUTCOME

• After completing Children’s Oncology Group protocol AHOD0031 between May and August of 2014, he is in remission
• Course was complicated by subclavian thrombus and bilateral pulmonary emboli. He was asymptomatic.
• Non-surgical management was successful.
• He feels well and wants to get back to his high school routines, including wrestling.
ACKNOWLEDGEMENTS & REFERENCES

- Dr. Zarah Ner, Pediatric Pulmonology Medicine & Sleep
- Dr. Michael Cooperstock, Pediatric Infectious Disease
- Dr. Barbara Gruener, Pediatric Hematology/Oncology


ACUTE ONSET PERSISTENT COUGH

HISTORY & PRESENTATION

- New patient, 10 year old male with reported allergic rhinitis and intermittent asthma with acute onset of persistent cough 2 weeks ago not associated with a cold or fever. Unresponsive to Robitussin and albuterol MDI. Cough occurs day and night. One episode of post-tussive emesis. Coughing associated with occasional problem with swallowing.
FINDINGS & INITIAL MANAGEMENT

- Unable/unwilling to perform forced expiratory maneuver
- Physical exam was normal. Patient noted to cough intermittently during exam. Dry, non-productive cough.
- CXR ordered

LOOK CLOSELY, LEFT MAINSTEM BRONCHUS

A LINEAR METALLIC DENSITY OVER PROXIMAL LEFT MAINSTEM, CONSISTENT WITH ASPIRATED FOREIGN BODY. LEFT PERIHILAR RETROCARDIAC STREAKY OPACITIES.
TREATMENT RESPONSE, ADDITIONAL FINDINGS

- Direct laryngoscopy, rigid bronchoscopy, removal of FB
- Left lung collapsed mid-procedure, FB acted as ball-valve
- Flexible scope and catheter were used to pass below the FB to re-inflate the left lung
- Sewing needle/dart, granulation
- Purulent material below carina, left
- PICU, Augmentin

OVERVIEW OF THE PROBLEM, PREVALENCE

- More than 3700 deaths due to FBA in 2007
- 80% involve children <3 years of age
- 90% hx of acute choking, fib coughing, wheezing or stridor
- Most FBs are radiolucent, organic, peanut most common
- Prior to age 15 right and left mainstem bronchi at equal risk
- Obstructive emphysema (62%), mediastinal shift (55%), pneumonia (26%), atelectasis (18%), radiopaque objects (3%)

WHAT WERE THE RED FLAGS?
THE PATTERN

- Acute onset cough of 2 weeks duration w/o URI
- Unresponsive to albuterol, anti-tussive
- Associated trouble swallowing
- Refused forced expiratory maneuver (spirometry)
- HE IS A 10 Y.O. BOY!
- HE HAS A BROTHER!
- BOYS DO CRAZY STUFF!

OUTCOME

- Normal CXR at one week follow-up!
- Now planning the next adventure…

ACKNOWLEDGEMENTS & REFERENCES

- Dr. Jim Acton, Pediatric Pulmonology
- Dr. Eliav Gov-Ari, Pediatric Ear, Nose and Throat


TEEN - DYSPNEA WITH EXERTION

HISTORY & PRESENTATION
• 13 year old with known asthma lost to follow-up for 18 months
• Marked worsening pattern of dyspnea with exertion, shortness of breath at rest, wheezing
• FVC only 60% of predicted, mild obstruction FEV1/FVC = .80/.82, 6% increase in FEV1 (1.95 L) after albuterol
• I/E wheezing, best heard posteriorly and centrally, harsh and prolonged inspiration, BP – 138/86
• CXR – radiologist phoned for CT w/contrast, distal trachea and bronchi are markedly attenuated

FINDINGS & INITIAL MANAGEMENT
• CT - A SOFT TISSUE MASS OCCUPIES THE MIDDLE MEDIASTINUM EXTENDING FROM THE THORACIC INLET INFERIORLY TO THE LEVEL OF THE HEART 3 CM BELOW THE CARINA WITH ATTENUATION OF THE AIRWAYS AND PULMONARY VEINS AS WELL. MARKED NARROWING OF THE TRACHEA JUST ABOVE THE CARINA. THE RIGHT MAIN STEM BRONCHUS IS ALMOST COMPLETELY OBLITERATED AS IS THE RIGHT PULMONARY ARTERY. THERE IS RIGHT MIDDLE LOBE ATELECTASIS AND RIGHT POSTERIORLY LOCATED GRANULOMA. THERE IS DECREASED LUNG PERFUSION - RIGHT SIDE DIMINISHED MORE THAN LEFT.
IMPRESSION: FINDINGS ARE CONSISTENT WITH FIBROSING MEDIASTINITIS WITH ATTENUATION OF THE AIRWAYS, THE PULMONARY ARTERIES AND SUPERIOR VENACAVA.
MARKED NARROWING OF THE TRACHEA

TREATMENT RESPONSE, ADDITIONAL FINDINGS
- Prednisone 30 MG po bid, itraconazole 100 mg po bid, albuterol Q4 prn, NO vigorous physical activity
- Over a 3 year tx period 6 CTs remained essentially unchanged
- IMPRESSION: the mediastinal adenopathy with narrowing of the trachea, right and left mainstem bronchus and right and left pulmonary arteries is unchanged the parenchymal findings are unchanged as well
- He developed osteopenia (DEXA scan z-score= -3.9) after almost 2 years of daily prednisone, Tx – Vit. D and calcium
- Required tx for hypertension
OVERVIEW OF THE PROBLEM, PREVALENCE

- Fibrosing mediastinitis is a rare, severe complication of histoplasmosis
- Infection occurs in childhood in endemic areas (Mississippi and Ohio River basins). FM usually begins between 20-40 YOA
- FM is an abnormal immunologic response to a soil-based fungus
- Invasive, calcified fibrosis of lymph nodes occluding major vessels or airways
- Use of anti-fungals is not proven, but usually part of tx
- Primary interventions address obstruction of vessels & airways

WHAT WERE THE RED FLAGS?

- Worsening dyspnea with exertion, disabling
- Shortness of breath at rest for months
- Harsh, prolonged inspiration suggesting upper airway obstruction
- 35% drop in FVC with only mild decrease in FEV1/FVC ratio

THE PATTERN

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OUTCOME

• At last follow-up 4 years after diagnosis he had returned to normal activity with some exertional wheezing he did not treat.
• His FVC was 103% predicted (4.03 L), FEV1/FVC = .81
• Maintenance medications included: itraconoczole 200 mg Q am and 100 mg HS, fluticasone 110 mcg 2 puffs BID by Aerochamber, albuterol MDI 2 puffs Q4 prn
• We referred him to Vanderbilt for evaluation and treatment. He moved out of state and was lost to follow-up.

ACKNOWLEDGEMENTS & REFERENCES

• Dr. Peter Konig, Pediatric Pulmonology
• Dr. Zarah Ner, Pediatric Pulmonology
• Dr. James Brown, Pediatric Radiology
• Dr. Ted Groshong, Pediatric Nephrology
• Dr. Burt Bachrach, Pediatric Endocrinology


HISTORY & PRESENTATION

• 17 year old female with longstanding severe persistent asthma, step 5 therapy, poor adherence and variable control, overweight, HbgA1c= 6.2, GERD managed with PPI
• Sick visit with cough and sternal pain over the preceding week
• CXR - Small plates of atelectasis in the left mid to lower lung field. Vague streaky density in the right lower lobe.
• Spirometry – 35% decrease in baseline FVC (post albuterol), diffuse lung disease???

FINDINGS & INITIAL MANAGEMENT

• Admitted with status asthmaticus & pneumonia
• IgE= 4953, Aspergillus Fumigatus = 28.1 (high)
• Discharged on prednisone 30 mg po BID, cefuroxime
• Baseline DEXA scan normal, began long term course of methylprednisolone for allergic bronchopulmonary aspergillosis
• CXR 90 days - Interval resolution of right basilar atelectasis/pneumonia and left sub-segmental atelectasis
• CT – normal (no bronchiectasis)

TREATMENT RESPONSE, ADDITIONAL FINDINGS

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OVERVIEW OF THE PROBLEM, PREVALENCE

- Allergic bronchopulmonary aspergillosis (ABPA)
- Almost exclusively in patients w/asthma, also cystic fibrosis
- A hypersensitivity reaction to Aspergillus (usually A. fumigatus)
- Sx – persistent cough, green or brown sputum, asthma exac.
- Lab- Aspergillus skin testing, total IgE levels, circulating precipitins, and A. fumigatus–specific antibodies
- Sputum positive for A. fumigatus, peripheral eosinophilia >1000
- Prevalence up to 13% of adult asthmatics, screen by prick test

WHAT WERE THE RED FLAGS?

- Acute cough, diminished breath sounds
- Precipitous drop in forced vital capacity
- CXR findings supporting diffuse disease
- Afebrile
- Chest pain
- Aggressive asthma therapy

THE PATTERN

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- Aggressive asthma therapy
OUTCOME

• ABPA in remission, IgE has remained below 700, 18 months
• Insulin-dependent diabetic
• Became pregnant 6 months after remission
• Mom and baby are doing well, expecting any day

ACKNOWLEDGEMENTS & REFERENCES

• Dr. Jim Acton, Pediatric Pulmonology
• Dr. Christopher Blue, Pediatric Endocrinology
• Dr. Hung Winn, Obstetrics


ABPA. Merck Manual, accessed 2/15/15
http://www.merckmanuals.com/professional/pulmonary_disorders/asthma_and_related_disorders/allergic_bronchopulmonary_aspergillosis_abpa.html

LESSONS LEARNED

• Some children & teens with severe impairment try to “carry on” ignoring their suffering and disability, their history doesn’t tell all
• Embarrassment or shame prevents people from disclosing what they know is source of their problem
• People sometimes achieve better outcomes than physical evidence supports, we should give it our best shot
• We should pay greater attention to FVC over time in patients with asthma, deficits point to risk of other pulmonary disease
• Side effects of potent therapies are sometimes worth the risk.
AS YOU GO BACK TO CARING FOR COMMON PROBLEMS...

REMEMBER TO LOOK FOR ZEBRAS

THE END

HERE’S A SHOUT OUT TO THE AMAZING YOUNG PEOPLE AND THEIR FAMILIES WHO THROUGH THEIR SUFFERING TAUGHT US ABOUT COURAGE AND BEING MORE VIGILANT IN CARING FOR OUR PULMONARY PATIENTS