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Pulmonology Second Opinion - Full Chart Review

Date: 2022-01-19

Patient: Jane Doe

Discussion:

January 18th 2017

Pulmonary Second Opinion- Jane Doe

Reason for Consultation: Suggest treatment plan and pain management.

HPI:

Jane Doe is a 25-year-old female from Bangladesh who presented, presumably at the end of September 2016, with a high fever during six days that was diagnosed as enteric fever. This was accompanied by a pruritic maculopapular rash on her shins and swelling of both ankles and followed by sequential peripheral facial nerve paralysis (around 11/10/2016; initially right, then left) that resolved within a month. Fever did not recur. She also complained of headaches.

She was seen by ophthalmology, presumably for visual abnormalities and was diagnosed with bilateral anterior uveitis. She was also found to have mediastinal lymphadenopathy in a CT chest.

She took five days of prednisolone from 9/10/16 to 13/10/16 and received an antibiotic course consisting of 2 g ceftriaxone and azithromycin 500 mg. (unknown duration or dates)

PPD was negative at 72 hours.

She was seen by a neurologist, Dr. Derek Soon, a pulmonologist, Dr. KC See and an ophthalmologist, Dr. Clement Tan.

Past medical history:

Asthma

Parotid enlargement (date?)

Allergies: Steroids (face swelling)

Laboratory results:

Pertinent negatives: HCV-Ab., HBVs-Ag, ANA, Anti-phospholipid Ab., p-ANCA, c-ANCA, anti-SSA, Anti-SSB, Anti-RNP, Anti Scl-70, Anti-Jo1TSH, Free T4, cortisol. Both CRP and ESR are normal. CBC, BMP, calcium, LFTs and lipid panel are essentially normal (there was a mild elevation of transaminases in 2010). HIV and treponemal testing were suggested, but I did not receive laboratory confirmation.

Although reference values are not provided, ACE level seems to be mildly elevated (73 U/L)

Imaging:

â€¢ CXR: two CXR images were submitted, but due to poor image quality it is not possible to comment on the results.
â€¢ CT chest (24/10/2016): (as per report-images not provided) mediastinal lymphadenopathy-right paratracheal and pre-carinal. Lymph node size not provided. No abnormalities in lung parenchyma, heart or great vessels.
â€¢ MRI Brain (23/10/2016): No significant intracranial abnormality is seen. Mucus retention cyst is detected within the right maxillary sinus and mild mucosal thickening in the bilateral ethmoidal air cells suggestive of sinusitis. Subtle enhancement in the IAC portion of the left facial nerve is consistent with Bell's palsy. No parotid or leptomeningeal involvement.
â€¢ Ultrasound Abdomen: minimal fatty change of the liver (as per Dr. Kansal's note- report not provided)

Specialty studies:

-Nerve conduction studies (NCS) were done in Bangladesh and repeated in Singapore. As per Dr. Soon's note, these did not show evidence of peripheral nerve involvement. However, an unsigned case summary from Birdem hospital states that an NCS of four limbs and facial nerves performed on 20/10/2016 is consistent with predominantly motor polyradiculoneuropathy that is axonopathic in nature, compatible with AMAN or other motor polyradiculoneuropathies.

Physical Examination description:

Height: 5 feet 4 inches; Weight 61 Kg
HR 104 bpm, BP 100/70 mmHg (on 25/10/2016)
No generalized lymphadenopathy
Bilateral facial palsy
Weakness of the left flexor muscles of the hand

Diminished knee and ankle deep tendon reflexes
No parotid enlargement

Assessment

Nafisa is a 25-year-old female with a PMH of asthma and parotid enlargement who presented acutely in September 2016 with fever (enteric fever?) followed by the development of cutaneous maculopapular shin lesions, sequential peripheral facial paralysis, and bilateral uveitis. A CT chest revealed the presence of mediastinal lymphadenopathy (paratracheal and pre-carinal) but no parenchymal lung disease. Her physicians have entertained the possibility of sarcoidosis, vasculitis, and tuberculosis.

It is possible that her cutaneous lesions did correspond to erythema nodosum (although a more thorough description is not available). The combination of fever, erythema nodosum, facial paralysis, bilateral uveitis and mediastinal lymphadenopathy is very suggestive of sarcoidosis. Additionally, there is a history of parotid enlargement for which she received steroids and ACE is mildly elevated, also supporting this possibility. Her presentation could correspond to a Heerfordt's syndrome (uveoparotid syndrome) and/or Lofgren's syndrome (erythema nodosum, hilar adenopathy, migratory polyarthralgia, and fever).

There are contradictory statements in her documents regarding the results of her nerve conductions studies. This should be clarified. However, in her summary it is stated that the NCS were consistent with acute motor axononeuropthy (AMAN). AMAN is frequently associated with Campylobacter infection and could have been related to her enteric fever a month prior. AMAN can also be a manifestation of sarcoidosis. Of note, Campylobacter infection can also present with erythema nodosum and reactive arthritis (ankle swelling in this patient).

Although tuberculosis and vasculitis could rarely present in this manner, workup has been negative for both of these conditions and no further workup is necessary at this time unless further signs or symptoms suggestive of these conditions develop. These could include but are not limited to, respiratory symptoms, cavitary or miliary pulmonary findings and pleural effusions in the case of tuberculosis or purpura, renal, neural or other organ involvement in that of vasculitis.
Additional causes of mediastinal lymphadenopathy include other infectious etiologies such as mycobacterial, fungal or viral disease (Epstein Barr, CMV, HIV) and malignant neoplasms, especially lymphomas or metastattic disease (breast, thymoma, Kaposi). Rheumatologic diseases are very unlikely given normal ESR and CRP and an atypical cliniacl presentation. An extensive workup was negative; However Rheumatoid factor (RF) and Anti-Sm was not included in the provided documents.

It is unclear whether the patient's symptoms have resolved. There is a suggestion of persistent pain in the request for consultation, but no description of the location or type of pain has been provided.

Recommendations:

Although the possibility of sarcoidosis seems likely, the diagnosis does not appear to have been formally made from the records provided. Biopsy of an easily accessible affected site would be the first step. Skin lesions (except lesions compatible with erythema nodosum that typically do not show granulomas) are the most easily biopsied sites. A close re-examination of the skin, especially around scars or tattoos should be undertaken to detect potential areas for biopsy. If the parotid or lacrimal glands are affected or if she has developed lymphadenopathy on other sites, these could also be biopsied. However, from the case description, it does not appear that she presented signs of involvement of these areas in October. Re-examination is appropriate at this time to prevent a more invasive diagnostic procedure. Also consider the possibility of nasal mucosa biopsy given the presence of sinusitis.

Because almost 3 months have elapsed since her last CT chest, it is appropriate at this time to perform a follow-up CT chest. This will determine whether the mediastinal lymphadenopathy is still present and rule out a transient infection as a potential cause if they have resolved. In the case of persistence, it is imperative to rule out other important causes of lymphadenopathy, especially lymphoma but also other metastatic malignancies (breast, thymic). Pathologic examination can reveal the presence of other rare pathologies such as amyloidosis, the presence of microorganisms such as mycobacteriae and fungi or caseating granulomas. In that case, Endobronchial Ultrasound (EBUS) guided transbronchial needle aspiration of the mediastinal lymph nodes is recommended. Alternatively, blind transbronchial needle aspiration can be attempted, but negative results need to be interpreted with caution as sensitivity decreases significantly using a blind technique. Finally, mediastinoscopy is another alternative but more invasive in nature.
EBUS needle aspiration also has an 80-90 % yield in the diagnosis of sarcoidosis. The presence of non-caseating granulomas would confirm the suspected diagnosis.

Although Campylobacter infection could explain many of the symptoms exhibited by this patient, it would not reasonably account for the development of mediastinal lymphadenopathy. Also, other than serologic tests which are not of routine practice, it is too late for conventional diagnostic tests (cultures). Furthermore, the results would not be useful clinically as they would not influence management.

Because it is possible that the patient has sarcoidosis, it is imperative to rule out cardiac involvement, and an ECG should be performed, if not already done. Of note, the patient was tachycardic in October. Further workup will depend on the ECG result and the presence of other symptoms consistent with cardiac involvement.

Additionally, an allergy to steroids has been reported. However, she took five days of prednisolone without adverse reaction. Because allergy to steroids is extremely rare and steroids are the mainstay treatment for sarcoidosis, an allergy consultation to determine whether this is a true allergy is recommended.

If the patient is diagnosed with sarcoidosis, she will need periodic follow-up with chest X-ray to screen for pulmonary parenchymal involvement, which occurs in about 80-90% of cases and could require more aggressive management. Pulmonary function tests with lung volumes and DLCO should be obtained as a baseline for future comparison.

Currently, the patient does not appear to require the use of steroids from the history provided and conservative management and follow-up observation is recommended. The most common indication for prednisone treatment is symptomatic pulmonary involvement, which this patient does not have. However, severe extrapulmonary manifestations that could lead to permanent organ damage such as vision loss, cardiac involvement (and risk of life-threatening arrhythmias), renal or neurologic involvement can also indicate the need for treatment with prednisone. In Nafisa's case, ocular involvement needs to be followed by her ophthalmologist who will determine its severity and appropriate treatment. Similarly, severely symptomatic patients suffering from fatigue, severe arthralgia, weakness or fever may also benefit from steroid therapy. However, the long-term side effects of steroids must be weighed against the potential benefits. I am not aware that Nafisa has any of these indications.

Summary of recommendations:

- Re-examination of the skin, lymph nodes, parotids and lacrimal glands; if affected, proceed with biopsy of these sites. Also, consider nasal mucosal biopsy.
- Repeat CT chest. If persistent mediastinal lymphadenopathy, consider EBUS guided needle aspiration.
- ECG
- HIV serology
- RF
- PFT with lung volumes and DLCO
-Allergy consultation regarding possible allergy to steroids

Questions:

Note:

Several notes recommended tissue biopsy, including skin, parotid and lymph nodes. However, no indication that these have been performed was submitted with her documents. I have also recommended performing an ECG , RFand HIV serologies which have likely been already done but not included in her medical record. If some or all of these results become available, I will be happy to add an addendum to my note to include them. Similarly, a request was made to provide advice on pain management. Further detailed characterization of her pain would be necessary including localization, duration, and type.

Evolution of the patient's symptoms from October to present can also be very useful.

Thank you for the opportunity to participate in Nafisa's care. Please do not hesitate to contact me with further questions through Second Opinions.

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