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Pleural Effusion in A Patient with Fahr's Disease

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Abstract

Background *Pleural effusion can be associated with a number of diseases. Here is a rare case report of a patient of pleural effusion associated with Fahr's disease.*

Introduction *Pleural effusion is a common disease of pleura characterised by excessive accumulation of pleural fluid in between parietal and visceral pleura. It can occur due to various reasons and can be broadly classified into exudative and transudative type. On the other hand, Fahr's disease is a very rare, dominantly inherited or sporadic neurodegenerative disorder characterised by idiopathic calcification of basal ganglia.*

Case Presentation *A 22 year old male presented to Chest OPD of Combined District Hospital, Shravasti with chief complaints of difficulty in breathing, dry cough, fever and right sided chest pain for 15 days. He was also having motor difficulty, gait disorder, seizure disorder and neuropsychiatric symptoms for one year for which he took medications for few months and then stopped. On auscultation, there was decreased breath sound on right side. Chest radiograph showed positive meniscus sign on right side and Non-contrast CT head showed bilateral symmetrical calcification of basal ganglia and cerebellum suggestive of Fahr's disease. Pleural fluid analysis showed raised ADA and presence of coagulum. Hence, a final diagnosis of Right sided Pleural Effusion (Tubercular) with Fahr's disease was made. 600 ml clear transparent was aspirated therapeutically under USG guidance and ATT was started under CAT1 DOTS. He was also advised anti-epileptic drugs and atypical anti-psychotics for seizure control and to improve his neuropsychiatric symptoms and referred to neurologist for further management.*

Conclusion *Every case of pleural effusion must be thoroughly investigated to know the underlying cause and to start the specific treatment. Although, a temporal association between pleural effusion and Fahr's disease could not be established in this case, yet causal relationship can not be completely overruled. Further study is warranted in this direction.*

Key words *Pleural effusion, Fahr's disease*

Background

Though the most common cause of pleural effusion is Pulmonary Tuberculosis in India **[**1**]**, yet association with rare diseases is also found. Here is a brief case report of a patient of pleural effusion associated with Fahr's disease.

Introduction

Pleural effusion is accumulation of excessive fluid in pleural space that occurs when pathological processes cause an imbalance of the hydrogen pressure gradient, capillary membrane permeability, and lymphatic capacity leading to poor protein metabolism or inflammatory exudates.**[**2**]**

Under normal conditions, pleural fluid is secreted by the parietal pleural capillaries at a rate of 0.6 mililitre per kilogram body weight per hour, and is cleared by lymphatic absorption leaving behind only 5-15 militres of fluid which helps to maintain a functional vacuum between parietal and visceral pleura **[**3**]**. Pathophysiologically, pleural effusion can be transudative and exudative. If a pleural effusion is mainly due to increased hydrostatic pressure, they are usually transudative. Increased mesothelial and capillary permeability or impaired lymphatic drainage usually causes exudates. **[**4,5**]**

Whereas, Fahr's disease is a rare, genetically dominant, neurodegenerative condition characterized by idiopathic calcification of the basal ganglia.**[**6**]** The pathogenesis is not fully known, but it is stipulated that it may be secondary to the impairment of the blood brain barrier or to a neuronal calcium phosphoric metaboliosm disorder. **[**7**]** It typically affects individuals in the 3rd and 4th decade of their lives8 with a prevalence of less than 1/1000000 population. It was first described by German neurologist Karl Theodor Fahr in 1930.**[**9-12**]**

Case Presentation

Chief complaints

On 26th February, 2022, a 22 years old male, accompanied by his father, presented to Chest OPD of Combined District Hospital, Shravasti with chief complaints of difficulty in breathing, dry cough, fever and right sided chest pain for 15 days.

History of present illness

The patient was asymptomatic a year ago when he first experienced walking in straight line. Subsequently, he developed difficulty in learning and memory and then developed multiple episodes of seizure attacks after which he consulted a neurophysician in Lucknow. He was then started on anti-epileptic drugs which he was taking for 3 months but stopped 15 days ago when he developed right sided chest pain, difficulty in breathing, fever and dry cough for which he came to Chest OPD.

Personal History

The patient is vegetarian, non-alcoholic and non-smoker.

Examination

There was dull note on percussion over interscapular and infrasacpular areas. On auscultation, there was decreased breath sound on right interscapular and infrascapular region while left side was clear with no added sound.

Neurological examination revealed movement disorder, gait disorder, cognitive impairment and speech disorder. He was unable to execute rapid alternating movements like pronation and supination of the forearm properly suggesting cerebellar lesion.

Radiological investigation

Chest X ray PA view(digital) revealed blunting of costophrenic angle and positive meniscus sign on right side and mediastinal shift towards left side.

NCCT head revealed bilateral symmetrical basal ganglia and cerebellar calcification suggestive of Fahr's disease.

Blood investigation

Total leukocyte count was raised (12,432 cells per cubic millimitre) with 84% neutrophils. Serum Protein was 6.7g/dL. Serum calcium was 7.1 mg/dL. Liver function tests and Kidney function tests were within normal range. Viral markers (HIV-I & II and HBsAg) were non-reactive.

Pleural fluid examination

10 ml of clear straw-coloured pleural fluid was aspirated from right side of the chest through 7th intercostal space mid-axillary line for and sent for examination. Pleural fluid examination results were-

Adenosine Deaminase (ADA) – 51.28 U/L

Protein- 3.41g/dL

Neutrophils- 35%

Lymphocytes- 60%

Ziehl-Neelson Stain- Acid Fast Bacilli not seen.

Coagulum- present

Cytology for malignancy- No malignant cell seen.

Final Diagnosis

The results of above investigations were in favour of exudative pleural effusion. So, a diagnosis of Right sided Pleural Effusion (Tubercular) with Fahr's disease was made.

Treatment

After taking valid consent, Therapeutic Pleural Aspiration was performed under USG guidance and nearly 600 ml of pleural fluid was aspirated from right side seventh intercostal space mid-axillary line after infiltrating 4 ml of 2% lignocaine through the same space. The patient was relieved of breathlessness and subsequent chest x ray showed lung expansion. Subsequently, patient was registered on NIKSHAY and

anti-tubercular therapy under Category 1 DOTS was initiated. The patient was also given supportive treatment for fever and cough.

The patient was also advised to take his anti-epileptic drugs previously prescribed by the neurologist namely-

Tab Clobazam 10mg BD

Tab Divalproex 500 mg BD

Tab Calcium 500 mg BD

Vitamin D3 60k units once weekly

The patient was further advised to consult his neurologist as he was experiencing neuropsychiatric symptoms after stopping his prescription drugs. It may be noted that the diagnosis of Fahr's disease was made at our hospital.

Since the patient refused admission, he was managed on OPD basis over 5 days.

Follow Up

The patient did not turn up for follow up, so follow up records are not available at the time of writing this article. However, patient has been contacted telephonically to come for follow up.

Discussion

Pleural effusion is a common disease of pleura characterised by pathological accumulation of excessive pleural fluid between parietal pleura and visceral pleura. The most common symptom of pleural effusion is dyspnea, severity of which is only loosely correlated with the size of effusion. Some patients also complain of dry cough, as was seen in this case, due to manifestation of pleural inflammation or lung compression due to effusion. Pleural effusions can also impair the quality of sleep.

Chest X ray PA view (Digital) is a routine radiological investigation done to detect pleural effusion although it is the not the most sensitive one. On an upright posteroanterior (PA) view, a minimum of 200 ml of fluid is required to obliterate the costophrenic angle, called the meniscus sign of a pleural effusion.

However, in a lateral decubitus view, 50 ml of fluid can be diagnosed with this sign. All unilateral effusion in adults needs thoracentesis to determine the cause of pleural fluid. Ultrasonography and Computed Tomography are far more sensitive in detecting small amount of effusions.**[**15,16**]**

Treatment of pleural effusion depends on underlying cause. Light's criteria are used to determine whether a pleural effusion is exudative or transudative. Satisfying any one criterium means it is exudative:

1. Pleural fluid Protein/ Serum Total Protein ratio > 0.5
2. Pleural lactate dehydrogenase/Serum lactate dehydrogenase > 0.6
3. Pleural LDH > 2/3 upper limit of serum LDH (lab reference value)

If a patient meets Light's Criteria of exudative effusion but has a clinical appearance suggestive of a transudative effusion, Dr. Light recommends a serum albumin – pleural albumin < 1.2 mg/dL, to confirm the effusion is exudative. Pleural fluid Adenosine Deaminase aids in diagnosing tuberculosis as raised ADA more than 40 IU/L is suggestive of exudative effusion, tuberculosis in particular as was seen in this case. **[**13,14**]**

Common causes of Transudative pleural effusion are congestive heart failure, cancer and pulmonary embolism while Exudative pleural effusion can occur due to Tuberculosis, which remains the most common cause of Pleural effusion in India, pneumonia, pulmonary embolism and Systemic Lupus Erythematosus.**[**17**]**

Bilateral Striopallido Dentate Calcinosis or Fahr's disease is a very rare neurodegenerative disorder characterised by bilateral symmetrical intracranial calcification.**[**18**]** The most common presentations as per the Fahr's Disease Registry are movement disorders (55%). Other neurological symptoms include cognitive impairment, cerebellar signs, psychiatry symptoms, gait disorders and sensory changes some of which were present in this case.**[**19**]**

The disorders of calcium metabolism may occur in association with the intracerebral calcification. Other causes of the intracranial calcification include infectious diseases like Toxoplasmosis and Syphilis and inflammatory illnesses like SLE. But Fahr's disease represents a heterogeneous group of disorders that are not associated with any known disorder of the calcium metabolism.**[**20**]**

Genetic studies have shown an autosomal dominant inheritance in the familial cases.**[**21**]** The imaging findings of the symmetric and extensive calcification of basal ganglia are usually typical, as was seen in this case. Cerebellar calcification explains presence of cerebellar signs (inability to do rapid alternating

movements like pronation and supination of forearm) in this case. Computed Tomography remains the most effective screening tool.[22]

Fahr's disease is a progressive disease with no definite cure. The treatment is mainly symptomatic that may include anti-epileptic medications, atypical antipsychotics, SSRIs, pain killers, calcium and vitamin D supplementation.[23]

Conclusion

Though, the patients with pleural effusion are commonly seen in chest clinics, every case should be thoroughly investigated to know the underlying cause. In this case, Fahr's disease precedes pleural effusion but without cause effect relationship, yet possibility of causal relationship cannot be overruled completely. Further study is warranted in this direction.

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