A case study on Contarini’s syndrome: A rare case due to autoimmune disorders

Sir,

Bilateral pleural effusions are usually caused by a single factor, such as congestive cardiac failure or malignant diseases. However, Contarini’s syndrome is an exceptional case characterized by bilateral pleural effusion of different etiologies with different fluid characteristics of the lungs. In this case, bilateral diagnostic thoracentesis is performed because the patient is devoid of any underlying causes.

A 68-year-old female patient was admitted to the hospital with C/O breathlessness for 2 weeks (mMRC Grade 2), weight loss, and loss of appetite over the previous 3 months. She had a low-grade history of fever 2 days ago. She was already a known case of rheumatoid arthritis in 2007 and was initially treated with indomethacin, once weekly methotrexate, and prednisolone for 2 years. In view of left sided macular retinopathy, maintained hydroxychloroquine 200 mg OD for three years before discontinuing, and maintained wysolone 2 mg until one week before admission.

On examination, she is conscious, oriented, and tired. She had tachycardia, but her other vitals were normal. Pallor and bilateral pitting edema are evident. The first arterial blood gases revealed primary respiratory alkalosis with subsequent metabolic alkalosis. A chest X-ray (CXR) reveals left-sided costophrenic angle blunting with heterogeneous opacity in the left mid and lower zones.

Laboratory tests revealed leukocytosis (12820), elevated infective markers (CRP 142 and ESR 106), hypoalbuminemia (1.8), dyselectrolytemia, and proteinuria (urine protein 3+) with an ANA strong positive >3200-mixed pattern. Positive SSA, ribosome P, and AMA-M2 markers are seen in the ENA profile (in 2007, positive for SSA, Ro-52KD, and Jo-1). Antibiotics were given to her (injection Piperacillin/tazobactam, injection amikacin, and tablet azithromycin).

A diagnostic thoracentesis was performed, and pleural fluid analysis revealed exudative fluid with low glucose (36), high LDH 1720, and low ADA (21) levels, with neutrophilic predominance. Atypical cells were not found in cytology (repeat). On December 29, 2020, an intercostal drainage tube was placed in the left sixth intercostal space, and postprocedure vitals were stable. On December 31, 2020, a bronchoscopy revealed no mucosal/endobronchial lesions, and the bronchoalveolar lavage submitted for examination was inconclusive. Bacterial cultures were negative. All the specimens tested negative for GeneXpert.

A repeat left-sided pleural fluid examination revealed exudative fluid with lymphocytic predominance, as well as glucose (121) and LDH (254).

CECT chest reveals a well-defined intrapulmonary fluid collection with peripheral enhancing walls in the lingular lobe and regions of consolidation in the left apicoposterior and lingular segments, as well as bilateral pleural effusion.

The right-sided pleural fluid investigation revealed transudative pleural effusion. The daily ICD drain was...
monitored (it drained about 1.5 L) and was withdrawn when the drainage was <50 ml/day for the next 3 days. CXR after removal revealed resolution of the effusion with left-sided MZ heterogeneous opacity. The patient has improved symptomatically and has been discharged.

The most common cause of Contarini’s syndrome is the development of transudative effusions due to congestive cardiac failure (opportunistic infections) with preexisting pneumonia/parapneumonic effusion. A common combination is a parapneumonic effusion that causes heart failure, which results in a contralateral transudate. In the book of pulmonary diseases, Hinshaw lists those secondary infections of serous pleural effusions which lead to empyema.

The etiology of the parapneumonic effusion in our instance is unknown, although it can be hypothesized on. The best explanation in this immunotherapy patient is gravity movement of contaminated buccal secretions from the lip abscess down to the lung, leading to pneumonia and eventual empyema through direct extension. Because of the noninfected effusion, negative blood cultures, and lack of a lung abscess, a hematogenous route of infection is less plausible. Hypoalbuminemia can induce transudative pleural effusion.

Contarini’s syndrome is a rare and unique disease that is likely underdiagnosed. When patients with autoimmune diseases who are receiving immunotherapy come with imaging scans indicating diverse types of pleural effusions, this condition should be considered. Due to a greater number of risk factors, immunocompromised people may be more susceptible to develop pleural effusions of various etiologies.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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