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Cryptogenic Fibrosing Alveolitis Necessitating Therapeutic Termination of A First Trimester Pregnancy
A Case Report

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Twenty-seven years old Bahraini lady, known to be suffering with fibrosing alveolitis for some eight years, presented to the accident and emergency department of Salmaniya Medical Complex. Recently her respiratory function had deteriorated and she had become house-bound on home oxygen therapy and nebulizers. She was taking Prednisilone tablets and Immuran. She was admitted to the Intensive Care Unit (ICU) in extreme respiratory distress for the past two days. While she was in the hospital, she complained of breast tenderness and supra pubic pain, her period being twenty days overdue. BHCG pregnancy test proved to be positive. Management and outcome of her case is herewith discussed.

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Cryptogenic fibrosing alveolitis (CFA) is uncommon among young women in their childbearing years[1-6]. Very little so far is known about its aetiology[1]. The course of the disease is progressive and the prognosis is generally poor. Most patients try steroids but only ten to twenty percent show any improvement[2]. Alternative medical treatment is available, but the results are not encouraging. Single lung transplantation may be offered to suitable patients with life-threatening disease.

To date few studies have examined pregnant women with carefully defined restrictive lung disorders[3]. The majority of restrictive pulmonary diseases have their onset after the childbearing years. When present they do not alter fertility.

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Furthermore, these disorders are only relative contraindication to pregnancy unless the case is too advanced and the patient is receiving oxygen continuously as was our patient. Other conditions which worsen the prognosis for these patients include advanced maternal age, advanced radiographic staging and the presence of extrapulmonary sarcoidosis[4].

Most women with CFA are advised to avoid pregnancy by using non-oestrogenic compounds because of the concern that they will lead to worsening of the disease[5,6]. Thus, contraceptive devices of progesterone preparations are recommended. Naturally if indicated, vasectomy of the husband or sterilization of the patient is an alternative.

THE CASE

Twenty-seven years old woman (G4 P2 Ab1), known to be suffering with fibrosing alveolitis for some eight years, presented to the accident and emergency department of Salmaniya Medical Complex. Recently her respiratory function had deteriorated, and she had become house-bound on home oxygen therapy and nebulizers. She was taking Prednisolone tablets and Immuran.

She was admitted in extreme respiratory distress for the previous two days. She had a productive cough with white sputum and a high grade fever. Her previous history consisted of frequent admissions to the hospital including time in the Intensive Care Unit (ICU) because of similar symptoms. She has two living children, the last delivery being 1.4 years ago. Subsequently, she has practiced no contraception because her G. P. had advised her against steroidal contraception.

On admission, she was very dyspneic, tachypneic and exhibited tachycardia. The examination of the lungs showed limited air entry bilaterally with inspiratory and expiratory crepitations associated with rhonchi. There was no bronchial breathing. Auscultation of the heart revealed no gallop, S 1 and S 2 were normal. A preliminary diagnosis of pneumonia complicating her condition necessitated her admission once again to the ICU.

In the ICU, she was monitored continuously and further investigated and placed on the following medications: Heparin 5000 units twice daily subcutaneously; Rocephin 2 G in an IV drip; Erythromycin 500 mg Q 6h p.o.; Septrim tabs, 1 tab bid p.o.; Immuran tabs 100 mg OD p.o.; Ranitidine 150 mg bid p.o. (for peptic ulcer prophylaxis).

She remained in the ICU for another two days, then was transferred to the medical ward.

Two days after being transferred from the ICU the sputum became greenish and the patient became more dyspneic and cyanosed. Blood and urine cultures were sterile, but frequent ABG's showed evidence of deterioration of her pulmonary function. The antibiotic was changed to Tazocin. Over the next ten days she improved slowly but progressively. She had tongue ulcerations and skin lesions, which were diagnosed to be iatrogenic as a result of drug therapy.

On the eighteenth day following her admission, she complained of breast tenderness and suprapubic pain. Her period was twenty days overdue. BHCG urine pregnancy test was positive. Ultrasound revealed an intrauterine pregnancy of some seven weeks. The pulmonary consultant was concerned about this pregnancy and the advanced stage of the restrictive lung disease. He suggested termination of the pregnancy and sterilization. The patient and her husband were counseled and together they agreed to the termination and sterilization. A vacuum aspiration of the eight week pregnancy and the suprapubic tubectomy were performed under regional anesthesia. The postoperative period was uneventful. Six weeks after admission the patient is still in the medical ward but her condition is stable.

DISCUSSION

It is uncommon to see cases of fibrosing alveolitis in women at Salmaniya Medical Complex. The cryptogenic type is more common among young females in their reproductive years, while the collagen type of the disease is more common among the older patients. This patient was a known asthmatic for ten years, and later had superimposed fibrosing alveolitis for the past two years. Despite treatment with Prednisolone and Immuran, her condition had progressively deteriorated and she had become house-bound on oxygen therapy.

Her latest episode of fever, breathlessness, tachypnea and tachycardia was precipitated by pneumonia, which responded very slowly to intensive therapy with antibiotics. The discovery of an early pregnancy under these circumstances constituted a further challenge to her critical condition. Why she has not taken any kind of contraceptive measures is incredible, considering the repeated advice of her attending pulmonary physician.

The termination was requested by her Consultant and discussed with two gynaecologists and a senior anaesthetist. There was an agreement on the need to do the operation combined with sterilization under regional anaesthesia.

Similar cases have been reported in the medical literature in which therapeutic termination was performed at much later stages of pregnancy[2,4]. There is minority of physiologists who believe that pregnancy will not necessarily cause deterioration of pulmonary function in cases of CFA. However there are no reliable studies amongst women with CFA in their reproductive phase to compare the prognosis before and after. Most specialists agree with the need for termination of pregnancy in women with severe cases of CFA[1-6].

CONCLUSION

Continuation of pregnancy is not contraindicated in all forms of restrictive pulmonary disease, but advanced cases of cryptogenic pulmonary fibrosis is a strong indication for therapeutic termination. The availability of unilateral lung transplants is still scarce and difficult in the Gulf region, so sterilization is indicated if the woman has completed her family, otherwise non-steroidal contraceptives such as intrauterine devices or progesterone orally are indicated in women afflicted with this condition.

In reference to this case we have presented a young woman in the childbearing phase with worsening of severe cryptogenic fibrosing alveolitis complicating an eight weeks pregnancy. A combined decision to terminate her pregnancy was made by the gynaecologists and her chest physician.

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