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Thoracoscopic management of recurrent spontaneous pneumothorax and incidentally detected eventration of diaphragm in a case of Marfan's syndrome: A case report

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Abstract: Recurrent pneumothorax has been reported in patients with Marfan syndrome and is more common in such patients. We report a case of a child who had 2 episodes of spontaneous pneumothorax. Patient was treated by intercostal drainage tube on each episode. After 2nd episode, patient was investigated by CT thorax. No evidence of bullae/emphysema was found. After 3 weeks he again presented with 3rd episode of spontaneous pneumothorax. Thoracoscopy was done, no bullous/emphysematous lesion found, incidental finding of eventration of left dome of diaphragm was found which was corrected by plication. Pleurodesis was done on post-op day 5 to prevent recurrence of spontaneous pneumothorax. Patient had uneventful recovery and was discharged and is on regular follow-up.This case is rare as association of Marfan syndrome with eventration of diaphragm has not been reported in pediatric patient while recurrent spontaneous pneumothorax is a known complication in this syndrome.
Keywords: Marfan syndrome, pneumothorax, Thoracoscopy, eventration, Pleurodesis

INTRODUCTION

Spontaneous pneumothorax is reported to occur in patients of Marfan syndrome. We present this case as association of Marfan syndrome with eventration of diaphragm has not been reported. Plication of left dome of diaphragm was done thoracoscopically. Talc powder put into left pleural cavity for achieving Pleurodesis. Patient is on 3 monthly regular follow up without any symptoms.

CASE REPORT

11 years old male child presented with acute onset of left side chest pain and breathlessness. He had reduced air entry on left side. Chest x-ray showed left side pneumothorax .Intercostal drain tube (ICD) was placed in 5th intercostal space. Past history revealed 1 similar episode of pneumothorax 2 months back and it was treated by putting intercostal drain tube.

Patient had thin built with poor muscular development. His height was 162 cm, upper segment being 79 cm and lower segment 83 cm. Ratio of two is 0.95. Patient had arachnodactyly. His arm span was 170 cm. Patient had asymmetric left sided pectuscarinatum and high arched palate. His ophthalmic examination was normal. Cardiovascular examination revealed systolic murmur at apex.

2-D echo revealed dilated aortic root and mitral valve prolapse. Patient was diagnosed as a case of Marfan syndrome based on these morphological features[1-3].

CT thorax was normal except abnormal shape of bony thorax. No evidence of bullous lesions or emphysema found (fig 1).

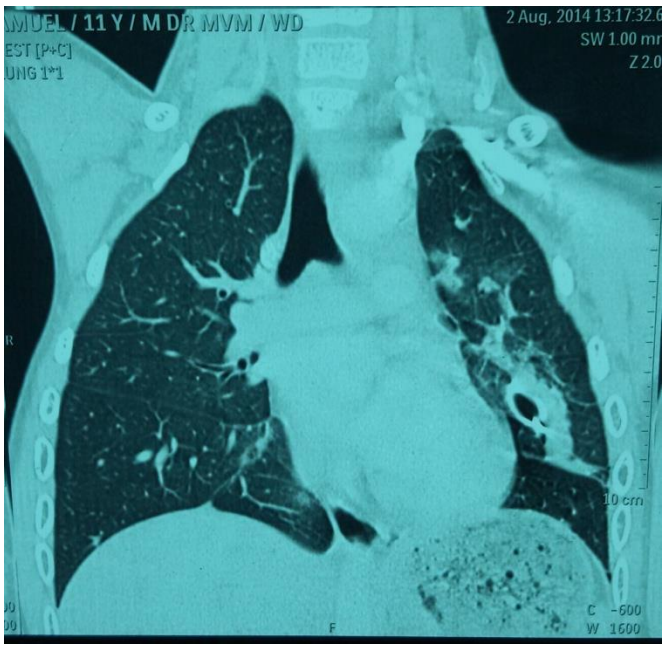

Fig. 1: CT thorax - No evidence of bullous lesions or emphysema found

Patient was discharged. He came back after 3 weeks with same complaint of acute onset pain in left side of chest and breathlessness. Chest x-ray showed left side pneumothorax which was managed by putting intercostals drainage tube in left 5th intercostals space. As this was 3rd episode of spontaneous pneumothorax,patient was evaluated by Thoracoscopy. No evidence of bullous lesion or air leak was found (fig 2).

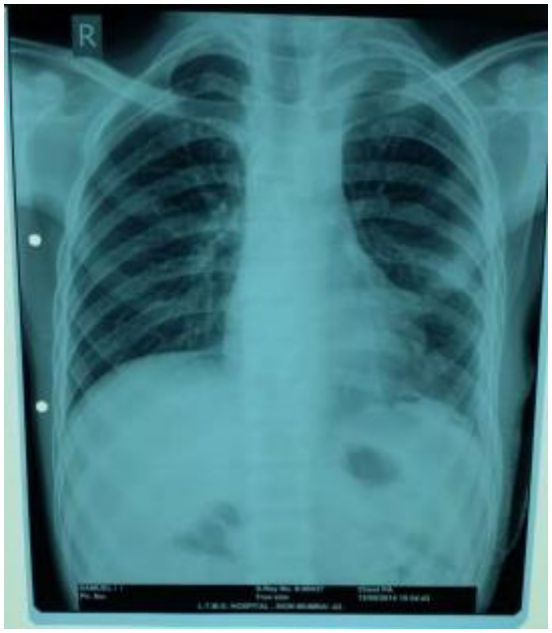

Fig. 2: Post-op chest X-ray showing expanded lungs and plicated left dome of diaphragm seen at lower level than right dome

However incidental finding of eventration of left dome of diaphragm found and it was corrected thoracoscopically by plication of left dome of diaphragm. (Fig.3) Pleurodesis of left chest cavity was done on post-op day 5 by putting 20 grams of talcum powder mixed with 20 ml NS through ICD. ICD kept clamped for 1 day and patient monitored for 1 day. Left dome of diaphragm was seen at lower level compared to right dome of diaphragm. ICD was removed on post-op day 7. Patient had uneventful recovery and discharged and is on 3 monthly follow-up.

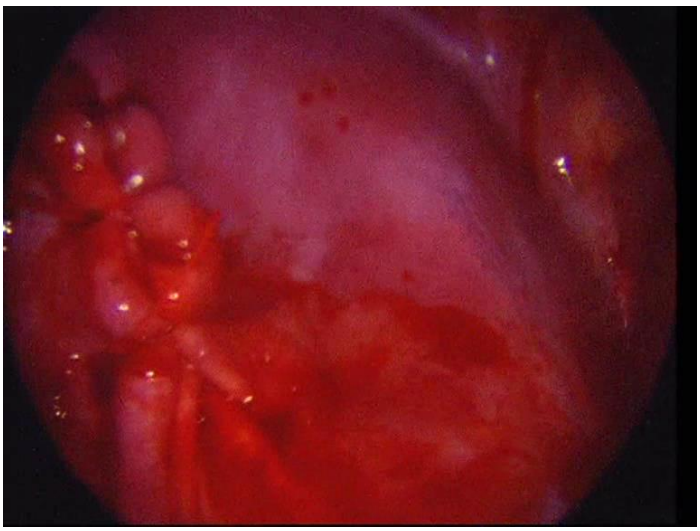

Fig.3: Thoracoscopic plication of left dome of diaphragm

DISCUSSION

Marfan syndrome (also called Marfan's syndrome, MS) is a genetic disorder of human connective tissue. It has different manifestations ranging from mild to severe: the most serious complications which often lead to early death involve heart and aorta. The syndrome also affects lungs, eyes, hard palate. About 24 instances of spontaneous pneumothorax (SP) have been reported in individuals affected by the MS [1-3].

MS patients are prone to form bullae or blebs, and SP has been described in about 10% of them. Spontaneous pneumothorax is the most acute pulmonary manifestation[4,5].Because of their connective tissue defect, tall stature, and thoracic cage deformities, Patient with MS is 10 times more likely to sustain SP than healthy individual [4].

Marfan syndrome patients have higher risk of bulla rupture because of the increased forces generated at the lung apices[5,6].

It is recommended that management of pneumothorax be initiated by chest tube drainage. Lung parenchyma appearance on chest radiographs and the air leak in intercostal drainage tube should be observed. If either bullous changes or a persistent air leak is present, then resection of the offending bulla and apical pleurodesis should be performed. The role of talc as the agent to achieve pleurodesis has become popular in treating patients with primary spontaneous pneumothorax (PSP) [7].

Talc pleurodesis in young patients with a primary pneumothorax appears to have minimal long-term adverse effects[8].

Our experience with this case of Marfan syndrome with recurrent spontaneous pneumothorax with incidentally detected eventration of left dome of diaphragm showed that exact cause for recurrent pneumothorax may not be found in these patients. Recurrence of spontaneous pneumothorax warrants stabilization of patient first by ICD insertion and then definitive surgical intervention in the form of VATS and treatment of underlying pathology. In absence of bullous lesion, pleurodesis has been effective to prevent recurrence of spontaneous pneumothorax in our patient.

People with the Marfan syndrome should avoid contact sports (to protect both the lung and the aortic root) and scuba diving (to protect against rupture of lung bullae). Pneumothorax along with aortic dissection should be considered, in any patient with Marfan’s syndrome in whom acute chest pain, with or without dyspnea, develops.

Association of eventration of one dome of diaphragm in a child of Marfan syndrome has not been reported previously, making this a rare case.

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