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Case Report

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Co-existince of Sickle Cell Disease and Hemidiaphragm Paralysis

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Prominent right hemidiaphragma elevation was observed on chest radiograph of a 14 years' old female patient with sickle cell disease. Her medical history yielded neither trauma nor intra-thoracic surgery. She didn't have either motor deficit or sensation disorder on any region of her body. Thorax CT yielded no lession except the significantly elevated right diaphragma. Her cranial CT showed no lesion, too. Diagnosis of right hemidiaphragm paralysis was confirmed by positive Hitzenberg sniff test on fluoroscopy. Although several pathopysiologic mechanisms are known to be involved and lead to central neurologic complications in sickle cell disease, involvement of peripheric nerves have not been reported. Here we present a 14 years' old female patient with sickle cell anemia and unilateral diaphragm paralysis, co-existince of which have not been reported so far.

Key words: Sickle cell, diaphragma paralysis, complication

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INTRODUCTION

Sickle Cell (SC) anemia is a disease caused by production of abnormal hemoglobin, which binds to other abnormal hemoglobin molecules within the red blood cell to cause rigid deformation of the cell[1]. The obstruction of small vessels by sickle cells results in repeated infarctions, leading to gradual involvement of all organ systems, most notably spleen, lungs, kidney and brain[2]. Infection, acute splenic sequestration crisis, aplastic crises, acute chest syndrome, stroke, cholelithiasis, renal disease and pain are the major complications of this disease[3]. Many neurologic complications other than stroke have also been described in SC disease including drowsiness, coma, convulsions, headache, temporary or permanent blindness, cranial nerve palsies and parestesias of the extremities[3]. The lungs are also commonly affected, with infarcts, emboli (from marrow infarcts and fat necrosis) and a markedly increased propensity for pneumonia[3].

The most important respiratory muscle is the dome-shaped diaphragm, which is innervated by cervical motor neurons C3-5 via the phrenic nerves. Diaphragma paralysis can involve either the whole diaphragm (bilateral) or only one leaflet (unilateral). Unilateral or bilateral diaphragm paralysis maybe seen following phrenic nerve injury and with a variety of motor-neuron diseases, myelopathies, neuropathies and myopathies[4]. Hemidiaphragm paralysis is more common than bilateral paralysis and is usually diagnosed from unilateral elevation of the hemidiaphgram on chest radiograph[4].

Here we present a 14 years' old female patient with sickle cell anemia and unilateral diaphragm paralysis, co-existince of which have not been reported so far.

Case report: A 14 years' old female patient known to have sickle cell anemia and being periodically followed up since she was 6 months' old, admitted to Pediatry Clinic for her routine control visit. She did not have any complaints other than fatigue and dyspnea on exertion. She had suffered from 5-6 painfull vaso-occlusive crisis per year until she was 10 years' old and only 5 in total since then. A pale skin was observed on inspection. Increased dullness and decreased intensity of pulmonary sounds on posterior wall of right hemi-thorax were detected. Physical examination of her cardio-vascular and other systems were entirely normal. She had neither motor deficit nor sensation disorder on any region of her body including dermatomes of cervical 3-5 from which phrenic nerves originates. Her Complete Blood Count (CBC) results were as follows WBC:10.32, RBC:2.62, HGB:8.6, MCH:32.8 MCV:96.1 fL, MCHC:33.9 pg, PLT:337, MPV:10.1.

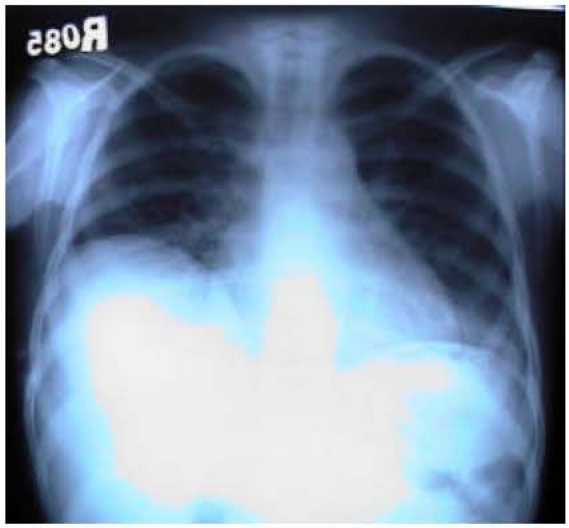

Fig. 1: PA chest radiograph of the patient shows the elavated right diaphragm with its apex significantly (> 4 cm) higher than that of the left

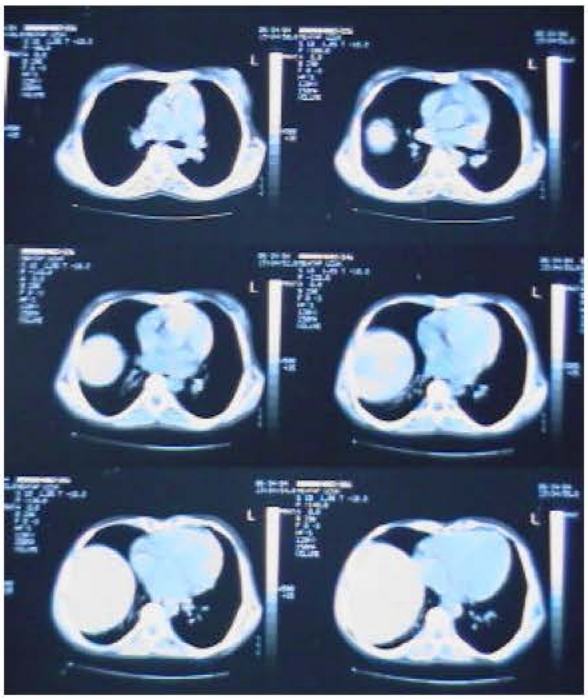

Fig. 2: CT slices of her thorax shows only the elevated rightdiaprgma without any mediastinal mass or parenchymal lessions

Hemoglobin electrophoresis yielded HbS:86.4, HbF:5.1 and HbA2:5.4. Prominent right hemi-diaphragma elevation was observed on her routine Postero-Anterior (PA) chest radiograph. The apex of the right hemi-diaphragma was significantly (> 4 cm) higher than that of the left (Fig. 1).

Any other radiological abnormality was not observed. Spirometry showed moderate to severe restriction in respiratory function with her FEV_1=53%, FVC=55%, FEV_1/FVC=97%, PEF=43% and $FEF_{25-75\%}$=58% of predicted. Her medical history yielded neither trauma nor intra-thoracic surgery, as a possible cause of diaphragm paralysis. So Computerized Tomography (CT) of thorax and upper abdominal ultrasonography were performed in order to rule out any possible reason for phrenic nerve paralysis such as a mediastinal mass and for right diaphragma elevation such as a hepatic tumor, respectively. Thorax CT yielded no lession except the significantly elevated right diaphragma (Fig. 2). Upper abdominal USG was normal with absence of any hepatic tumor and subpulmonary effusion. Her cranial CT showed no lesion, too. Diagnosis of right hemidiaphragm paralysis was confirmed by positive Hitzenberg sniff test on fluoroscopy.

DISCUSSION

Pulmonary complications lead to mortality and morbidity in patients with SC disease[5]. In addition lung function abnormalities are present in young children with SC disease and restrictive abnormals become more prominent with increasing age[6]. This patient has moderate to severe restrictive type dysfunction dysfunction but this situation is probably due to unilateral diaphragm paralysis in major part rather than the SC disease itself.

Diaphragm paralysis maybe seen following phrenic nerve injury and with a variety of motor-neuron diseases, myelopathies, neuropathies and myopathies[4]. None of these possible reasons could be identified in our case. She didn't have any motor deficit or sensation disorder on any part of her body, either. She had exposed neither to trauma nor to intra-thoracic surgery. Besides CT of thorax did not show any mediastinal mass or any lessions such as pneumonia, pleurisy, aortic aneurysm, substernal goiter, neoplasm adjacent to a phrenic nerve all of which may lead to phrenic nerve injury or compression. Thorax CT revealed the significantly elavated right diaphragm (Fig. 2).

Patients with SC disease have an approximately 25% chance of developing some type of neurological complication in their lifetime[7]. The neurological complications result from one point mutation that causes vasculopathy of both large and small vessels. Anemia and the resultant cerebral hyperemia produce conditions of hemodynamic insufficiency. Sickled cells adhere to the endothelium, contributing to a cascade of activated inflammatory cells and clotting factors, which result in a nidus for thrombus formation. Because the cerebrovascular reserve becomes exhausted, the capacity for compensatory cerebral mechanisms is severely limited. There is evidence of small-vessel sludging and a relative deficiency of nitric oxide in these vessels further reduces compensatory vasodilatation. Both clinical strokes and silent infarcts occur, affecting motor and cognitive function[7]. Nevertheless these may explain only central neurological complications in SC patients. In our case Cranial CT was normal as well as her motor and cognitive functions. We suspect, if similar mechanisms which are involved in the repeated vaso-occlusive crises may also lead to peripheric nerve dysfunction which has not been reported so far.

Therefore, we present this case as a coexistence of sickle cell disease and hemidiaphragma paralysis.

REFERENCES

1. Lonergan, G.J., D.B. Cline and S.L. Abbondanzo, 2001. Sickle cell anemia. Radiographics, 21: 971-974.
2. Beutler, E., 1998. Disorders of Hemoglobin. In: Fauci A.S., E. Braunwald and K.J. Isselbacher *et al.*, Eds. Harrison's Principles of Internal Medicine. 14th Edn. McGraw-Hill Companies, pp: 648-650.
3. Lee, G.R., J. Forester and J. Lukens, Eds., 1999. Wintrobe's Clinical Hematology. 10th Edn. Lippincott Williams and Wilkins Company, pp: 130-135.
4. Criner, G.J. and S.G. Kelsen, 1998. Effects of Neuromuscular Diseases on Ventilation. In: Fishman A.P., J.A. Elias and J.A. Fishman *et al.*, Eds. Fishman's Pulmonary Diseases and Disorders. 3rd Edn. McGraw-Hill Companies, pp: 1575-1576.
5. Greenough, A., 2004. Sickle cell disease-Pulmonary complications and a proinflmmatory state. AJRCCM., 169: 663-665.
6. Sylvester, K., R.A. Patey and M. Dick *et al.*, 2004. Pulmonary function abnormalities in children with sickle cell disease. Thorax, 59: 67-70.
7. Prengler, M., S. Pavlakis and I. Prohovnik *et al.*, 2002. Sickle cell disease: The neurological complications. Ann. Neurol., 51: 543-552.