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Case Report

Hodgkin lymphoma patient presenting as superior vena cava syndrome in a patient suffering from myotonia congenita: A rare case scenario

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ABSTRACT

A 14-year-old male who was a known case of myotonia congenital (MC) (SCN4A mutation related) on Tab phenytoin therapy presented with progressive shortness of breath, swelling of face, and intermittent fever for 2 months. On investigation, a middle mediastinal mass was diagnosed and histopathological examination along with immunohistochemistry revealed it to be Hodgkin lymphoma (HL). The patient was started on Inj. dexamethasone for superior vena cava syndrome followed by doxorubicin, vincristine, etoposide, and prednisolone chemotherapy and phenytoin dose adjustment. The patient responded well to treatment. This case summarizes the management in patients of HL presenting as superior vena cava syndrome in a background of MC on enzyme inducer therapy.

Keywords: Superior vena cava, Hodgkin, Lymphoma, Myotonia, Phenytoin

INTRODUCTION

The superior vena cava syndrome (SVCS) refers to the constellation of clinical symptoms and signs resulting from increased venous pressure upstream due to complete or partial superior vena cava (SVC) obstruction resulting in impaired venous return to the heart from the head, neck, thorax, and upper extremities, and subsequent development of venous shunts to bypass the obstruction. The most common causes include non-small-cell lung cancer (50%) followed by small-cell lung cancer (25%), device-related (25%), and non-Hodgkin lymphoma (HL) (10%).[1] The relationship between HL and SVCS is ill-defined. Myotonia congenita (MC) is a group of congenital neuromuscular channelopathies characterized by typical facies and delayed relaxation of the muscles following voluntary or induced contraction. It is caused by mutations in the skeletal muscle chloride channel gene (CLCN1) and the skeletal muscle sodium channel gene (SCN4A) causing hyperexcitability of muscle fibers.[2]

CASE REPORT

A 14-year-old male presented with shortness of breath, swelling of the face, and intermittent fever for the past 2 months. The patient was a diagnosed case of MC (SCN4A mutation related) at the age of 3 years and was on Tab Phenytoin therapy since then.

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On examination, hatchet facies [Figure 1] was present along with facial puffiness, sternal dullness, and decreased air entry right middle, and lower zone with oxygen saturation of 84% at room air was present.

Baseline hematological and biochemical parameters are mentioned in [Table 1]. Chest X-ray showed ill-defined homogenous opacity in the right lung middle zone obscuring the right heart border. High-resolution computed tomography thorax revealed a large ill-defined heterogenous necrotic mass lesion in the middle mediastinum with moderate right-sided pleural effusion. Pulmonary function test showed forced expiratory volume in 1 s/forced vital capacity 83%. Suspecting SVCS patient was started on Inj. dexamethasone, Inj. furosemide, and IV broad-spectrum antibiotics with head end elevation and oxygen support. Histopathological examination of trucut biopsy from mediastinal mass showed extensive areas of necrosis, focal areas of stromal infiltration by a heterogenous population of cells comprising of polymorphs, lymphocytes, plasma cells, and histiocytes; scattered large cells having vesicular nuclei and distinct nucleoli. The immunohistological examination was suggestive of classical HL. Positron emission tomography computed tomography (PET CT) scan whole body suggested a metabolically active (SUV max 7.5) large lobulated mass in the anterior and middle mediastinum on the right side causing displacement of mediastinal structures toward the left along with fluorodeoxyglucose (FDG) avid enhancing enlarged prevascular, right upper and bilateral lower paratracheal, subcarinal, and right hilar lymph nodes (SUV Max 10.8) with low-grade FDG avid (SUV Max 1.7) soft-tissue density lesion seen in the posterior basal segment of the right lung lower lobe [Figure 2].

The patient was started on the first cycle of OEPA (Vincristine, Etoposide, Prednisolone, Doxorubicin) chemotherapy with a dramatic improvement of respiratory symptoms both clinically and radiologically. Neurology opinion was sought for the effect of enzyme inducer drug phenytoin on the efficacy of chemotherapeutic agents and for the doses of myopathy aggravating drugs vincristine and steroids. As per advice, the dose of phenytoin was tapered over a span of 7 days, and normal doses of vincristine and steroids were given with watchful observation. The patient tolerated the chemotherapy well and is planned for 1 more cycle of OEPA followed by a repeat PET CT scan.

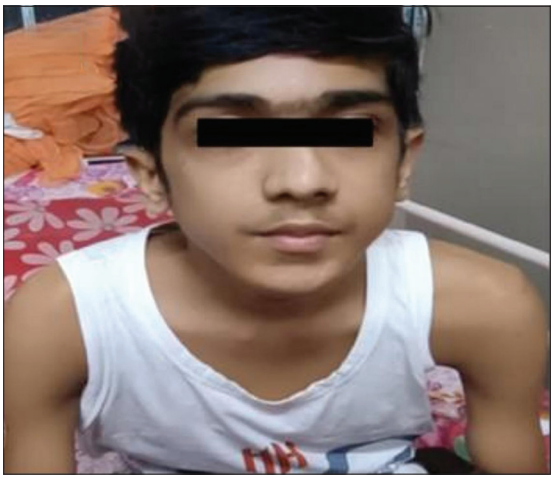

Figure 1: Hatchet facies of myotonia congenita.

Table 1: Baseline parameters.

Hemoglobin	10.3 g/dl
Total leukocyte count	23500/cmm (no immature cells and neutrophilic predominance)
Platelet count	180000/cmm
Urea	3 g/dl
Creatinine	0.8 g/dl
Serum sodium	135 meq/l
Serum potassium	4 meq/l
Serum calcium	8.5 g/dl
Serum magnesium	1.8 g/dl
Serum phosphate	4.5 g/dl
Liver function tests	Total bilirubin: 0.4 Aspartate transaminase: 45 U/L Alanine transaminase:42 U/L Alkaline phosphatase:103 U/L Total protein: 6.0 g/dl Serum albumin: 3.8 g/dl
LDH	1210 U/L
Immunohistochemistry of truecut biopsy of mediastinal mass	Large cells positive forCD30,CD15 & PAX-5 Negative for CD20 & CD45 Background cells positive for CD3 Suggestive of classical Hodgkin lymphoma

DISCUSSION

SVCS is a medical emergency. Its association with classical HL is rare. Supportive measures such as head end elevation, oxygen inhalation, and diuretics are helpful in patients of SVCS due to any cause, dexamethasone also provides symptomatic relief by decreasing edema and tumor burden in malignant causes of SVCS. Thrombolysis is indicated in the presence of acute thrombus.[3] To date, no case has been described of SVCS with classical HL in a setting of MC. The activity of taxanes, vinca alkaloids, methotrexate, teniposide, and camptothecin analogs is known to be impaired by enzyme-inducing sodium channel-blocking drugs like phenytoin. Phenytoin increases the metabolism of corticosteroids and increases the clearance of taxanes and

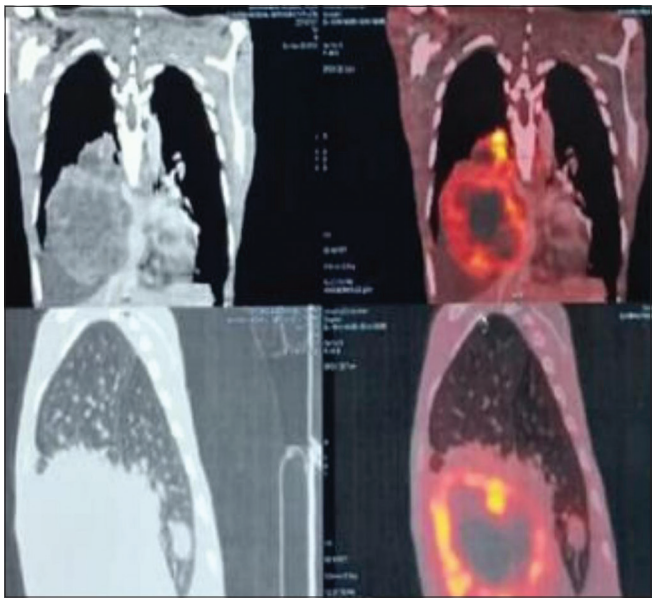

Figure 2: Positron emission tomography- computed tomography scan showing metabolically active (SUV max 7.5) large lobulated mass in anterior and middle mediastinum on the right side displacing mediastinal structures to the left.

vinca alkaloids. Hence, comprehensive medical history along with knowledge of chemotherapeutic drug interactions is required to avoid potential drug interactions that may reduce the efficacy or enhance toxicity of chemotherapy drugs.[4]

CONCLUSION

SVCS is a rare presentation in patients with HL. To date, no case of HL in a background of MC (SCN4A mutation related) has been described. This case summarizes the approach to the management of HL-associated SVCS in a background of MC on phenytoin (enzyme inducer) therapy.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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