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CASE REPORT

The Unspoken Presentation of the Uncommon: Superior Vena Cava Syndrome

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ABSTRAK

'Superior Vena Cava Syndrome' merupakan diagnosa perubatan yang kritikal. Pengesanan and perawatan penyakit tersebut pada peringkat yang awal bukan sahaja boleh mengurangkan kadar kematian dan juga mobiditi pesakit. Kes klinikal ini memaparkan seorang pesakit lelaki yang berumur 53 tahun, berpenyakit 'Parkinson' dan strok iskaemia telah datang ke Jabatan Kecemasan dengan simptom penyakit 'Superior Vena Cave Syndrome' selama empat hari. Cabaran yang dialami dalam kes ini adalah pesakit tidak boleh bercakap dan kehidupan seharian beliau bergantung kepada ahli keluarga. Kebanyakan informasi adalah diperolehi daripada pemerhatian ahli keluarga yang menjaga pesakit. Oleh itu, kesangsian klinikal berdasarkan sejarah klinikal adalah sangat penting dalam diagnosa dan perawatan awal penyakit 'Superior Vena Cava Syndrome'.

Kata kunci: mediastinum, superior vena cava syndrom, tumor

ABSTRACT

Superior vena cava syndrome is a medical emergency. The morbidity and mortality of superior vena cava syndrome can be decreased by early recognition, early establishment of primary cause, early supportive and definitive treatment. We illustrate a case of 53-year-old male with underlying advanced Parkinsonism and ischemic stroke who presented with superior vena cava syndrome over 4 days period. Diagnostic challenge in this patient lies in the fact that patient had aphasia and his daily living activity was dependent. Most of the information was based on caretaker's observations. A strong clinical suspicion based on clinical history is crucial for early diagnosis and treatment of superior vena cava syndrome.

Keywords: mediastinum, superior vena cava syndrome, tumour

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INTRODUCTION

Superior vena cava syndrome is a constellation of clinical presentations that is caused by partial or complete obstruction of the superior vena cava. It was first reported in 1757 in a patient with syphilitic aortic aneurysm (Schindler & Vogelzang 1999). Malignant neoplasm is now the commonest etiology, accounts for more than 90% of the cases (Beeson et al. 2014). Non-malignant causes such as catheter related thrombosis, vascular disorder, infections, mediastinum benign tumour and mediastinum fibrosis accounts for the remaining cases. With increase in use of intravenous catheter, incidence of catheter related superior vena cava syndrome is increasing (Rice et al. 2006). Untreated malignant superior vena cava carries mortality rate of 30% (Hassikou et al. 2002). Despite of the high mortality rate, it is often left undiagnosed until the later stage due to its gradual increase in symptomatology. Therefore, prompt recognition of superior vena cava syndrome is very important to ensure a better outcome of the patient.

CASE REPORT

A 53-year-old male with underlying ischemic stroke and Parkinsonism stage IV was brought in by family members to Emergency Department complaining of sudden onset of bilateral upper limbs and neck swelling for 4 days duration. It was associated with rapid breathing and rash over the upper torso region. Otherwise, there was no fever, prolonged cough or any constitutional symptoms. There was no history of trauma. Patient was activities-of-daily-life (ADL) dependent and aphasic for one year due to ischemic stroke. There was no recent change in conscious level.

On examination, patient was able to open eyes spontaneously, obeying simple command but only making incomprehensible sounds. His vital signs were as follows: blood pressure: 121/79 mmHg, heart rate: 110/min, temperature: 36.7˚C, oxygen saturation: 95% under room air, respiratory rate: 28/min. There was no stridor heard. On inspection, patient had facial puffiness with diffuse neck swelling extending from the base of neck to angle of mandible. There is right supraclavicular lymphadenopathy measuring 6cm x 4cm. Pitting oedema was noted over right upper limb. Dilated veins were noted over the chest and the neck. His lungs are clear on auscultation, cardiovascular examination reveals dual rhythm no murmur. Neurological examination revealed positive signs of Parkinsonism and residual neurological deficit over the right upper and lower limbs. There were no signs of Horner's syndrome.

Initial investigations show total white count: 18 x 10^9 per litre with 80% of neutrophils, hemoglobin level :12g/dL, platelet: 340 x 10^3 per l and C-reactive protein: 5 mg/L indicative of on-going inflammation. Her renal profile shows urea: 4.5 mmol/L, Sodium: 140 mmol/L, Potassium: 3.5 mmol/L and creatinine: 60 mol/L but associated with elevated corrected serum calcium level of 2.80 mmol/L. Liver function

test showed hypoalbuminaemia with albumin level of 30 g/L and the rest of parameters were normal. Chest X-ray showed widened mediastinum and clear lung field. CT thorax revealed matted mediastinal lymphadenopathy associated with superior vena cava and left brachiocephalic vein thrombosis.

Patient was then started on initial intravenous dexamethasone 4 mg with the aim to reduce soft tissues swelling. Hydration with intravenous 0.9% normal saline was initiated for hypercalcaemia. Patient was then referred to medical team and Ear Nose and Throat team. Diagnosis of superior vena cava obstruction secondary to mediastinum mass was made. The primary cause of malignancy was still under investigation.

DISCUSSION

Superior vena cava is a thin-walled structure with low pressure. Therefore, it is susceptible to extrinsic pressure and intra-luminal thrombosis. Obstructed superior vena cava will result in retrograde flow via collateral venous system and interstitial oedema. Obstruction of superior vena cava by external compression tends to present more gradual compared to those caused by intra-luminal thrombosis (Higdon & Higdon 2006). The most frequent signs and symptoms are face or neck swelling (82%), upper extremity swelling (68%), dyspnoea (66%), cough (50%), and dilated chest vein collaterals (38%) (Rice et al. 2006).

This case illustrates the classic clinical presentations of dyspnoea, collateral vein dilatation on chest wall, neck, facial and upper limb swelling. Superior vena cava syndrome is a clinical diagnosis that does not have definitive diagnostic criteria. Clinical presentations vary depending on the acuity of obstruction and adequacy of collaterals development. Therefore, it is often missed during the early stage of the disease especially during the first encounter to medical facility. In this case, the diagnosis is solely established based on the clinical signs identified during physical examination due to the limited history. The rapid onset of symptoms is also misleading as Superior Vena Cava Syndrome tends to occur in a more gradual manner. On the other hand, rapid progression of symptoms over a period of 4 days suggests malignancy or thrombosis as the likely cause of obstruction. It is further confirmed by CT thorax which showed the co-existence of new onset mediastinum mass and thrombosis of superior vena cava and left brachiocephalic vein thrombosis. Obstruction of superior vena cava by external compression tends to present more gradual as compared to those caused by intra-luminal thrombosis (Higdon & Higdon 2006). Subtle signs such as cough, dyspnoea, hoarseness, chest pain, jugular vein distension, and oedema of the hands, face, or neck can be very non-specific. Normally, symptoms worsened in the morning or with certain posture such as bending forward or lying flat. Late signs and symptoms can be life-threatening, such as respiratory distress, stridor, mental status changes, syncope, and cyanosis of the face and upper body (Colen 2008). Complications such as

oesophageal varices (Greenwell et al. 2007) and pleural effusion (Rice 2007) were reported in patients with superior vena cava syndrome. Pleural effusion occurred in nearly 60% patients with superior vena cava. However, this feature was absent in this patient. It usually resolves upon correction of superior vena cava obstruction (Rice 2007).

Approach to superior vena cava syndrome in Emergency Department includes prompt recognition of superior vena cave syndrome and stabilization of airway, breathing and circulation. Early recognition of superior vena cava syndrome is of utmost importance as it will expedite the establishment of primary cause. This is crucial as the prognosis this condition largely depending on the underlying cause. It should be prioritised in the list of differential diagnoses when patient presented with signs and symptoms mentioned above especially in patient with underlying malignancy and intravenous catheter. Investigations such as plain chest X-ray or CT Thorax will help to identify the primary cause. Invasive contrast venography is the gold standard to define the etiology of blockage (Nickloes et al. 2017). Treatment in Emergency Department are mainly supportive. They include airway, breathing stabilization, intravenous administration of glucocorticoid (dexamethasone) and diuretics. However, the use of steroid and glucocorticoid is still controversial (Lewis et al. 2011).

CONCLUSION

Superior vena cava syndrome is a rare medical emergency but potentially life-threatening. Early recognition is vital to ensure a better outcome of such patients. Superior vena cava syndrome has a wide variety of clinical presentations. Early presentations can be non-specific and misleading. Therefore, high index of suspicion is important to avoid misdiagnosis of the patients.

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