

Management of Malignancy Associated Superior Vena Cava Syndrome in the Emergency Department

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Abstract: Objective: The objective of this article was to do a review of the available literature on Malignancy associated Superior Vena Cava Syndrome (SVCS), and provide a quick effective guide on the latest approaches on management of patients presenting in the Emergency Room with SVCS.

Methods: Multiple documents were identified which were published with regards to SVCS, Emergency Management of SCVS and case reports. All the papers were studied and latest diagnostic methods and management protocols were identified and extrapolated from the literature.

Results: Of approximately 30 papers shortlisted that were written on SVCS, newer ways of identifying and managing SVCS in the ER (specifically) were collected and presented. Newer therapies i.e. External Beam Radiation Therapy via Photons in the Emergency setting, Multidisciplinary approach to Malignancy associated SVCS were discussed along with traditional symptoms relief measures.

Conclusion: Both adults and children who present with the signs and symptoms of SVCS in the emergency room require extra care and multidisciplinary management.

Keywords: Superior Vena Cava Syndrome, Oncological emergencies, Superior mediastinal obstruction, Radiotherapy, Cancer, Malignancy.

INTRODUCTION

Superior Vena Cava Syndrome (SVCS) refers to the constellation of symptoms that occur due to obstruction of the Superior Vena Cava [1,2]. SVCS can have multiple etiologies, but malignancy has shown to be the most common causative factor after which increased use of central venous catheters and numerous other intravascular devices [3,4]. Originally SVCS was thought to be a sequel of infection (Tuberculosis/Syphilitic Aortic Aneurysm), in recent times it has been found to be commonly due to malignancies or thrombosis [5-7]. Cases SVCS have also been reported with patients who have pacemakers or endocardial defibrillators placed [8, 9]. The majority of SVCS cases are due to lung cancer (80%), while lymphomas are approximately 15%. If the etiologies of lung cancer causing SVCS are broken down, 50% are due to Small Cell Carcinoma (SCC), as SCC primarily arises usually in the perihilar or central locations of the lung. Another study showed Bronchogenic carcinomas to account for 46% of all Superior Vena Cava Syndrome cases [10, 11]. Albeit not commonly, SVCS progresses to tracheal obstruction (more frequently in children due to their lesser chest volume); this development is referred to as Superior Mediastinal Syndrome (SMS) [1]. Although not reported as frequently, but important to keep in differentials when a patient presents with SVCS like symptoms are Aortic Pseudoaneurysm and mediastinal Hematoma [12, 13].

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In this review we will discuss; identification via signs and symptoms and initial management of Malignancy associated SVCS and SMS in the Emergency Room (ER).

SYMPTOMATOLOGY, SIGNS AND DIAGNOSIS

Diagnosis of SVCS in the ER relies mainly on clinical findings [14]. The hallmark symptom is mainly neck and facial edema (82%); but other signs and symptoms recorded include upper limb swelling (68%), dyspnea (66%), cough (50%), and dilated veins (38%) [2, 3]. Stridor is also an alarming sign that points towards edema of the airways. These symptoms have been recorded to be more in severity when lying down, complaints of orthopnea are commonly found since the supine position increases blood flow to the upper body [14]. If the patient presents acutely with symptoms of increased severity the location of obstruction should be suspected to be below the level of the azygous vein, as the gradual development of symptoms (over months or weeks) is usually associated with collateral vasculature formation, the most important of which is the azygous vein [3]. It should be kept in mind that increased venous pressure due to SVCS can cause laryngeal/bronchial and cerebral edema which are life-threatening consequences, thus prompt identification through relevant focused patient history and physical examination can minimize inpatient mortality [15]. All cases of confirmed malignancy shall be discussed in site-specific multidisciplinary team tumor boards, therefore, previous records of consensus recommendation of the joint experts'

team shall always be available in the patient's medical records [16, 17]. It is advisable to discuss the SCVS/SMS cases with the oncology team and get up-to-date details of the previous and present management of the patient. In certain instances, no electronic medical record is available for emergency room clinicians.

MANAGEMENT

Symptomatic cases of SCVS/SMS presenting in the ER should be managed initially with head elevation, supplementary oxygen therapy along with a course of parenteral steroids (according to institution guidelines), and airway protection (e.g. intubation) [15, 18]. If no alleviation of symptoms occurs, interventional steps are merited including endovascular stenting which has shown to provide immediate relief of neurological symptoms/airway compromise and generally does not hamper further diagnostic modalities [17]. Diagnostic maneuvers such as venography can be used alongside endovascular interventions, as it is considered to be the gold standard in localizing obstructions in the venous passages. Other radiographic imaging techniques and magnetic resonance imaging (MRI) also play an important part in the identification, spread, and etiology of the SVC blockage. The CT chest has shown a sensitivity of 96% and specificity of 92% in the diagnosis of SVC obstruction along with collateral vessels [19-21].

External Beam Radiation Therapy via photons can also be offered in the emergency setting. This recommendation would be made by the Radiation Oncology team after reviewing the history and physical assessment of the patient. If the patient already had previous radiation to the region it would become too difficult to offer a second course of radiation with any meaningful tumoricidal dosage in an acute setting. Certainly, SVCS is regarded as an oncological emergency for radiation oncologists [22].

Certain particular issues are faced by the clinicians who are serving in emergency departments in Low and Middle-Income Countries (LMIC). One special group of patients is those who belong to the pediatric age [23, 24]. Their primary malignancies are quite different from the adult population. Therefore, pediatric cancer patients presenting with the signs and symptoms of SVSC deserve extra care in the emergency rooms. One of the positive points which go in favor of pediatric oncological emergencies is that almost all children with cancer are being treated by standard protocols. Like elective management plans, emergency treatment is also mentioned in internationally accepted pediatric clinical practice guidelines [25]. Clinicians serving in the emergency rooms can consult the pediatric multidisciplinary team to offer emergency management to the child.

Published literature supports the use of urgent external beam radiation treatment in these patients. An old retrospective

study was published by Armstrong *et al.* in 1987, which showed clinical benefits of emergency radiotherapy [26]. Symptom relief was achieved in 85-90% patients within 3 weeks of treatment. Relief of symptoms also enabled improved blood flow through the superior vena cava as well as development of collateral pathways of venous blood flow due to the decrease in pressure on the mediastinal contents. Depending on the duration and total dose of radiotherapy and the performance status of the patient, some mild side effects are expected. Patient may face potential side effects including dyspnea, skin changes, fatigue, cough, pneumonitis, pharyngitis, esophagitis, anemia, leukopenia and anorexia [27]. Standard total dosage of radiation offered with palliative intention varies from three to four Grays per session for five to ten fractions. The total dose and fractionation of radiotherapy can vary according to histopathological diagnosis of malignancy [5]. Overall management plan comprised of steroids therapy through use of prednisolone or dexamethasone, chemotherapy, external beam megavoltage radiotherapy, insertion of an expandable metal stent into the superior vena cava (with or without thrombolysis or anticoagulation) or a combination of any of these treatments [28]. Radiation Treatment remains the most effective treatment for patients suffering from SVCS attributed to either non-small cell lung cancer or caused by any other malignancy. It is crucial to undertake tissue or cytologic diagnosis prior to providing treatment to the patient. This is undertaken to ensure that the treatment field is composed of the tumor with necessary margins for mediastinal and hilar lymph nodes. Patients suffering from non-small cell lung cancer with mediastinal adenopathy as well as without any distant metastases usually have supraclavicular nodes included within the radiation treatment field [29]. Immediate symptomatic relief is being observed after completion of external beam radiation treatment. The intention of treatment is palliation of symptoms for immediate improvement in the clinical condition of SVCS patients [30].

CONCLUSION

In summary, both adults and children who present with the signs and symptoms of SVCS in the emergency room require extra care and multidisciplinary management.

CONFLICT OF INTEREST

Declared none.

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