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The Clinical-Pathological Evaluation of SVC Syndrome in northwest of Iran between 2003-2007

Mohammad Reza Ghaffary*, Shamsi Ghaffari and Maryam Mordadian

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Abstract

Background and objectives: Superior vena cava syndrome results from the obstruction of blood flow through the superior vena cava causing a collection of clinical signs and symptoms including dyspnea, upper limb edema , chest pain and distension of cervical and chest veins. The present study aimed to evaluate the epidemiologic, clinical and pathological characteristics of SVC syndrome in north west of Iran between 2003 and 2007. Materials and methods: A total of 180 cases of SVC syndrome selected for the study. The prevalence of SVC syndrome, sign, symptoms and causes of SVC syndrome were analyzed using SPSS software version 16 via descriptive statistics. Results: From 180 patients with SVC syndrome, 125 were male (69.5%), 55 were female (30.5 %). The mean age of patients were 52.7 ± 18.3 years (male: 51.76 y, female 54.8%). In the majority of patients, super venous syndrome was diagnosed using bronchoscopy and in a few patients, CT guided biopsy and chest X-ray were the main tools of diagnosis. In total, a diagnose of SVC syndrome was made through combining clinical symptoms and paraclinical findings. The commonest cause of SVC syndrome was lung cancer (67.74%) followed by lymphoma (22.58%), other malignancy (3.22%) and tissue diagnosis was not performed in 6.45% of patients. In addition, the underlying pathology of lung cancer was small cell lung cancer (38.9%), squamous cell carcinoma (47.61%) and adenocarcinoma (14.28%). Conclusion:.In contrast to other studies lymphoma and SCC are most common cause of SVC syndrome.

Keywords: Super Vena Cava Syndrome, Malignancy, Bronchoscopy.

Introduction

Superior vena cava (SVC) syndrome was first described in 1757 in a patient with syphilitic aortic aneurysm presenting with a collection of symptoms including dyspnea , upper limb edema , chest pain and distension of cervical and chest veins. This syndrome is now defined as the obstruction of blood flow through the superior vena cava causing a wide array of clinical manifestations including head fullness, upper limb edema, chest pain and distension of cervical, chest veins, dyspnea and cough. SVCS is mainly caused by advanced malignant tumors such as lung cancer and lymphoma and other malignancies. SVC syndrome has been viewed as a relative emergency, which mostly manifests in patients with malignant tumors requiring immediate treatment. In 1954, Schechter reviewed 274 well-documented cases of SVCS and found that 40% were due to syphilitic aneurysms or tuberculous mediastinitis. Since the early reports, these infections have gradually decreased as the primary cause of SVC obstruction. Lung cancer is now the underlying process in approximately 70% of patients with SVCS (1&2). SVC is the major vessel for drainage of venous blood from the head, neck, upper extremities and upper thorax. The SVC is located in the anterior right superior mediastinum surrounded by firm structures including chest pain, trachea, right main bronchus, aorta, pulmonary arteries and lymph nodes. It extends from the junction of the right and left innominate veins to the right atrium, a distance of 6-8 cm. The distal 2 cm of the SVC lies within the pericardial sac and has a relative junction to the pericardium. It is composed at the lower border of the right 1st costal cartilage by the union of left and right brachiocephalic (innominate) veins. Then, the left brachiocephalic vein traverses an approximately 4 cm long behind the first right rib cartilage to join the right brachiocephalic veins, thereby forming superior vena cava (SVC) (Vincent et al., 1999; Schindler & Vogelzang, 1999). The aim of this study was to determine the prevalence of epidemiologic factors involved in the superior vena cava (SVC) syndrome, such as the prevalence of signs and symptoms, the etiologic factors involved, and the rate of survival for patients with superior vena cava syndrome. In addition, given the incidence of lymphoma, this research seeks to address the following question: Does lymphoma have the highest prevalence in the early pathology of the superior vena cava?

Materials and Methods:

This descriptive, retrospective, cross-sectional study was performed based on convenient samples and information in

Mohammad Reza Ghaffary*

Tuberculosis and Lung Disease Research Center, Tabriz University of Medical Sciences, Tabriz, Iran.

Shamsi Ghaffari

Pediatric Cardiologist and Echocardiologist: Tabiz University of Medical Scinces, Azarbayjan, Iran.

Maryam Mordadian

Pediatric Cardiologist and Echocardiologis: Rajaie Cardiovascular, Medical and Research Center, Iran University of Medical Science, Tehran Iran.

patients' medical records referred to the oncology and radiotherapy wards at Imam Khomeini teaching hospital between 2003 and 2007 . The medical records of patients with superior vena cava refereed to the center were taken from the archived records and the data were completed in accordance with the research form for evaluating patients with superior vena cava. The relevant demographic, epidemiological data, primary signs and symptoms, diagnostic and pathological approaches and the number of radiotherapy sessions were obtained from patient's medical records. In addition, data on the survival rate of patients with this syndrome were collected through correspondence or telephone from a limited number of patient companions. However, the process was incomplete due to the lack of access to the address or telephone number of these patients. A total of 180 cases of SVC syndrome were referred to radiotherapy ward. The results were analyzed using SPSS software version 16 via descriptive statistics.

Results

From 180 patients with SVC syndrome, 125 were male (69.5%), 55 were female (30.5 %). The mean age of patients were 52.7 ± 18.3 years (male: 51.76 y, female 54.8%). Clinical symptoms in patients with this syndrome included dyspnea, head fullness and upper limb edema, cough, chest pain and dysphagia. Symptoms of patients with SVC syndrome were as follows: distension of cervical veins, facial edema, distension of chest veins, arm swelling and cyanosis. In the majority of patients, SVCS was diagnosed using bronchoscopy and in a few patients, CT guided biopsy and chest X-ray were the main tools of diagnosis. In total, a diagnose of SVC syndrome was made through combining clinical symptoms and paraclinical findings. Of the 180 patients with this disease, there were 155 cases of pathologic reports in the medical records with the diagnosis of the underlying disease: 105 patients (67.74%) had lung cancer, 35 (22.58%) lymphoma, 5 with acute lymphoblastic leukemia L2 and 10 patients without pathologic diagnosis. Out of 105 cases of SVC syndrome patients with underlying cause of lung cancer, SCLC (small cell lung cancer) was seen in 40 cases, squamous cell carcinoma in 50 cases and 15 cases with adenocarcinoma.

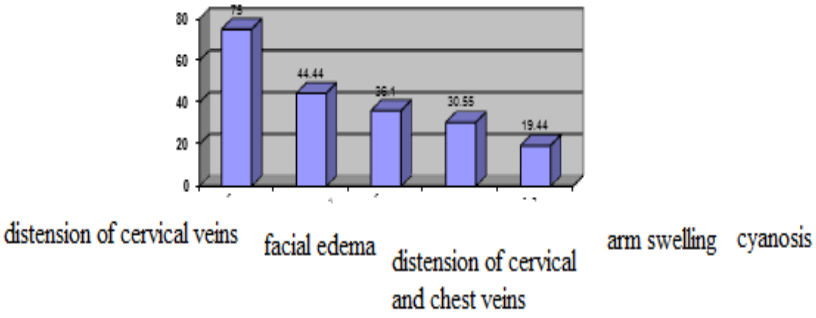

Figure 1. Histogram of the incidence of SVC syndrome symptoms

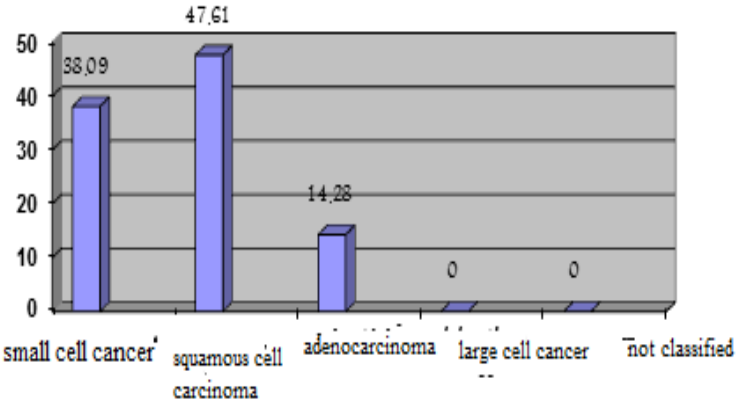

Figure 2. Histogram Pathologic diagnosis of the primary disease causing SVC syndrome

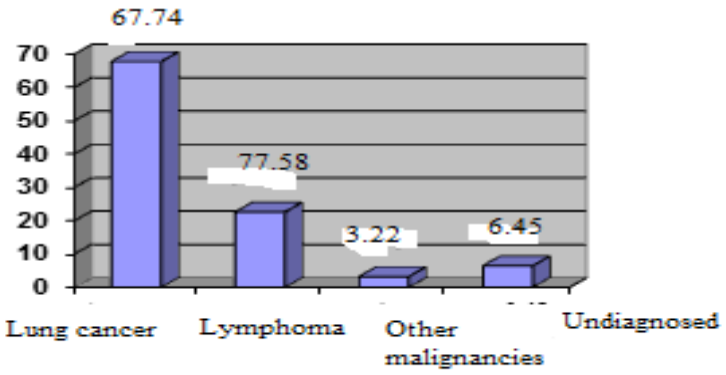

Figure 3. Histogram of subgroups of lung cancer in 21 patients with SVC syndrome

Discussion

According to the results, the most common cause of superior vena cava syndrome is malignant cancers and numerous studies have shown that 78 to 86 percent of cases of SVC syndrome are caused by malignancy. The following table shows the primary pathological diagnosis of SVC syndrome in 415 patients collected by several groups.

Table 1. Primary pathological diagnosis of SVC syndrome

Tissue diagnosis	Belletal	Schraurnagel	Parishetal	Yellinetal	Total
Lung cancer	129 (81%)	67 (63%)	45 (52%)	30 (48%)	271 (65%)
Lymphoma	3 (2%)	10 (9%)	8 (9%)	13 (21%)	43 (8%)
Other malignancies Primary or (metastatic)	4 (3%)	14 (13%)	14 (16%)	8 (13%)	40 (10%)
Non-neoplastic	2 (1%)	16 (15%)	19 (22%)	11 (18%)	50 (12%)
Undiagnosed	21 (31%)				21 (5%)
Total	159	107	86	63	415

In a study by Armstrong et al., on 4100 patients with bronchogenic carcinoma, 99 patients (2.4%) were identified with SVCS. Similarly, Salsali and Cliffton observed SVCS in 4.2% of 4960 patients with lung cancer; 80% of the tumors inducing SVCS were of the right lung. Small cell carcinoma was also found the most common cause of the syndrome and the tumor was found in 38% of patients with lung cancer caused by SVCS. It was also shown that the second most common histologic subtype was squamous cell carcinoma, found in 26% of lung cancer patients with SVCS. In another study, Perez-Soler et al. identified 36 cases (4%) of SVCS among 915 patients with non-Hodgkin's lymphoma treated at the M.D. Anderson Cancer Center. Out of 36 patients, the histologic types associated with SVCS were diffuse large cell in 23 patients, lymphoblastic in 12, and follicular large cell in one patient. In addition, SVCS were observed in 37% of patients with B-cell lymphoma that had sclerosis. HL involves the mediastinum more commonly than any other lymphoma but rarely causes SVCS. Thymoma and germ cell tumors are other malignant mediastinal tumors causing SVCS. Further, metastatic breast cancer is the most common disease leading to SVCS. One study found that the most common metastatic disease that causes SVC syndrome is breast cancer, accounting for 11% of cases. According to the literature, SVCS typically caused by non-malignant cancers is not rare and today benign tumors have grown more rapidly than before, because the rate of peripheral IV cannulation has been increased. Data collected from all hospitals showed that SVCS was among the underlying causes of disease in more than 22% of patients. Obstruction in SVCS is infrequent in the pediatric age group, but there are numerous causative and etiological agents in this age group. The etiological agent of SVCS are primarily therogenic arising after surgery for congenital heart disease (CHD) and atrial fibrillation shunt, ventriculoperitoneal shunt surgery , and insertion of a catheter in the SVCS for intravenous feeding. According to the findings of a study, of the 175 patients with SVCS, the majority of SVCS cases (70%) are caused by surgery or therogenic. Meanwhile, of the remaining 53 cases, 37 cases (70%) were mediastinal tumors, 8 (15%) were benign granuloma and 4 (7.5%) were caused by congenital anomalies of the cardiovascular system. Another study reported that of the 16 patients with superior vena cava syndrome at presentation, eight had non-Hodgkin's lymphoma, four had acute lymphoblastic leukemia, two had Hodgkin's disease, one had neuroblastoma, and one had a yolk sac tumor (Prez & Luther, 1998; Spencer & Ammar, 2007). Intrathoracic goiter is a very rare cause of SVCS syndrome. In a research study between 1984 and 1997 year 4985 patients underwent surgical treatment due to various thyroid gland diseases, among them were 28 (0.6%) patients with intrathoracic goitre, but only in one case (0.002%) the signs of superior vena cava syndrome (SVCS) were observed (Cengiz et al., 1990). Painless aortic dissection caused by a chronic dissecting aortic aneurysm after aortic valve replacement may present itself with superior vena cava syndrome (SVCS), which is one of the very rare causes of the SVCS. A review of the clinical symptoms of patients with SVCS suggests that dyspnea is the most common symptom of the disease, observed in 63% of patients. Headache and swelling of

the face have been reported in 50% of the patients. Other complaints include cough (in 24% of cases), arm edema (in 18% of cases), chest pain (in 15% of cases) and dysphagia (in 9% of cases). Moreover, the common symptoms of SVCS are: dyspnea (63%), headache and facial edema (50%), cough (24%), arthritis (18%), chest pain (15%) and dysphagia (9%). The characteristic physical findings were venous distention of the neck (96%), chest wall (54%), facial edema (46%), plethora (19%), and cyanosis (19%). These symptoms get worse during bending or twisting. Clinical findings in 370 patients with SVCS included cervical dilatation (66%), chest wall distension (54%), facial edema (46%), cyanosis (20%), facial inflammation (19%), and arms edema (14%).

Conclusion:

According to the results, the epidemiological data obtained from this study are consistent with the information in reference books and articles; 69.5% of patients were men and 30.5% were women. (Vincent et al., 1999; Prez & Luther, 1998; Gray, 1987; Qanadli SD EHM, 1999). The mean age of patients with this syndrome was 52 years and 8 months, which is justified by reasoning that cancers are age-related, much more frequent in the old than in the young (Schindler & Vogelzang, 1999; Cengiz et al., 1990; Hayashi et al., 1998). If the average age of patients with this syndrome was considered according to the benign origin, it was expected that the mean age of the subjects were younger. However, since malignancy is the main cause of the disease in all patients with this syndrome and cancer incidence in humans rises exponentially in the final decades of life, so there is no significant difference between the subject's mean age and other research. The most frequent signs and symptoms in SVCS patients include shortness of breath (69.44%), facial swelling and head fullness (44.44%), cough (36.11%), arm edema (30.55%), chest pain (30.55%), and dysphagia (77.2%), which are partially consistent with findings of earlier studies. The signs and symptoms of SVCS syndrome are: dilatation

of cervical veins (75%), facial edema (44.44%), dilatation of chest veins (36.1%), arm edema (30.55%) and cyanosis (19.44%), respectively. These results are to some degree congruent with those results reported by other studies and articles. Regarding the pathology of underlying disease, lung cancer (67.44%) was the leading cause of the disease, which to some extent this is consistent with data of other research. Lymphoma accounts for the 22.58% of the diseases, showing a high proportion than those for other surveys. This inconsistency may be due to the high prevalence of lymphoma in this region compared with other regions of the country. Other malignancies accounted for 22.3% of cases and undiagnosed causes (45.5%). Additionally, small-cell carcinoma (38.09%) and adenocarcinoma (14.28%) were among the main causes of lung cancer, which are completely consistent with our previous findings. However, the squamous cell carcinoma accounted for 47.71% of lung cancer cases in this study, which is much higher than those findings of other studies, requiring further study.

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