Clinical radiological and etiological profile of patient with superior vena cava syndrome in Indian scenario

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SJ YSR conceived idea, YSR MM drafted the study, YSR MLG collected data, YSR SJ did satisfied analysis & interpretation of data, YSR SJ critical reviewed manuscript, All approved final version to be published

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The Authors declares that there is no conflict of interest.

Abstract

Background: Superior vena cava syndrome (SVCS) may occur with an acute or sub acute onset producing a syndrome with characteristic feature including facial edema, plethora, dilatation of vein of chest wall and neck, mild to moderate respiratory difficulty and less commonly headache or more rarely visual disturbance and altered state of consciousness. Literature on different aspect of SVCS from India are lacking. So a study was undertaken at Institute of Respiratory Disease, Jaipur.

Objective: To study clinic-etiological profile of patients with SVC obstruction, etiological distribution of SVCS and to assess the impact of etiological treatment on resolution of SVC obstruction and the symptoms related to it.

Methodology: A prospective type of clinical study was conducted with 26 patient. A detailed clinical history with presenting symptom, sign and radiographic presentation were noted.

Results: Non small cell carcinoma (46.15%) was most common etiology followed by small cell carcinoma (42.30%), lymphomas (7.58%) and fibrosing mediastanitis (3.84%).Commonest radiological presentation was right upper lobe lung mass in 46.15% cases. Bronchoscopy was most productive investigation in establishing the tissue diagnosis.

Conclusion: Etiology of SVCS is static since last three decades. None of solo diagnostic tool is helpful in all cases. However bronchoscopy and relatively safer procedure like FNAC are helpful in quicker diagnosis. These are safer, cheaper and easier alternative to thoracotomy. Furthermore doppler imaging also help in detection of vascular complication like thrombus in great vein, that may be important factor in patient with Intractable SVCS.

Key words: Superior vena cava syndrome; Non small cell carcinoma; Bronchoscopy

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Introduction

Superior vena cava is major drainage vessel for venous blood from head, neck, upper extremities and thorax.[1] It is located in middle mediastinum and is surrounded by relatively rigid structures such as sternum, trachea, right bronchus, aorta, pulmonary artery, and perihilar and paratracheal lymph nodes. It extends from junction of right and left innominate vein to right atrium, a distance of 6-8 cm. It is thin wall, low pressure vascular structure. This wall is easily compressed as it traverse the right side of mediastinum. Suprior vena cava syndrome may occur with an acute or sub acute onset producing a syndrome with characteristic feature including facial edema, plethora, dilatation of vein of chest wall and neck, mild to moderate respiratory difficulty and less commonly headache or more rarely visual disturbance and altered state of consciousness.[2] Spectrum of SVCS has changed since the original etiological classification and reviewed by Hudson in 1978. It has changed from infectious etiology to non infectious due to availability of effective chemotherapy. With increase in smoking trend carcinoma lung has become an important cause of SVC obstruction contributing 80-85 % of cases

followed by lymphoma and metastasis of non pulmonary origin. The introduction of transvenous catheter, central venous line and swan ganz catheter has seemingly created new benign cause of SVC obstruction.[3] Other less common causes are mediastinal tubercular lymphadenitis[4], fibrosing mediastinitis[5], endocardial defibrillation and pace maker leads.[6] Paucity of literature on different aspect of SVCS from India prompted us to take this prospective study.

Methodology

This prospective clinical study was conducted in department of respiratory medicine, SMS Medical college Jaipur between 2013 to 2015. Patient from age group between 30 to 70, all sex and both urban and rural back ground were included to make sample more representative. A detailed clinical history with presenting symptom, sign and radiographic presentation was taken. Apart from general symptoms patients were also looked for paraneoplastic syndrome, vascular airway and nerve compression. A thorough general physical examination and respiratory system was done along other systemic examination. Diagnosis of SVCS was established by clinical examination followed by imaging studies and later on etiology of SVCS was established with help of tissue diagnosis. Usual approach was applied as shown in table 1.

Results

Our study included 26 patients with SVCS, age of subjects ranged from 35 to 80 with mean age of 54± 11. There were only 3 female among.[26] More than 2/3 of patient (73.07%) had symptoms of 1 – 6 month duration. 19.07% (5) of study subject had illness of less than one month, remaining 7.69% (2) were symptomatic from more than 6 month duration. Mean duration of illness is around 2 to 3 month. Maximum number of patients (50%) were agricultural farmer followed by masonry worker (7.69%), office worker (7.692%) and house wife (7.69%), six patients

Algorithm adopted for Diagnosis

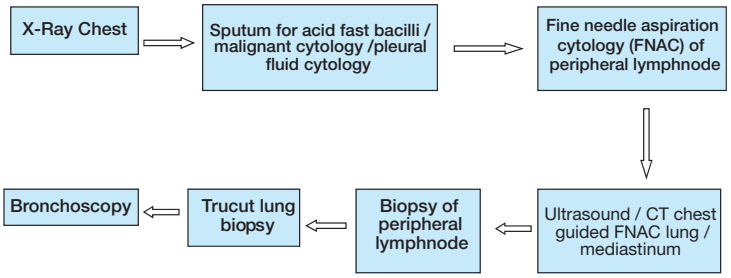

Table 1: Signs and Symptoms of SVCs

S.No	Observation	No. of Patients
1	Swelling	
	Face	26
	Neck	26
	Chest	23
	Arms	12
2	Dilated tortuous and non pulsatile veins	
	Neck	26
	Thorax	20
	Abdominal wall	26
3	Conjunctival Congestion	2
4	Hoarsness of Voice	26
5	Headache	16
6	Dysphagia	6
7	Facial Flushing	10
8	Dizziness	4
9	Visual Disturbance	2

belonged to other occupation. Majority of patients were current smoker (84.61%) while 11.53% were non smoker. Only one patient was ex-smoker. Cough was most common mode of presentation (92.31%) followed by chest pain (61.54%). Cough was equally distributed in small and non small carcinoma while chest pain more common in small cell carcinoma (81.82%) than non small cell carcinoma (41.67%). Expectoration was found more common in non small carcinoma (58.33%) than small cell type (27.27%). Hemoptysis and dysnoea were distributed equally between small and non small type. No significant relationship was observed between symptom and type of malignancy. Facial and neck swelling were present in all patients followed by chest and arm swelling in 88% and 46.15% respectively. Headache was found in16 patients (61.54%), 10 of these patient had small cell carcinoma. Facial flushing was seen in 10 patients. Visual disturbance was found only in one patient (Table 1). There was no significant association between type of malignancy and specific symptoms. Clubbing was found in 50% of patients, it was almost equally distributed in both small and non small carcinoma (Table 2). Lymphadenopathay was more common in small cell carcinoma (45%) than non small type (25%) (Table 3). Dilated vein below umbilicus-(post azygous) was seen only in 3 cases, 2 of them were of small cell carcinoma. Commonest radiological presentation was right upper lobe lung mass 12 (46.15%)of total patients followed by right upper lobe collapse in 6 (26.09%),mediastinal mass was found in only 6 cases. Out of them one had benign etiology while two had lymphoma. Pleural effusion was seen in 4 patient with other radiological finding (Table 4). Doppler imaging of both in nomminate vein & SVC showed intraluminal thrombus in 8 of 26 patients, while in 18 patients, signs of external SVC compression by media stinal lymph node or primary mass lesion were present (Table 5). Bronchoscopy was commonest mode to establish the diagnosis in 11 patient (42. 31) followed by FNAC lung and lymph node 6 (26.09%) and 4(17.39%) respectively. Open lung biopsy was needed in single case of benign cause of fibrosing media stinitis. On histological classification non small cell carcinoma 12 (46.15%) was most common etiology followed by small cell carcinoma 11 (42.30%) (Table 6). Lymphomas was found in 2(7.58%) patient. Single patient (3.84%) was diagnosed as fibrosing media stanitis.[8] Patients could survive beyond 6 month of follow up. More than half of study patient died during 6 month follow up. Follow up was not available in 4 patients while one patient refused for treatment.

Discussion

This was a prospective study showing clinico radiological and etiological profile of patients with SVCS. Diagnosis of syndrome was often made straight forward. Present study not only describes clinical radiological profile of SVCS but also tells about determinants of survival. Mean age of patient was 54±11 years (range 35-71 year). More than 50% of cases were of more than 55 year, it was similar to study.[7] Probable explanation for this age distribution is that the most common underlying etiology of SVCS was bronchogenic carcinoma which is more often in older age group. Only 2 case were below age of 45

Table 2: General physical examination

S.No	Observation	No. of patient
1	Clubbing	13
2	Lymph node	9
3	Pallor	11
4	Voacal cord palsy	2

Table 3: Mode of etiological diagnosis and yield in SVCS

S.No	Mode of diagnosis	Type of disease			Total
		Bronchogenic carcinoma (n 23)	Benign n1	Others n2	
1	Bronchoscopy	11(47.83%)	0(0.00%)	0(0.00%)	11(42.31%)
2	FNAC lung	6(26.09%)	0(0.00%)	0(0.00%)	6(23.08%)
3	FNAC lymph node	4(17.39%)	0(0.00%)	0(0.00%)	4(15.38%)
4	Lung biopsy	2(8.69%)	0(0.00%)	0(0.00%)	2(7.69%)
5	Lymph node biopsy	0(0.00%)	0(0.00%)	2(100%)	2(7.69%)
6	Open lung biopsy	0(0.00%)	1(100%)	0(0.00%)	1(3.85%)

Table 4: Radiological presentation and type of disease

S. No	Radiological presentation	Type of disease			Total
		Bronchogenic carcinoma (n 23)	benign	other	
1	Right upper lobe collapse	6 (26.09%)	0(0.00%)	0 (0.00%)	6 (23.08)
2	Pleural effusion	4 (17.39%)	0 (0.00%)	0 (0.00%)	0 (15.38)
3	Right lung mass	12 (52.17%)	0 (0.00%)	0 (0.00%)	12 (46.15)
4	Hilar prominance	4 (17.39%)	0 (0.00%)	0 (0.00%)	4 (15.38)
5	Mediastinal Mass	3 (13.04%)	1 (0.00%)	2 (66.67%)	6 (23.08)

Table 5: Level of obstruction and Doppler study of neck vein

S.No	Level of SVC obstruction	Preazygos	Post azygos	Total	
1		23	3	26	
	Doppler study showing		Type of malignancy		
			Small cell	Non small cell	Total
2	Presence of thrombus		2	6	8
3	No thrombus		9	6	15

Table 6: Histological Diagnosis and duration of survival

S.No	Disease	Duration (in month)			Total
		<3	3-6	>6	
1	Bronchogenic carcinoma	4 (22.22)	10 (55.56)	4 (22.22)	18 (100)
2	Benign causes	0 (0.00%)	0 (0.00%)	1 (0.00%)	1 (100%)
3	others	0 (0.00%)	0(0.00%)	2 100)	2(100)
4	Total	4 (19.05)	10(47.62)	7(33.33	21(100%)

year at diagnosis and both were female. [1] out of these 2 was of nonhodgkin lymphoma while other case was non small cell carcinoma. Etiology for early onset of disease likely due to environmental exposure to carcinogen or genetic predisposition. Our study dominated by male patients . 23 out of 26 were male. It may due to more smoking in male or under utilization of health service by female.Similar observation was made by Perez R et al.[8] 23 out of 26 patients were smoker. Only three patients were non smoker. This may be due to fact that SVCS is more commonly found in patients of bronchogenic carcinoma associated with smoking. Although detailed occupational history was taken but we did not find occupational exposure to carcinogens. Most of the patients were farmer and labourer with history of smoking .Most common respiratory symptom in present study was cough (80.76%) followed by chest pain (65.38%) with similar observation in study by Little et al.9While some other studies showed that cough was less common but it is not clear that these studies considered new onset of cough or ignored existing cough as usual feature of chronic smoker.[10,11] Dyspnea in bronchogenic carcinoma is multifactorial and it not caused by simple obstruction of SVC. Factor like associated pleural effusion, pericardial effusion, tracheal obstruction and level of bronchial obstruction are other determinants of dyspnea in patients with SVCS. Probably difference in under lying extent of disease may account for difference in prevalence of dyspnea in patient of SVCS. In our study dyspnea was observed in 9 patient (34.61%) while was reported as predominant symptom 62.79%12 and 83.33 %13.This may be because only four of our study patients had pleural effusion while none of patient had tracheal compression and endobronchial growth was also seen distal to carina. Similar discrepancy was noted in hemoptysis also. It was higher in our study (42.30%) as compare to little et al (11.90%). This may be due to more number of patient having endobronchial growth in our study. In specific symptoms 7 out of 26 had arm edema while facial and neck swelling was found in all 26 patient. Headache was next common symptom in 38.46%. Similar observation was made by different studies.[11-14] Obstruction of SVC lead to inadequate drainage through sub clavian and internal and external jugular vein. Arm edema was less possibly because of better collaterals for subclavian vein which help in relatively better drainage as compared to jugular veins. In our study major constitutional symptom was weight loss (76.92%) followed by anorexia (61 .53%). Similar result was observed by scaratino etal15and little et al.[9] This is because of the fact that dysphagia is symptom of posterior mediastnal mass whereas in our study only 6 patient had involvement of posterior mediastinum. Regarding

common signs present in our study, clubbing and lymphadenopathy were present in 46.15% and 34.61% respectively. This may be due to etiologic factor of bronchogenic carcinoama. Doppler ultrasound showed presence of thrombus in SVC innominate in 8 patient. This is likely due to stagnation of blood flow in vessel rather hypercoagulable state because 6 patient having thrombus in situ were of non small carcinoma. Hypercoagulable state is manifestation of small cell variety. Among radiological finding lung mass was seen in 46.15% cases followed by collapse and mediastinal mass in 23.08% similar observation was made in another study 16 out of 6 mediastinal mass, 2 patient had lymphoma while one has fibrosing mediastinitis. Bronchoscopy was most productive investigation in establishing the tissue diagnosis. It was done in 11 of 26 patient and histological diagnosis was obtaining in all these 11 patient. This is perhaps because most of lesion were centrally located . This is consistent with available literature.[17] Less invasive means like FNAC peripheral lymph node or lung mass also proved important tool for diagnosis in 6 and 4 patient respectively. Earlier studied have used mediastinoscopy 19 and thoracotomy in large number but with availability of better image guided modality , these are not used very often now. Benign cause was found in only one case. Among malignant etiology non small cell carcinoma (46.15%) followed by small cell carcinoma 42.30%. Only one patient of adenocarcinoma was diagnosed. Rest 2 patients were of lymphoma. Study by davenport D etal18 state that most common etiology was bronchogenic carcinoma 74.28% followed by lymphoma (20.63%). In another study by Yellin et al 14 47.61 % and 20.63% were of bronchogenic carcinoma and lymphoma respectively. While 17.46% had benign etiology. There was selection bias in our study as no case of placement of central line and pacemaker induced thrombosis or secondary to vascular disorder were not found. Our study was respiratory centre based study which only include adult patient with predominantly chest symptoms. Figures of increased bronchogenic carcinoma would have been definitely diluted if there would have no selection bias . Therefore the relative frequency of different etiology can not be extrapolated for community. There was no significant difference observed between age ,sex respiratory, constitutional and specific symptoms, radiological presentation, level of obstruction duration of survival with malignancy ($p<.05$). Prevalence of various histological type of bronchogenic carcinoma varies in different geographical areas. In our study most common variety was non small cell carcinoma followed shortly by small cell carcinoma. Most studies in other part of word showed non small cell carcinoma as predominant etiology of SVCS. No Indian study available to best of our knowledge is available till date. More studies are needed to answer to prevalence of lung malignancy causing SVCS. Prognosis of SVCS depend upon underlying etiology, median survival was 100±48 days. Six month survival was only 22.22% with bronchogenic carcinoma. Lymphoma patient could survived beyond 6 month of treatment . In a study by Urban etal 20 small cell carcinoma treated with chemotherapy , median survival time with or without SVCS was 40 weeks. In our study less median survival was probably because many of our patients had stage 4 disease . Those who died before three 3 month were having mainly brain metastasis . Further survival period of 5 patient was not available . Had it been included, possibly median survival could have longer. It is clear from our study that lesser the duration of illness lesser the survival. As out of 7 patients who survived more than 6 month, 6 patients had illness of more than 3 month. Prognosis of smokers was poor as compared to non smoker. Presence of thrombus in vessel was poor prognostic factor with SVCS but these are statistically non significant ($P<.05$)

Conclusion

Etiology of SVCS is static since last three decades. None of solo diagnostic tool is helpful in all cases. However bronchoscopy and relatively safer procedure like FNAC are helpful in quicker diagnosis to begin treatment. These are safer, cheaper and easier alternative to thoracotomy. Furthermore doppler imaging also help in detection of vascular complication like thrombus in great vein, that may be important factor in patient with Intractable SVCS.

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