

1 The Role of Multislice CT in Evaluation of Superior Vena Cava 2 Syndrome

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5 *Received: 8 December 2013 Accepted: 4 January 2014 Published: 15 January 2014*

6

7 **Abstract**

8 The superior vena cava (SVC) syndrome is a clinical entity caused by obstruction of the
9 superior vena cava by infiltration, compression or thrombosis. Cancer is the most common
10 underlying cause of superior vena cava obstruction. The incidence of catheter-induced
11 superior vena cava obstruction is rapidly increasing. Fibrosing mediastinitis and Behcet
12 disease are rare causes of SVC syndrome. Clinical presentation of SVC syndrome may include
13 cough, dyspnea, dysphagia, and swelling or discoloration of the neck, face and upper
14 extremities. Aim of this study is to evaluate the role of Multislice CT in study of superior
15 vena cava obstruction syndromes and assessment of collateral circulation in different causes of
16 superior vena caval obstruction

17

18 *Index terms—*

19 **1 Introduction**

20 The superior vena cava (SVC) syndrome is a clinical entity caused by obstruction of the superior vena cava by
21 infiltration, compression or thrombosis. Although clinical symptoms of the disorder were first described in 1757 in
22 a patient with a syphilitic aneurysm of the ascending aorta, vascular causes are now rare and approximately 90%
23 of cases are associated with a cancerous tumor that is compressing the superior vena cava, such as bronchogenic
24 carcinoma including small cell and non-small cell lung carcinoma, Burkitt's lymphoma, lymphoblastic lymphomas,
25 acute lymphoblastic leukemia (rare), and other acute leukemias (Krimsky et al, 2002). Tuberculosis has also
26 been known to cause superior vena cava syndrome (SVCS). SVCS can be caused by invasion or compression by a
27 pathological process or by thrombosis in the vein itself, although this latter is less common (approximately 35%
28 due to the use of Benign Causes of Superior Vena Cava Obstruction Iatrogenic superior vena cava obstruction the
29 incidence of catheter-induced superior vena cava obstruction is rapidly increasing e.g. large central venous
30 catheters, such as dialysis catheters, Hickman catheters, and parenteral nutrition catheters (3). Fibrosing
31 mediastinitis is a rare histologically benign disorder caused by proliferation of collagen tissue and fibrosis in
32 the mediastinum (4). Behcet disease is a rare systemic disease in which superior vena cava stenosis or occlusion
33 may result (5).

34 Clinical presentation of SVC syndrome: SVC syndrome is caused by gradual compression of the SVC, leading to
35 edema and retrograde flow, but it can also be caused more abruptly in thrombotic cases. Symptoms may include
36 cough, dyspnea, dysphagia, and swelling or discoloration of the neck, face and upper extremities. Often, collateral
37 venous circulation causes distension of the superficial veins in the chest wall. Although SVC syndrome is usually
38 a clinical diagnosis, plain radiography, computed tomography (CT) and venography are used for confirmation.
39 (6).

40 CT Findings: CT can diagnose SVC affection in SVC syndrome and can detect subclinical superior vena cava
41 obstruction in patients who are relatively asymptomatic (Yu JB et al, 2008). Regardless of its cause, the CT
42 diagnosis of superior vena cava obstruction includes lack of opacification of the superior vena cava, an intraluminal
43 filling defect or severe narrowing of the superior vena cava, and visualization of collateral vascular channels ??Eren
44 et al, 2006). MDCT, with its multiplanar and 3D imaging capabilities, allows thorough anatomic delineation of

7 DISCUSSION

45 the various collateral pathways diverting the blood from the site of obstruction. Although axial images allow
46 evaluation of potential causes of superior vena cava obstruction, such as a mediastinal mass, MDCT provides
47 information about the level and degree of superior vena cava obstruction, the length of the affected segment,
48 and the presence or absence of intraluminal clot distal to the obstruction, thereby allowing the interventional
49 radiologist to choose the optimal treatment option. ??Plekker et al, 2008).

50 2 II.

51 3 Aim of the work

52 4 Patients and methods

53 5 a) Patients

54 This study involved 20 patients; 13 males and 7 females, age range 15-69 (average of 39.3years). Cases were
55 referred from the chest department to the radiology department in Kasr Al-Aini for MSCT of the chest.

56 Twelve patients presented clinically with mediastinal syndrome; of whom six cases were biopsy proved central
57 Bronchogenic carcinoma, five were known cases of Behcet disease and one case with unknown etiology.

58 Two cases had right upper limb DVT. Two cases had lymphoma and one case was surveyed for chest metastases.
59 One case had fever of unknown origin after chemotherapy. One case with antiphospholipid antibody syndrome
60 where echo detected calcified right atrial thrombus. One case with bilateral pleural effusion. (See table 1).

61 6 Results

62 See the summary of MSCT results in tables 2-4.

63 7 Discussion

64 Multi-detector row computed tomography (CT) with 2D and 3D reconstructed images provides a unique
65 perspective on thoracic anatomy and disease. Multidetector row CT allows shorter acquisition times, greater
66 coverage, and superior image resolution (7).

67 In vascular imaging, this would provide image quality that equals or surpasses that of conventional angiography.
68 Its use has expanded to aid in diagnosis and surgical planning, as it is reliable in depicting clot, thoracic
69 vasculature and may also be used to evaluate thoracic venous anomalies and to plan therapy (8).

70 Multi-detector row CT has been used for venous angiography. Superior vena cava obstruction, often related to
71 tumor, is frequently seen at multidetector row CT. This technique may be used to establish the extent of tumor
72 involvement and document the extent of collateral vessel formation (9).

73 MDCT, with its multi-planar imaging capabilities, allows thorough anatomic delineation of the various
74 collateral pathways diverting the blood from the site of obstruction. Although axial images allow evaluation
75 of potential causes of superior vena cava obstruction, such as a mediastinal mass, if intervention is warranted,
76 MDCT provides valuable information about the level and degree of superior vena cava obstruction, the length of
77 the affected segment, and the presence or absence of intra-luminal clot distal to the obstruction, thereby allowing
78 the interventional radiologist to choose the optimal treatment option (10).

79 Superior vena cava syndrome usually presents more gradually with an increase in symptoms over time as
80 malignancies increase in size or invasiveness (11).

81 The severity of the syndrome depends on the rapidity of onset of the obstruction and its location (12). (see
82 table 5). SVC syndrome can lead to the formation of downhill esophageal varices and pleural effusion. Numerous
83 case reports have described pleural effusions in conjunction with the SVC syndrome. These effusions occur
84 in 60% of SVC syndrome cases. The effusions are small, usually occupying less than one-half of the affected
85 hemithorax, and occur approximately equally on either side or bilaterally. Although previously thought to be
86 largely transudates, a large case series found that 18% of the effusions were chylous, with the remainder being
87 exudates. None of the effusions sampled in the series were transudates. Occluded lymphatic flow from increased
88 hydrostatic pressure in the SVC and left brachiocephalic vein probably contributes to the development of chylous
89 pleural fluid. The pathophysiology of the exudative effusions, however, remains unknown. Many factors, including
90 diuresis, small pulmonary emboli, and the underlying inflammatory or malignant condition all likely contribute.
91 Chylous or exudative pleural effusions occur in most patients with SVC syndrome. The effusions are usually
92 small and resolve upon correction of the underlying SVC obstruction. ((13).

93 Five to 10 percent of cases of SVC obstruction are due to benign causes. Most result from invasive monitoring
94 techniques, such as the placement of central venous lines, Swan-Ganz catheters, and interventional techniques,
95 such as the placement of pacemakers and central venous catheters for chemotherapy (14). Thrombi in the SVC
96 were detected by transesophageal echocardiography in 30% of patients who had single lumen silicone rubber
97 hemo-dialysis catheters. Polyvinyl chloride, polyethylene and teflon catheters are associated with increased
98 thrombogenicity, as compared to silicone rubber. Furthermore, the SVC stenosis may be induced by persistent
99 trauma of the endothelium by the catheter tip and the higher blood flow during dialysis hours ((17).

100 Fibrosing mediastinitis (fig ??)-it is a rare benign disorder caused by proliferation of acellular collagen
101 and fibrous tissue within the mediastinum. Although many cases are idiopathic, many (and perhaps most)

102 cases in the United States are thought to be caused by an abnormal immunologic response to Histoplasma
103 capsulatum infection. Affected patients are typically young and present with signs and symptoms of obstruction
104 or compression of the superior vena cava, pulmonary veins or arteries, central airways, or esophagus. There
105 may be two types of fibrosing mediastinitis: focal and diffuse. The focal type usually manifests on computed
106 tomographic (CT) or magnetic resonance (MR) images as a localized, calcified mass in the paratracheal or sub-
107 carinal regions of the mediastinum or in the pulmonary hilae. The diffuse type manifests on CT or MR images
108 as a diffusely infiltrating, often non-calcified mass that affects multiple mediastinal compartments. CT and MR
109 imaging play a vital role in the diagnosis and management of fibrosing mediastinitis (18).

110 Behçet disease (fig 6,7) -Behçet disease is a multisystem disease of unknown etiology. The syndrome carries
111 the name of the Turkish dermatologist Hulusi Behçet, who, in 1937, described a syndrome of recurrent aphthous
112 ulcers, genital ulcerations, and uveitis leading to blindness. Although the cause of the disease is still unknown, it
113 has become recognized as a multisystemic inflammatory disease (19). Enlargement of the thyroid gland (goiter)
114 (20). The aim of this study was to provide evidence that MSCT is useful in patients with superior vena caval
115 obstruction, detecting the level and the cause of obstruction.

116 Multi-slice CT scans were reviewed in 20 patients referred from chest department to radiology department in
117 Kasm Al-Aini.

118 Twelve patients presented clinically in our study with mediastinal syndrome, Two cases had right upper limb
119 DVT. Two cases had lymphoma and one case was surveyed for chest metastases. One case had fever of unknown
120 origin after chemotherapy. One case with antiphospholipid antibody syndrome where echo detected calcified right
121 atrial thrombus. One case with bilateral pleural effusion

122 The twelve patients presented clinically in our study with mediastinal syndrome, six cases were biopsy proved
123 central Bronchogenic carcinoma, five were known cases of Behçet disease and one case with unknown etiology.
124 These findings are more or less consistent with the study made by Bagheri et al, 2009 which stated that among 45
125 patients with SVCS, their diagnostic pathological reports showed 26 (57.8%) were compatible with bronchogenic
126 carcinoma (small cell lung cancer [SCLC] in 19 cases and non-SCLC in 7) which was the most common etiology of
127 superior vena cava syndrome. Lymphoma was reported in 14 cases (31.1%), (12 of non-Hodgkin's lymphoma and 2
128 of Hodgkin's lymphoma), and germ cell tumor and malignant thymoma were observed in 3 (6.7%) and 2 (4.4%) of
129 patients (21). In our study, 5 Behçet cases revealed partial SVC obstruction and eminent mediastinal collaterals,
130 6 cases with central infiltrating Bronchogenic carcinoma revealed partially infiltrated SVC with few collaterals
131 noted in 5 cases and eminent chest wall collaterals in one case (the case was a follow up case of Bronchogenic
132 carcinoma after radiotherapy), 1 Case of mediastinal syndrome of unknown etiology showed diffuse affection of
133 the mediastinum with soft tissue density lesion causing effacement of SVC and marked attenuation of the right
134 pulmonary artery ; a picture suggesting fibrosing mediastinitis with eminent chest wall collaterals, 2 cases with
135 left upper limb DVT seen extending into left brachiocephalic vein and normal contra-lateral brachiocephalic vein
136 and SVC, 2 cases with lymphoma showed huge anterior mediastinal mass, displaced and compressed SVC with
137 no collaterals, one case of chest assessment for metastasis revealed lung, pleura and mediastinal metastases with
138 SVC distortion and partial infiltration and no collaterals, one case of fever of unknown origin after chemotherapy
139 showed partial SVC obstruction with a heterogenous thrombus with air density, enlarged SVC, no collaterals, one
140 case of Antiphospholipid antibody syndrome revealed partial SVC obstruction at lowest portion , no collaterals,
141 one case of bilateral pleural effusion revealed partial SVC thrombus in relation to CV-Line catheter, no collaterals.

142 To conclude, the rate of obstruction and its location greatly affects the severity of symptoms of SVCS, this
143 depends on the development of the collateral circulation. Meaning that when the onset of obstruction is slow (as
144 in benign causes of obstruction e.g. Behcet's disease and with central venous catheters), the collateral circulation
145 will have time to distend and accommodate the increased blood flow. On the other hand, diseases causing rapid
146 onset of obstruction (malignant tumors), produce more severe symptoms because the collateral veins will not
147 have time to distend. L Longmore et al, 2007 suggested that the general recruitment of venous collaterals over
148 time may lead to remission of the syndrome although the SVC remains obstructed (24).

149 Wilson, 2007 stated that not all cases of SVC obstruction must present with SVCS (e.g. facial edema, venous
150 distension in the neck, upper limb edema) and that there is 10% of cases may have radiographic SVC obstruction
151 in absence of symptoms. This is consistent with our study which includes twelve of the twenty patients presented
152 clinically with mediastinal syndrome, two cases had right upper limb DVT, two cases had lymphoma and one
153 case was surveyed for chest metastasis. One case had fever of unknown origin after chemotherapy, one case with
154 anti-phospholipid antibody syndrome and one case with bilateral pleural effusion (25).

155 VI.

156 8 Summary and Conclusion

157 Superior vena cave syndrome (SCVS) is a constellation of signs and symptoms resulting from obstruction of
158 the SVC or its major tributaries by intraluminal occlusion or by extrinsic compression and/or invasion from
159 malignant and benign diseases.

160 SVC obstruction leads to increased venous pressure and edema of the neck, arms, upper chest, and head
161 causing increased intracranial pressure. Patient may present with headache, syncope or presyncope, nausea and
162 vomiting, hoarseness, dysphagia, cough, dyspnea and chest pain. Severity of symptoms depend on the time
163 course of obstruction. As obstruction develops, venous collaterals develop to find alternate pathways for venous

8 SUMMARY AND CONCLUSION

164 return to the right atrium. In the post-antibiotic era malignancy remains the commonest etiology. Lung cancer
165 is the commonest malignancy. SVCS is most common with small cell lung cancer as it grows rapidly in central
166 airways. CT chest is the investigation of choice which provides information on location, possible etiology, extent
167 of collaterals and guide biopsy attempts.

168 The advent of multi-detector CT has revolutionized imaging of the mediastinal vascular structures. In
169 comparison to single-detector helical CT scanners, multi-detector scanners not only provide faster speed, greater
170 coverage, and improved spatial resolution, but also have the unique ability to create images of thick and thin
171 collimation from the same data set.

172 One of the greatest benefits of this new technology is the improved quality of two-dimensional (2D) multi-planar
173 and three-dimensional (3D) reconstruction images.

174 MSCT can easily prove or exclude the affection of SVC by partial or complete obstruction, the development of
175 collateral circulation as well as detecting the cause of obstruction whether thrombosis, compression or infiltration
and its extent. ¹



Figure 1:

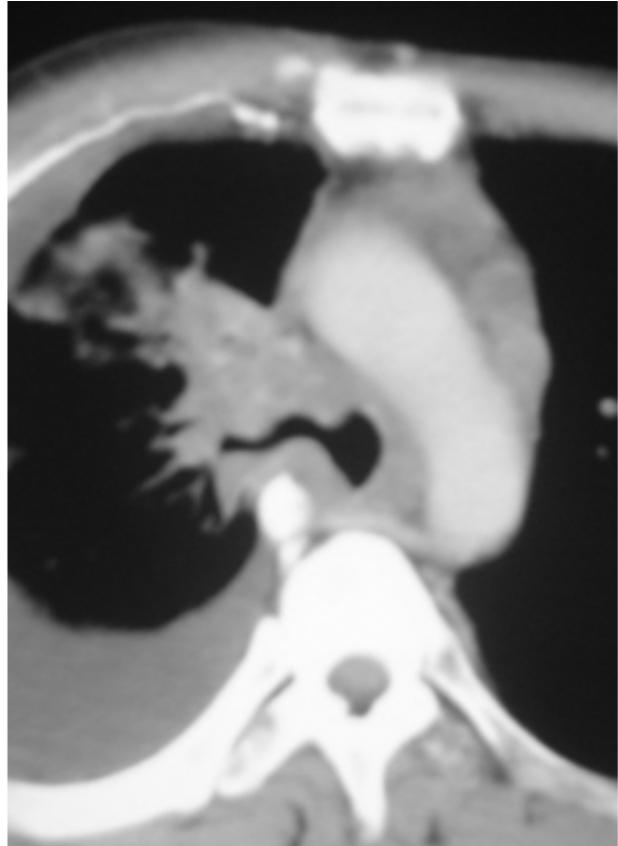
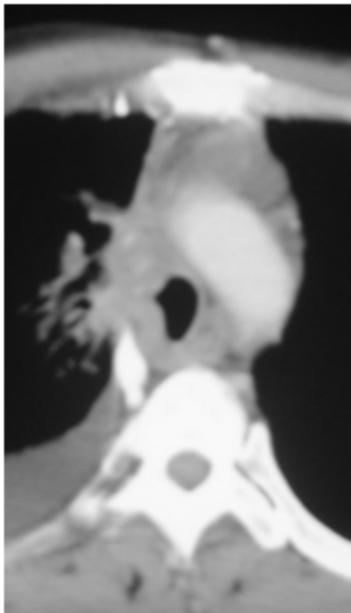


Figure 2:

176

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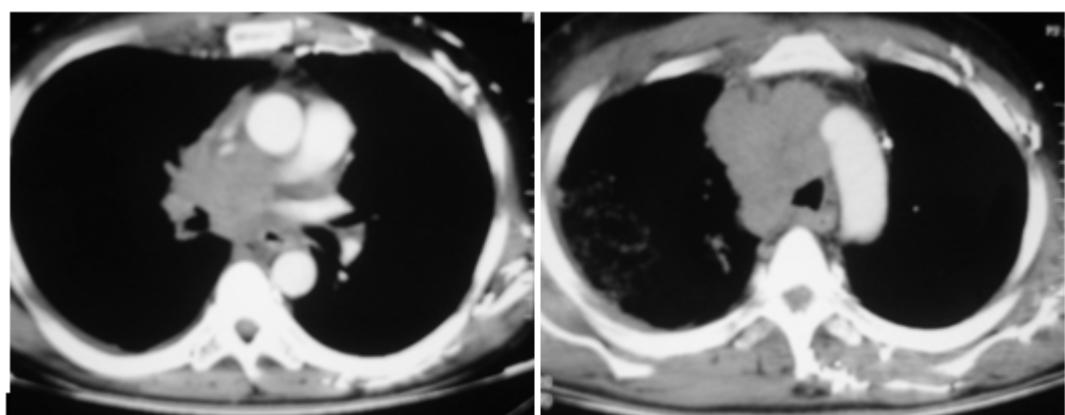


Figure 3:

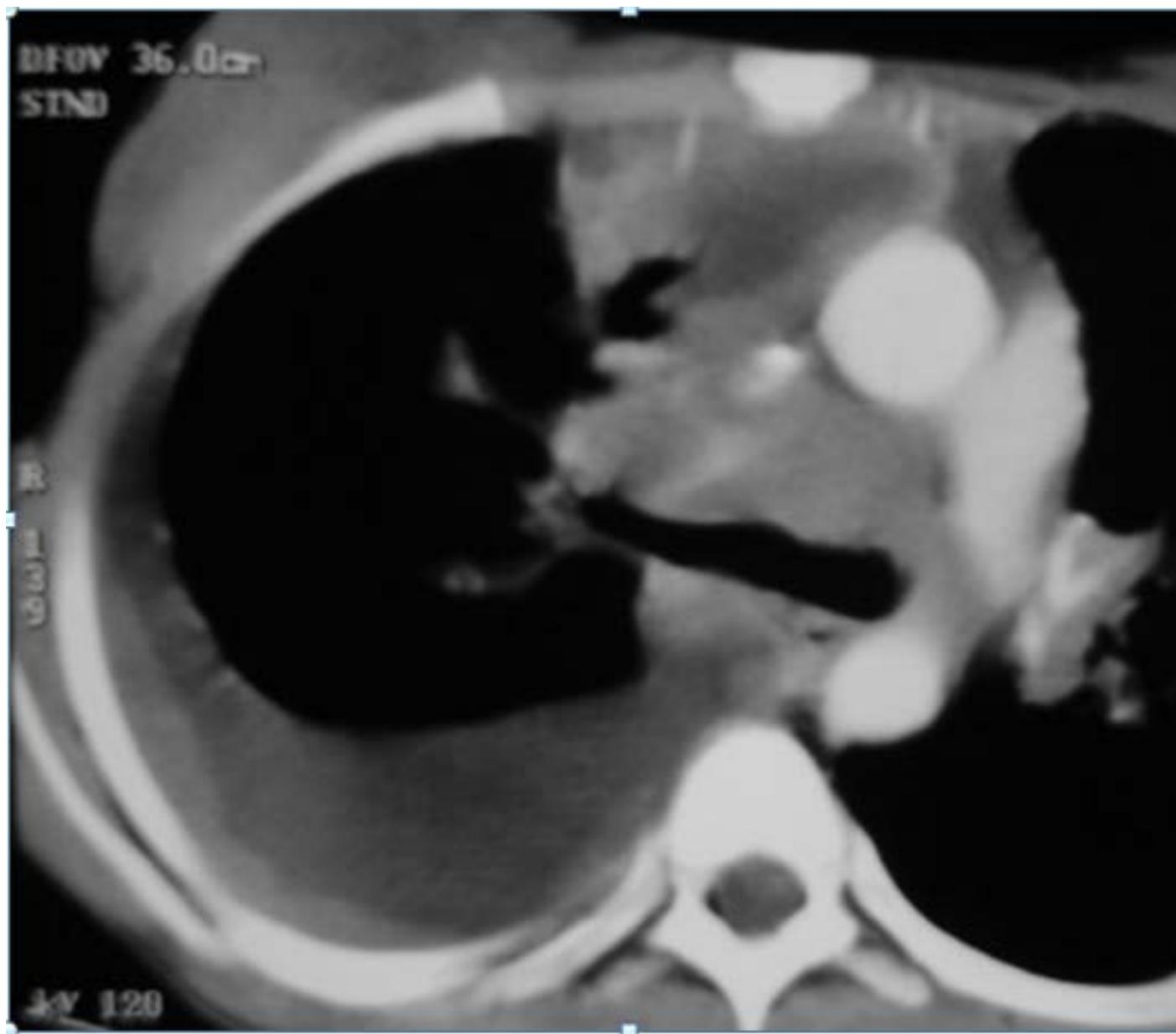


Figure 4:

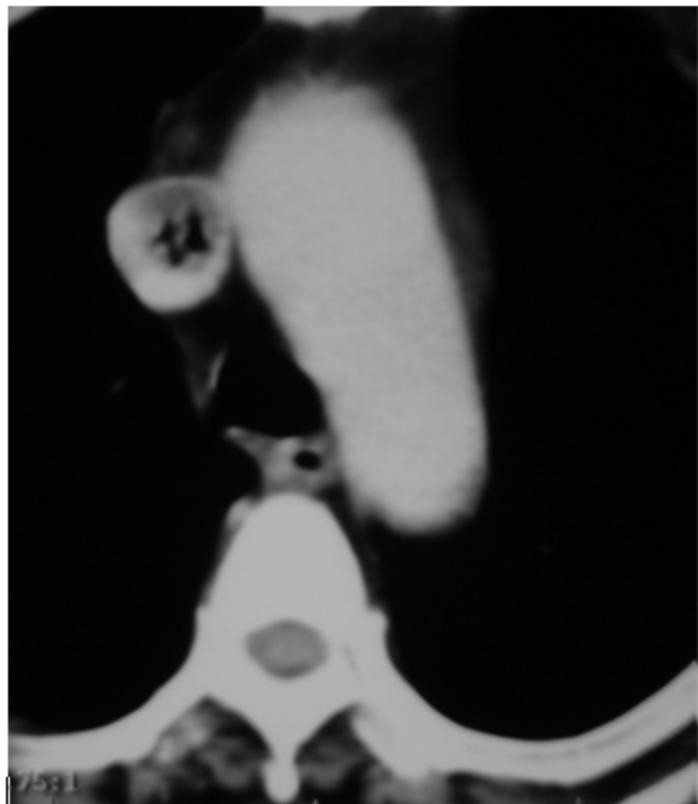


Figure 5:

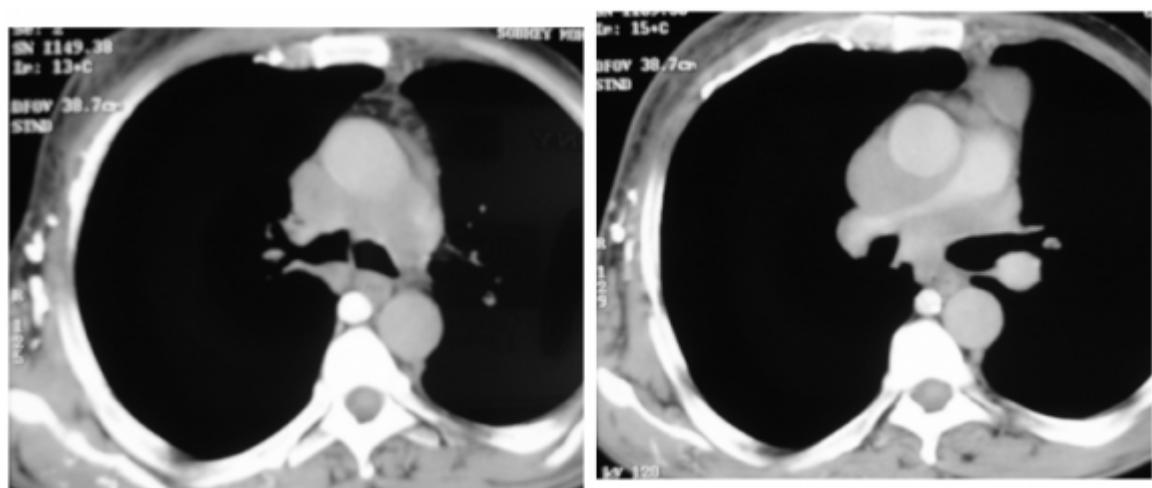


Figure 6:

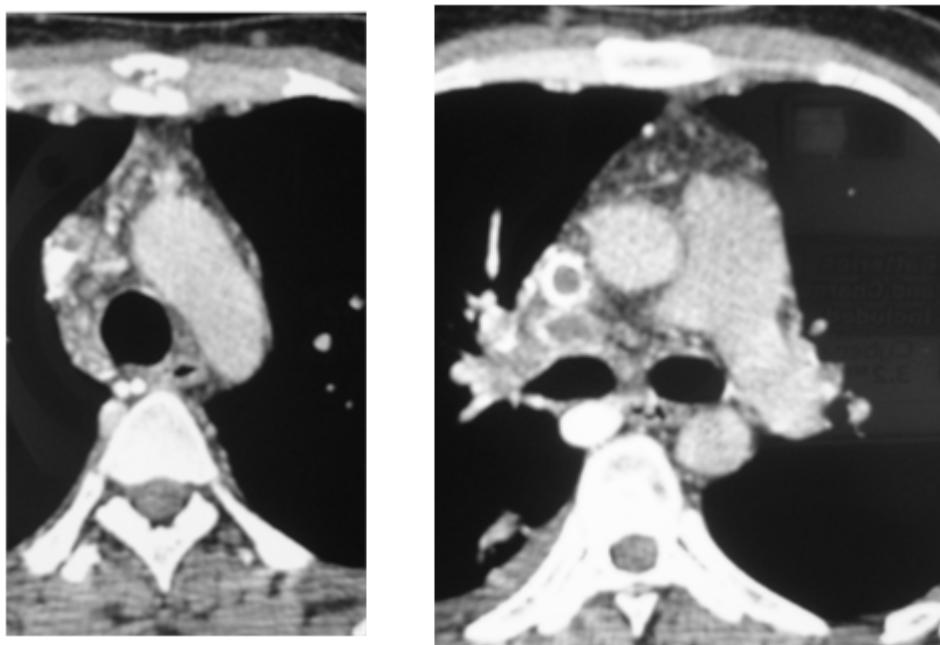


Figure 7:

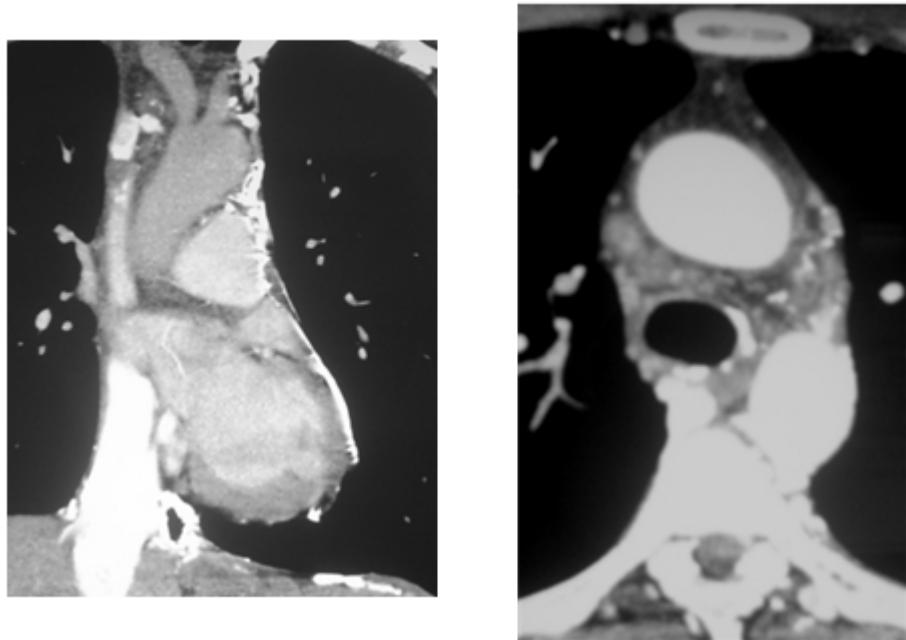


Figure 8:

8 SUMMARY AND CONCLUSION

1

Showing patients' presentation

R Referred case

Mediastinal syndrome

Number of patients

12

Upper limb DVT

2

Chest assessment in case of lymphoma

2

Chest assessment for metastases

1

Fever of unknown origin after chemotherapy

1

Antiphospholipid antibody syndrome

1

Bilateral pleural effusion

1

b) Methods

administration of 40 ml of omnipaque

All patients were subjected to:

a rate of 3

ml/sec.

IV.

1. Thorough clinical examination with history taking, general and

chest examination.

2. Routine laboratory tests mostly complete blood
picture, other tests were considered according to
case e.g. culture and sensitivity.

3. MSCT of the chest: Toshiba Aquilion MSCT 64
channels was used. All cases were given pump IV

Figure 9: Table 1 :

2

shows summary of the MSCT findings in our 20
cases, while table 3 gives summary of the SVC different
MSCT appearances and table 4 shows collateral
circulation in MSCT.

Figure 10: Table 2

1	
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IV.	
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Case presentation	MSCT finding
ation	
5 Behcet cases	-Partial SVC obstruction
6 central infiltrating	-Eminent mediastinal collaterals
Bronchogenic carci-	Partially Infiltrated SVC with few collaterals noted in 5 cases
noma	and
Case of mediasti-	eminent c chest wall collaterals in one case (the case was a
nal syndrome of un-	follow up case of Bronchogenic carcinoma after radiother-
known etiology un	apy).
	Diffuse affection of the mediastinum with soft tissue density
	lesion causing effacement of SVC and marked attenuation of
	the right pulmonary artery ; a picture suggesting fibrosing
	mediastinitis*
	Eminent c chest wall collaterals seen.
2 cases with left up-	DVT seen extending into left brachiocephalic vein. Normal
per limb DVT	contralateral brachiocephalic vein and SVC.
2 cases with lym-	Huge anterior mediastinal mass .Displaced and compressed
phoma	SVC.
One case of chest as-	No collaterals
essment for metas-	Lung, pleura and mediastinal metastases .SVC distortion and
tases	partial infiltration .
	No collaterals
One case of fever of	Partial SVC obstruction with a heterogenous throm-
unknown origin af-	bus**with air
ter chemotherapy	

[Note: density, enlarged SVC, no collaterals One case of Antiphospholipid antibody syndrome Partial SVC obstruction at lowest portion , no collaterals One case of bilateral pleural effusion Partial SVC thrombus in relation to CV-Line catheter, no collaterals *case was proved fibrosing mediastinitis. ** Case was proved to have MRSA infection.]

Figure 11: Table 2 :

2le3

M MSCT SVC findings	No. of cases
Partial obstruction	9 cases
Infiltration	7 cases
Normal	4 cases

Figure 12: Table 2 : le Table 3 :

4

No of cases	HRCT finding
5 Behçet cases	Eminent collaterals
1 case of fibrosing mediastinitis	Eminent collaterals
1 case of infected thrombus	No collaterals
2 cases with partial thrombus	No collaterals
5 cases of malignant mediastinal infiltration by Bronchogenic carcinoma	Few collaterals mostly chest
1 case of follow up Bronchogenic carcinoma with SVC infiltration	Eminent chest wall collaterals
1 case of 2ry malignant mediastinal infiltration.	No collaterals
V.	

Figure 13: Table 4 :

Figure 14: Table 5 :

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