A 68 years Old Man with Compressive Chest Pain and Breathlessness

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A 65 year old Bangladeshi male presented with compressive chest pain and respiratory distress for 2 days. He had hoarseness of voice and breathlessness for 1 year. He was hypertensive, diabetic and dyslipidaemic, and had a past history of ischaemic.

Physical examination revealed a pulse rate of 92/min, blood pressure (BP) of 130/100mmHg, unremarkable praeordial and lung auscultation. ECG showed right bundle branch block. Troponin I and NTPro-BNP were normal. Echocardiogram revealed concentric LV hypertrophy with normal LV systolic dysfunction, and grade I diastolic dysfunction. Chest X ray showed widened mediastinum with prominent aortic knob (Fig.-1).

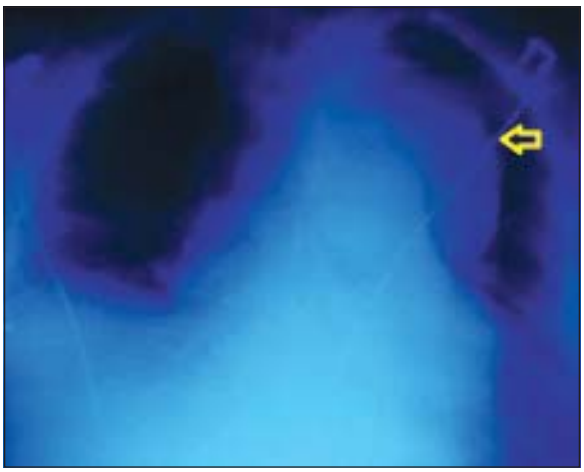

Fig.-1: *Chest X ray AP view showing widened mediastinum with prominent aortic knob.*

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CT aortography showed a large saccular dilatation of the arch of aorta (Fig.-2) distal to the left subclavian artery measuring 8.3cm transversely with mural thrombus. Another focal fusiform dilatation of proximal thoracic aorta measuring about 6.9cm at maximum was seen, with eccentric mural thrombus leaving a patent lumen of ~2.7cm. Fusiform diataion was also seen in the abdominal aorta (Fig.-3). No dissection seen. Extensive workups for thrombophilia were negative.

He was given beta blockers and ACE inhibitor for BP control, aspirin, and statin, in line with ACCF/AHA recommendations[1]. Given the mural thrombus, he was treated with enoxaparin, and discharged on warfarin. He was offered thoracic endovascular repair of aneurysm (TEVAR) or open surgical repair of thoracic aortic aneurysm (TAA).

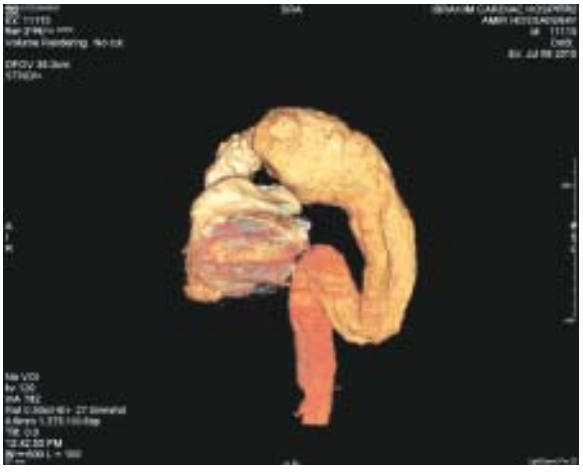
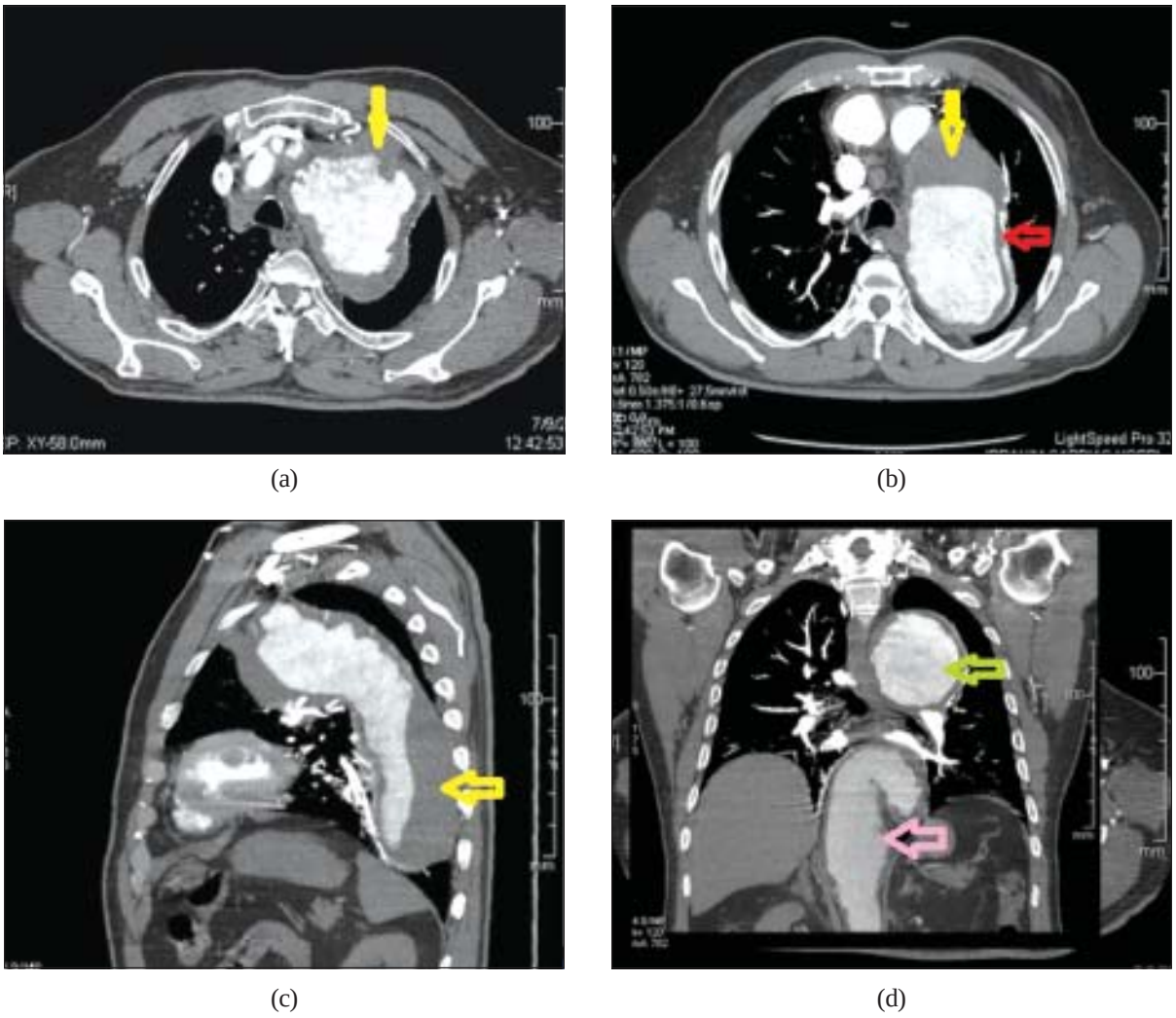


Fig.-2: *CT aortography image showing large saccular dilatation of the arch of aorta and focal fusiform dilatations of descending thoracic and abdominal aortae.*



(a) (b)

(c) (d)

Fig.-3: *CT scan of chest (a) : Axial plane: Ascending aortic aneurysm showing filling defect (yellow arrow) denoting thrombus. (b): Axial plane: Descending TAA (red arrow) with thrombus (yellow arrow). (c): sagittal plane: descending TAA showing thrombus (yellow arrow). (d): coronal plane showing ascending aortic aneurysm (green arrow) and descending TAA and abdominal aortic aneurysms (pink arrow).*

Discussion:

An aortic aneurysm is diagnosed when the ascending aorta is larger than 5 cm and the descending aorta is larger than 4 cm[2]. Aortic root or ascending aortic aneurysms (~60%) are the most common TAA, followed by descending aorta (35-40 %) and aortic arch (<10%) [2,3]. Thoracoabdominal aneurysms constitute approximately 3% of all aortic aneurysms and are usually diffuse and atherosclerotic in nature[3]. Atherosclerosis is the overall most common cause of aneurysm, accounting for 70%[2]. In contrast to the ascending aorta, the majority of descending TAAs are atherosclerotic[2,3]. Most of them present in the sixth and seventh decades of life, with a male predominance, and involvement of abdominal aorta in one-third of patients[2].

The most likely aetiology in this case was atherosclerosis, as for most descending TAA, compounded by risk factors of smoking, hypertension, and older age[4]. Descending TAAs are typically fusiform, often begin distal to the origin of the left subclavian artery[3] and coexist with abdominal or arch aneurysms[3].

Most TAA and abdominal aortic aneurysms are clinically silent, with the aneurysm discovered incidentally on

chest radiography, echocardiography, CT or MRI[4]. Alternatively, rupture may constitute the first manifestation[3]. 5%-10% of patients experience symptoms, such as chest or back pain. Aortic arch or descending TAAs may also produce hoarseness of voice ("dysphagia lusoria") from compression of the recurrent laryngeal nerve[3].

Atherosclerotic aneurysms are commonly associated with mural thrombosis, posing the additional risk of systemic embolization, causing occlusion of distal vessels[3-6]. Other serious complications of TAA are dissection and rupture.

Most TAAs are evident on chest radiographs[3,4] but chest X rays cannot exclude their diagnosis. Trans thoracic echocardiography may not thoroughly characterise arch and descending TAA. Transoesophageal echocardiography can usually image most of the aorta[4]. However, the best imaging modality for TAA is Computed tomography (CT) or magnetic Resonance Imaging (MRI). It has several advantages, including rapid scan times, wider availability and the ability to image the three-dimensional structure of the aorta along its entire course[1-3]. Furthermore CT angiography and contrast-enhanced MRI are highly accurate in the evaluation and follow up of patients undergoing endovascular TAA therapy, and are also preferred over aortography in most cases[3].

The optimal management of TAA mural thrombi has not been clearly established and is influenced by the localisation of the thrombus and patient co-morbidities. Therapeutic strategies include anticoagulation, thrombolysis, interventional modalities such as thromboaspiration, or balloon-catheter thrombectomy, and open surgical procedures such as thrombectomy, thromboendarterectomy, and aortic prosthetic replacement[7]. Strict control of hypertension (target goal <130/80mmHg in diabetics), optimization of lipid profile with statins (target LDL <70mg/dL), smoking cessation and glycaemic control should be instituted. Beta-blockers and ACE inhibitors or Angiotensin receptor blockers are recommended anti-hypertensive choices (class IIa)[1].

Intervention is recommended at larger diameters for aneurysms of the descending aorta[1]. Symptomatic aortic aneurysms at any level should be resected regardless of size. Modalities include open surgical repair (OSR) or Thoracic Endovascular aneurysm repair (TEVAR). TEVAR is a far less invasive alternative to OSR of descending TAAs, with lower morbidity and mortality rates, provided the aortic anatomy has adequate landing zones to accommodate the endograft[1,8]. Long term surveillance of the aorta with imaging is imperative, with re-evaulation at 6 months after discovery of the aneurysm to document its stability. For degenerative TAAs bi-annual imaging is recommended for aneurysms between 4.5 to 5.4cm, and annual if 3.5-4.4cm[4].

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LETTER TO THE EDITOR

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To

Editor-in-Chief

Journal of Bangladesh College of Physicians and Surgeons.

Sir,

I would like to thank you for publishing the article " Haemophagocytic Lymphohistiocytosis in Adult- A Case Report and Literature Review" in your journal . I have gone through it and appreciate the authors for reporting on such a rare and important case. I would like to share some of my observations and comments regarding this case.

Secondary haemophagocytic lymphohistiocytosis (HLH) occurs after strong immunologic activation which can occur with systemic infection, immunodeficiency or underlying malignancy. Epstein-Barr virus infection is most common one linked with HLH. Patient with dengue fever can sometimes develops unusual manifestation in the form of expanded dengue syndrome. HLH is one of the important expanded dengue syndromes.

That 65 years old male presented with drowsiness for 1day with a recent history of high grade intermittent fever in the month of June which is a peak month for dengue infection. In this case report clinical features suggestive of dengue i.e muscle and joints/bones pain, retro orbital pain was not mentioned clearly. Whether patient was febrile throughout the illness or became afebrile within a short period. Initial investigation was suggestive of dengue haemorrhagic fever. Author mention ICT for dengue was negative but I am not fully satisfied with only this statement. Whether NS1 antigen for dengue was done or not, was not mentioned. I would like to know if authors took every step to exclude possibilities of dengue in this case. As dengue fever is a burning public health problem in our country features mimicking dengue should be thoroughly investigate to confirm or refute the diagnosis.

Overall I think the case report and literature review is very much updated, informative. I would like to thank the authors for their hard work.

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Reply

Thank you very much for going through the article and making your observation.

Our patient presented with drowsiness for 1 day with recent history of high grade intermittent fever and

generalized erythroderma for 6 days. He had no history of headache, body ache, retro orbital pain, myalgia or joint pain which typically suggestive of Dengue fever. Initially dengue fever was one of our differential diagnosis and we did ICT dengue on the day of his admission (6th day of his illness) and it was negative. We know dengue NS1 Ag remain positive for initial 5 days of illness. This test may become negative from day 4-5 of illness. Hence we did not do dengue NS1 Ag as he came to us on 6th day of his illness. His dengue NS1 Ag was done on 2nd day of his illness before admission in our hospital and it was negative. We did repeated ICT dengue but both IgG and IgM were negative.

He was afebrile for 3 days after starting the chemotherapy. Again he became febrile and passed away after 2 days.

I fully agree with you that Dengue is one of the cause of secondary HLH. Clinicians should be aware of the fact that the occurrence of haemophagocytosis could be due to dengue virus infection in areas where the disease prevalence is more like our country.

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