Case Report

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A rare case of azygos vein arch aneurysm: A case report

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Abstract

Background: The aneurysm of the azygos vein (AVA) is a dilatation of the azygos vein (AV) over 1,5cm; it is very rare, mostly fusiform and affecting people between 19 and 70 years old. Aetiology can be traumatic, inflammatory/infectious, cardiogenic, neoplastic or idiopathic. The weak points for the development of an AVA are essentially two: the confluence of the AV in the superior vena cava (SVC) and the convergence among its embryologic constituents. AVA is often asymptomatic, therefore it can be an incidentaloma. Symptoms, usually due to compression of neighbouring structures (such as trachea, bronchi, SVC, inferior vena cava (IVC), oesophagus or nerves), can simulate a mediastinal mass. The main complication of AVA is thrombosis, which can extend to pulmonary vessels. Other complications include rupture, fistulae and dissection. Most of the authors consider the occurrence of complications the only indications to treat it.

Case report: We present the case of an 87-year-old woman with chronic atrial fibrillation, which came to our emergency department for wheezing and dyspnoea. Though the chest radiograph only found a pleural effusion, the subsequent CT scan showed a voluminous mediastinal mass compressing the oesophagus, which proved to be an AVA during the dynamic phase of the study.

Conclusion: The AVA represents a possible hypothesis in the differential diagnosis of mediastinal masses, that in most cases can be probed radiologically.

Keywords: Azygos vein; aneurysm; CT angiography; mediastinal mass.

Background

The aneurysm of the azygos vein (AVA) is a very rare pathology, with almost 50 cases in literature, mostly with a fusiform shape and identified in people between 19 and 70 years old. Since azygos vein (AV), normally, has a diameter of about 1cm, we define “aneurysm” a dilatation over 1,5cm [1]. There are two potential weak points for the development of AVA: the confluence of the AV in the superior vena cava (SVC) and the convergence among the vessels which constitute the embryologic origin of the AV (right supra-cardinal vein, right anterior and posterior cardinal veins). AVA is often asymptomatic, therefore it can be identified for the first time during exams or procedures performed for other reasons [2]. Symptoms are usually due to compression of neighbouring structures and can occur in case of enlargement of the aneurysm, which can simulate a mediastinal mass [3]. There is no consensus among authors on management of AVA, but most of them consider the occurrence of complications, such as thromboembolism, rupture, fistulae and dissection, the only indications to treat it [4].

Case Report

An 87-year-old Caucasian retired woman came to our emergency department because of sudden and progressively worsening wheezing, dyspnoea and dysphagia. The patient had a history of chronic atrial fibrillation in treatment. She had no fever; an initial assessment showed a heart rate of 90 bpm, a respiratory frequency of 20 breaths per minute, an oxygen saturation of 96% and a blood pressure of 138/88 mmHg. The auscultation of the right basal lung did not reveal any tactile vocal fremitus. A chest radiograph was performed to exclude pneumonia, but only a left pleural effusion was found. Since tumours are one of the principal causes of unilateral pleural effusion, the patient underwent a CT scan without contrast medium and cytologic pleural fluid examination.

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CT scan confirmed the presence of a left pleural effusion, without any sign of pneumonia. Nevertheless, two findings drew our attention: a thyroid goitre, which was believed to be implied on patient's dyspnoea, and a voluminous mediastinal mass behind the trachea which appeared to be in continuity with the AV. It showed axial diameters of about 30x28mm (**Figure 1**) and caused oesophageal compression. First, we thought to an enlarged lymph node, [5] however its densitometric values (typical of the fluids) and the apparent continuity with the AV seemed to suggest the presence of an AVA (**Figure 2**). After the thoracentesis, cytologic examination of the pleural fluid gave negative result, so the patient was treated with antibiotics. Subsequently a control contrast enhanced CT scan confirmed the diagnosis of saccular AVA (neck width = 22mm), showing a homogeneous contrast enhancement during the dynamic phase of the study, in addition to exclude the presence of pulmonary nodules that could have previously hidden by the passive atelectasis due to the presence of the fluid.

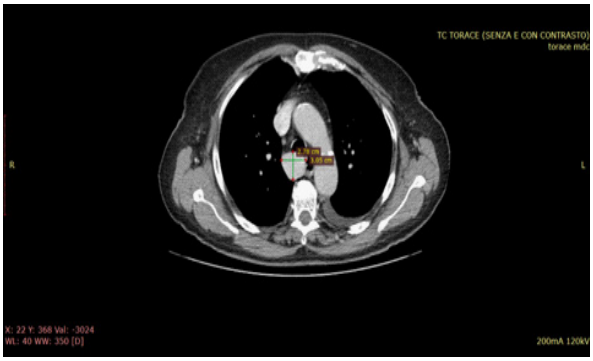

Figure 1: *Enhanced contrast CT (venous phase) demonstrates a saccular AVA behind the trachea with axial diameters of about 30x28mm, which causes oesophageal compression.*

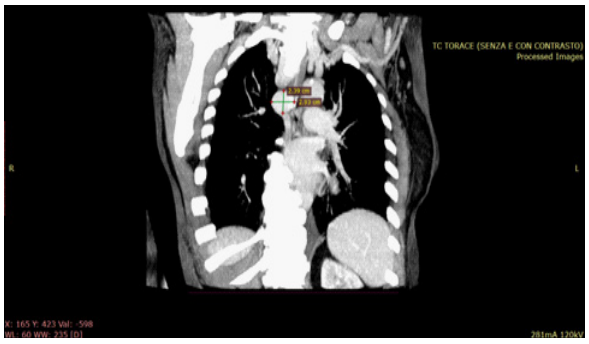

Figure 2: *Coronal reconstruction better demonstrates the continuity of the aneurysm with the AV.*

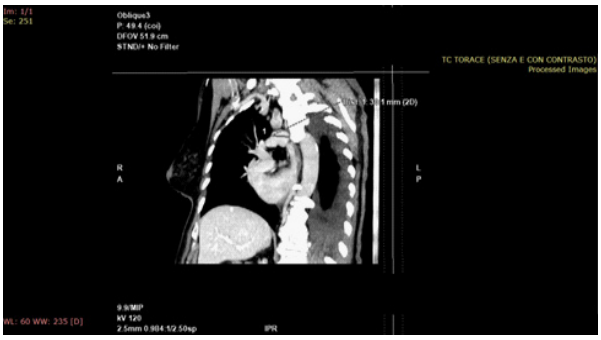

Figure 3: ***Oblique sagittal MIP confirms the diagnosis.***

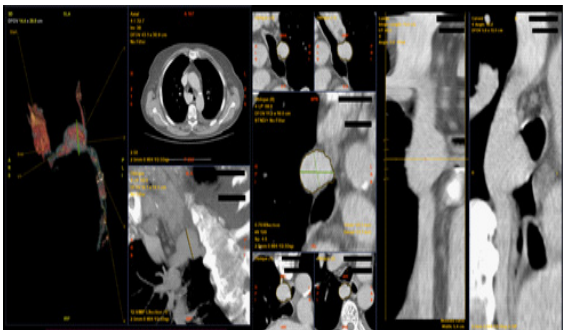

Figure 4: *Volume rendering (VR) and post-processing reconstructions can provide more specific information about the characteristics and the measurements of the lesion.*

Discussion

There is no evidence in literature to establish the distribution of AVA in age or sex groups; a small prevalence has been observed in women between 19 and 70 years old.

It has been reported the association between AVA and anatomic variants, such as double inferior vena cava (IVC), retroaortic left renal vein and an azygos vein which drains into the IVC.

AVA has different aetiologies:

- Traumatic (according to many authors, it is the most frequent), secondary to blunt traumas or catheterization
- Volume overload, caused by heart failure, portal hypertension, pregnancy, pulmonary sequestration with drainage in the AV, but also by congenital malformations such as obstruction and/or agenesis of the AV and/or venae cavae
- Connective tissue alterations (acquired or congenital); the association with the Ehlers-Danlos syndrome is reported in literature, especially with type 4
- Vascular tumours, such as haemangiomas, lymphangiomas and leiomyosarcomas of the AV
- Inflammations/Infections
- Phlebosclerosis (vascular fibrosis with parietal thickening) and/or endothelial phlebo-hypertrophy (which includes anomalies of the protein synthesis and of the connective tissue)
- Idiopathic, due to anomalies in the embryological development of the AV
- AVA can be asymptomatic, however this is true for small aneurysm, as the probability of being symptomatic increases with the diameter of the dilatation. Symptoms are often due to compression of neighbouring structures:
- Trachea and principal bronchus (cough and dyspnoea)
- SVC and/or IVC
- Oesophagus (dysphagia)

- Vagus nerve

Other symptoms include chest pain, pre-syncopal episodes, palpitations and hiccups.

Diagnosis of AVA can be very difficult. Radiological examinations combine the utilization of radiography, trans-oesophageal doppler ultrasonography, computed tomography (CT) and magnetic resonance imaging (MRI); doppler ultrasound can also differentiate between AVA and azygos lobe, highlighting the presence of blood flow.

Other investigations by means of venography or video-assisted thoracoscopy (VATS) can be required in doubtful cases; bioptical procedures are always not recommended, because of the risk of haemorrhage.

Chest X-ray can be frequently negative; a smooth-margin opacity situated at the right tracheobronchial corner, which changes its volume during the respiration or Valsalva maneuver, can be identified in the postero-anterior (PA) projection; the same entity can be suggested by the presence of a posterior prominence located in the mediastinum on the lateral view [6]. CT and MRI are useful to define the vascular nature of the lesion and its relationship with neighbouring structures. Differential diagnosis includes vascular/and or neurogical tumours, bronchogenic cysts, lymph nodes and other kinds of mediastinal masses.

Contrast enhanced CT shows us a progressively contrasted lesion (maximum enhancement during the equilibrium phase). Multi-planar reconstructions (MPR) help to identify the mass and to establish its communication with SVC. A cardio-CT study can better define the morphology and provide us lots of information about the right heart function and the possibility of thromboembolic events; this can influence the prognosis and the treatment [7].

MRI findings are different according to the sequence:

- T1 weighted imaging: iso-intense mass compared to soft-tissues

- T2 weighted imaging: heterogeneous mass with prevalent high intensity (fluid) and focal absence of signal, which is typical of vascular lesions

- Contrast enhanced T1 weighted imaging: progressive enhancement (maximum enhancement after 60 seconds); an angiographic MRI study will show us a homogeneously contrasted mass [8, 9].

The main complication of AVA is thrombosis, secondary to alterations in blood flow, which can produce acute or chronic pulmonary hypertension (the thrombus can extend to pulmonary vessels) and pulmonary embolism. Other complications include rupture, formation of fistulae and dissection.

AVA does not require treatment when asymptomatic; it is usually sufficient to control the etiological factors (eg. anticoagulants in patients with disorders of coagulation) and to perform follow-up examinations [10].

Most of authors agree to treat AVA when symptomatic, complicated or in case of a doubtful situation. The excision can be performed during video-assisted thoracoscopic surgery (VATS) or thoracotomy. It is reported in liter-ature that some authors recommend the excision only to idiopathic aneurysm, others in case of saccular aneurysm, even if asymptomatic, because of its lack of stability [11].

Interventional radiology includes many approaches to the treatment of AVA, such as stenting, amplatzer, embolization with coils and thrombolysis.

Conclusions

The hypothesis of AVA must be considered during the differential diagnosis of mediastinal masses, even if its occurrence is very rare. Contrast enhanced CT and MRI imaging can provide in most cases enough information for the diagnosis, but sometimes a video-thoracoscopic or open-surgical approach can help in doubtful cases. The insurgence of complications is currently considered by most of the authors the only indication to treat an AVA.

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