Abstract
Background: The incidence of large and medium pulmonary vasculitis is rare and insidious. The prognosis for such cases is poor with no established treatments. This case reports the differential diagnosis and surgical management of an immunoglobulin G4 (IgG-4) related granulomatous occlusive large pulmonary vessel.

Case Summary: A 67-year-old woman was hospitalized with complaints of “On and Off” exertional shortness of breath associated with dry cough and fever. Multiple diagnostic modalities were done to establish the differential diagnosis. Her multi-slice CT pulmonary angiography showed a hypodense area at the origin of the right pulmonary artery (RPA) causing complete occlusion with late thin filling of distal part at hilum suggestive of a chronic thrombosis or an underlying mass lesion. The echocardiography revealed mild mitral / tricuspid regurgitation and severe Pulmonary Arterial Hypertension (PAH). A mass excision from the RPA and its reconstruction using conegra conduit was done. Histopathology of the tissue inferred the diagnosis as IgG-4 related granulomatous vasculitis.

Discussion: The presence of IgG-4 related granulomatous vasculitis in the pulmonary large vessel is rare, underestimated and usually not investigated. Use of multiple modalities helped in the differential diagnosis. The prognosis of this patient appears improved following surgical management.

Keywords: Pulmonary vasculitis; Large vessel; Immunoglobulin-4 related; Case report; Differential diagnosis, Reconstruction using conegra conduit, Takayasu arteritis

Learning point: Multiple modalities helped in the differential diagnosis of large vessel pulmonary vasculitis. In selected cases, mass excision and reconstruction of the pulmonary arteries offers better prognosis for pulmonary vasculitis of large vessel.

Introduction
Vasculitis is the inflammation of the blood-vessel wall. It is responsible for the pathophysiology of a diverse group of individual diseases[1]. The infiltration of the immune effector cells results in constitutional complaints and end-organ damage[2]. The prevalence of the large and medium vessel vasculitis is relatively rare, and, the incidence of isolated pulmonary vasculitis is rarer[3].

The present case reports an unusual finding of a large vessel Pulmonary Artery (PA) vasculitis, wherein the mass arising from the right PA, extending to the middle PA and pulmonary valve along with enlarged para-aortic lymph node were excised and reconstructed. The Histopathological Examination (HPE) inferred the diagnosis as IgG-4 related granulomatous vasculitis, a very rare entity with very few cases surgically managed and reported so far.

Case Presentation
A 67-year-old female, with controlled hypertension was hospitalised with complaints of “On and Off” exertional shortness of breath associated with dry cough and fever. Three months earlier, she had complaints of retrosternal chest pain radiating to the right side of the back, which was treated as esophagitis and resolved with medications. During hospitalization, her condition deteriorated rapidly with the development of orthopnoea, severe right sided chest pain with right ventricular systolic pressure of 70 mmHg.

Diagnosis
Before hospital admission, she underwent several diagnostic investigations. High-Resolution Computed Tomography (HRCT) of the thorax was done with a suspicion of peripheral right lower lobe pneumonia. Her 2D-Echo, reported left ventricular ejection fraction (LVEF) ~ 60% with mild mitral regurgitation (MR)/mild pulmonary arterial hypertension. Her multi-slice CT pulmonary angiography showed a hypodense area at the origin of the Right Pulmonary Artery (RPA) causing complete occlusion with late thin filling of distal part at hilum suggestive of a chronic thrombosis or an underlying mass lesion. The contrast CT showed a filling defect in the RPA, probable thrombus, suspecting an infarct. Whole body Positron Emission Tomography (PET) was suggestive of a hypermetabolic lesion (2.9x1.6x2.9 cm) at the origin of the RPA, a lesion (1.5x1.5x1.4 cm) between the non-coronary sinus
projecting into the right atrium and a right supraclavicular lesion (1.2x0.6x0.9 cm), involving mediastinal lymph nodes with mild pericardial effusion (Figure 1). Ultrasonography (USG) of the neck was normal except tiny hypoechoic areas noted in both the lobes of the thyroid gland. Small thrombus in the segmental branch of the right lower lobar PA was also noted. Multiple small right pulmonary infarctions / consolidations and minimal right pleural effusion were also noted. Left Pulmonary Artery (LPA) was normal. There was no liver, adrenal and bone lesions.

On hospitalisation, coronary angiogram depicted, normal coronary arteries, mild mitral / tricuspid regurgitation, and severe PAH. Right ventricle angiography showed totally occluded RPA, normal LPA and coarsely trabeculated ventricle with good contraction (Figure 2). Digital Subtraction Angiography (DSA) revealed dilated long Main Pulmonary Artery (MPA) with normal opacification, totally occluded RPA from the origin, normal LPA and pulmonary valve with no abnormal branching pattern. Pre-operative chest X ray, USG of abdomen/pelvis, and venous doppler of both the limbs were normal.

Surgical procedure

She was counselled and consented for surgical treatment. Her surgical procedure was a mass excision from the RPA and its reconstruction using contegra conduit (Figure 3-5). The operating procedure included, median sternotomy, verticle pericardiotomy and entire thymus gland removal. Themass arising from RPA, extending into the MPA and reaching upto pulmonary valve, along with the enlarged para- aortic lymph node was excised and HPE was done. Reconstruction from MPA to RPA was done using No. 16 contegra conduit and roof of MPA was reconstructed with pericardial patch.

Hospital stay and management

Her intraoperative course was uneventful and she was mobilized on second day post operatively. Minimal inotropes were administered. She was discharged in a haemodynamically stable condition. Her discharge prescription included warfarin, antibiotics and analgesics. The HPE of the RPA showed transmural vascular inflammation composed of lymphocytes, plasma cells with many lymphoid follicles, suggestive of large vessel vasculitides (Takayasu arteritis/Giant cell arteritis) Figure 6). The histopathological findings inferred the diagnosis as Immunoglobulin G-4 (IgG-4) related granulomatous vasculitis (Figure 7,8a and b).
**Figure 3:** Contegra to right pulmonary artery

**Figure 4:** Contegra to right pulmonary artery anastomosis

**Figure 5:** Surgical outcome
Figure 6: Necrotising granulomatous vasculitis

Figure 7: Immunoglobulin G related immune-histocompatibility

Figure 8 a,b: Immunoglobulin G-4 related immuno-histocompatibility

Discussion

We describe here a case of pulmonary vasculitis who presented with exertional shortness of breath, right sided chest pain, with no history of haemoptysis, anorexia or weight loss. With an intention to provide rational treatment and improved prognosis, we investigated the clinical characteristics, pulmonary lesions, using several diagnostic modalities. The symptoms of PA vasculitis often suggest more prevalent diagnoses such as infection, malignancy or connective tissue disease[4]. The stenosis, occlusion, or embolism of PA's can cause PAH, perfusion defects, or even pulmonary infarction[5]. Differential diagnosis of large vessel PA vasculitis is tricky, because of the associated
heterogenous clinical symptoms, lack of a single reliable diagnostic test, and its low prevalence[6]. Multiple imaging modalities have to be employed to detect PA abnormalities including DSA, CTA, PET CT etc [7]. In the present case, multiple modalities helped to establish the differential diagnosis of PA vasculitis. Although recommended in only few cases, this patient was managed surgically for better prognosis[8]. Histological verification of the inflamed vessel wall is paradigm to the diagnosis of vasculitis[6]. This case of large vessel PA vasculitis inferred the diagnosis as IgG-4 related granulomatous vasculitis, whose occurrence is very rare. Its presence in the pulmonary circulation is underestimated and usually not investigated. In the present case, the differential diagnosis was aided with multiple diagnostic modalities and clinical characteristics. Mass excision and reconstruction of the pulmonary arteries was successfully done. The prognosis of this patient appears better following timely surgical management.

Consent: The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

Acknowledgements

The authors acknowledge the guidance and support of Parloop Bhatt in preparation of the manuscript and acknowledge the lab technicians and nurses for documentation of the case information.

References