

**Case Presentation**

# An Interesting Autopsy Case Report of Acute Respiratory Failure

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**Introduction**

Vasculitis is a group of rare but potentially serious disorders characterized by inflammation of blood vessels. This condition can affect blood vessels of all sizes, ranging from small capillaries to larger arteries. The inflammation can disrupt blood flow, leading to a variety of symptoms and potential complications [1]. Vasculitis can target various organs and body systems, making it a challenging condition to diagnose and manage. It is a rare autoimmune disorder with female preponderance causing the immune system to attack blood vessels. Its incidence is 6 in every 1,00,000 population and more commonly seen in elderly population. Amongst vasculitis, ANCA associated vasculitis is extremely rare, with a worldwide incidence of 1.2 to 2.0 cases per 100,000 individuals [2]. The estimated 1 year survival rate for ANCA associated vasculitis is 77% - 100%. The 5 and 10 year survival rates vary from 46% to 85% and 60% to 80% respectively. The mortality rates are falling because of better and advanced treatment options but remain elevated in severe cases. ANCA associated small vessel vasculitis includes microscopic polyangiitis, Wegeners granulomatosis, Churg-strauss syndrome and drug induced vasculitis. The exact cause of vasculitis is often unknown, but it is believed to involve an abnormal immune response. The immune system mistakenly attacks healthy blood vessels, leading to inflammation. Some cases of vasculitis are linked to autoimmune disorders, where the body's immune system attacks its tissues [3]. Other possible triggers include infections, certain medications, and exposure to toxins. The affected individual presents with fatigue, muscle pain, fever, cough, hemoptysis, abdominal pain, blood in urine or weakness and numbness in hands or feet. In severe cases there is presence of blood and protein in urine, making it appear brownish and foamy, high blood pressure and respiratory problem. The normal ANCA level may differ from lab to lab although the normal levels of ELISA are 0.00 units/ml to 0.22 units/ml.

**Clinical profile**

A 29 year old serving soldier was admitted with complains of fever, pain abdomen, hemoptysis, bodyache and dark coloured urine since 03 days. On admission patient

**More Information**

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was tachypneic and hypotensive. Blood investigations revealed decrease hemoglobin and platelet count. pANCA came out to be positive. CT scan was suggestive of bilateral nodular densities and x ray showed bilateral fluffy opacities. Despite management with oxygenation, iv fluids and iv antibiotics, there was no improvement in his symptoms and his condition further deteriorated. Bronchoscopy revealed diffuse bronchoalveolar hemorrhage. Biochemical evaluation revealed deranged kidney and liver function test. Patient on ventilation developed bradycardia, hypotension and desaturation. On examination blood pressure was not recordable, carotid pulse was not recordable, CPR was started and patient was declared dead at 1215hrs on 30 Oct 2020 Figures 1,2.

Patient was taken for autopsy and on external/ gross examination of organs, there was visible hepatomegaly,



Figure 1: X Ray shows bilateral fluffy opacities.

kidney showed multiple petechial hemorrhages and both lungs were boggy Figures 3-5.

Histopathology showed evidence of diffuse alveolar hemorrhages in both lungs. Evidence of vasculitis was noted in small vessels of kidney and lungs, with neutrophilic inflammatory infiltrate around these vessels. Hence it was concluded that p-ANCA associated vasculitis lead to diffuse alveolar hemorrhage, causing acute respiratory distress and eventually respiratory failure Figures 6-8.



Figure 2: CT scan shows multiple nodular densities.



Figure 3: Hepatomegaly, liver weighing 2300gm.



Figure 4: Petechial hemorrhages over both kidneys.

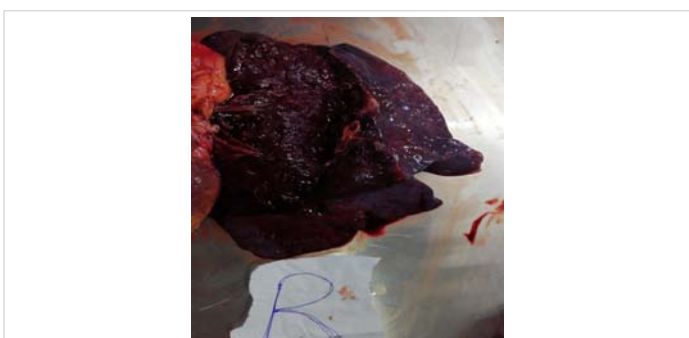


Figure 5: Bilateral lungs are boggy and heavy.

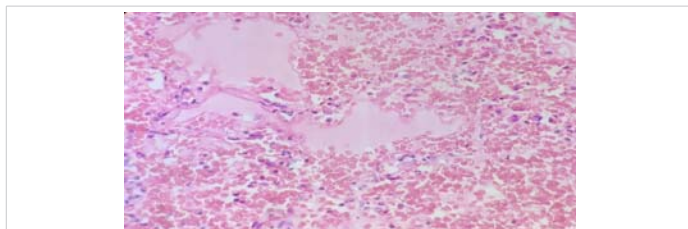


Figure 6: Section shows interstitial and intra-alveolar hemorrhage with denuded alveolar lining epithelium.

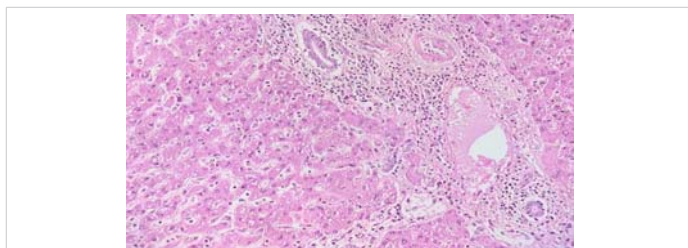


Figure 7: Section shows dilated vessels with peri-portal inflammation and dilatation of sinusoids.

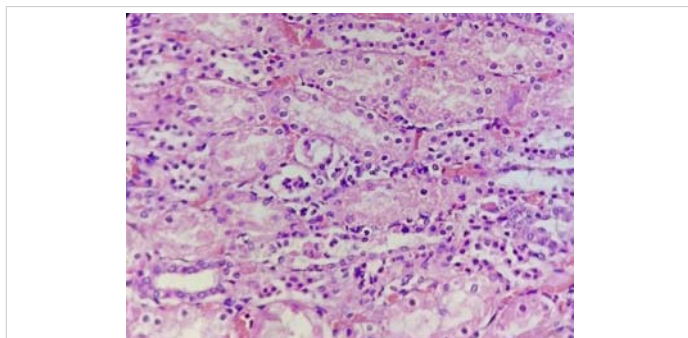


Figure 8: Section shows congested blood vessels and tubular nephritis with small interstitial capillary injury.

## Results

Diagnosing vasculitis can be complex, as its symptoms often overlap with other medical conditions. The prognosis is extremely poor if not diagnosed in time. Hence better clinical history and adequate tests like test for ANCA antibodies, LFT and KFT is extremely important [3]. Treatment for vasculitis aims to control inflammation, alleviate symptoms, and prevent further damage to organs. The approach often involves administration of Corticosteroids, Immunosuppressants, Biologic therapies Plasmapheresis and Supportive care. Hence, vasculitis is a complex and potentially severe condition characterized by inflammation of blood vessels. Early diagnosis and appropriate treatment are essential for managing symptoms, preventing complications, and improving the overall quality of life for those affected [4].

## Discussion

The exact cause of vasculitis is often unknown, but it is believed to involve an abnormal immune response. The immune system mistakenly attacks healthy blood vessels, leading to inflammation. Some cases of vasculitis are linked to autoimmune disorders, where the body's immune system



attacks its tissues [5]. Other possible triggers include infections, certain medications, and exposure to toxins. The affected individual presents with fatigue, muscle pain, fever, cough, hemoptysis, abdominal pain, blood in urine or weakness and numbness in hands or feet. In severe cases there is presence of blood and protein in urine, making it appear brownish and foamy, high blood pressure and respiratory problem. The normal ANCA level may differ from lab to lab although the normal levels of ELISA are 0.00 units/ml to 0.22 units/ml [6].

## Conclusion

Vasculitis is a rare autoimmune disorder and hence its diagnosis is extremely challenging. There is no clinical criteria to diagnose a case of vasculitis. The only gold standard investigation in a case of vasculitis is histopathology. Hence early diagnosis and medical intervention is extremely important to improve long term survival.

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