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An Interesting Autopsy case Report of Acute Respiratory Failure

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Abstract

Introduction: Acute respiratory distress comes with a lot of etiological factors. The least common among it is, diffuse alveolar hemorrhage which constitutes more than 50% of the mortality rate. One of the least common cause of diffuse alveolar hemorrhage is Small vessel vasculitis. Vasculitis is a group of rare but potentially serious disorders characterized by inflammation of blood vessels.

Clinical profile: Vasculitis presents with multiple etiological factors, 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. 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

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. Treatment for vasculitis aims to control inflammation, alleviate symptoms, and prevent further damage to organs. The approach often involves administration of corticosteroids, immunosuppressants, biological therapies, plasmapheresis and supportive care

Discussion: 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 was tachypneic and hypotensive. Blood investigations revealed decrease hemoglobin and platelet count. pANCA came out to be positive. 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.

Conclusion: Vasculitis is a rare autoimmune disorder and hence its diagnosis is extremely challenging. There is no god standard or clinical criteria to diagnose a case of vasculitis. Hence early diagnosis and medical intervention is extremely important to improve long term survival.

Keywords - Acute respiratory failure, vasculitis, diffuse alveolar hemorrhage, ANCA associated vasculitis

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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 1,00000 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 100000 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 to 0.22units/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 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.

Date	28/10/2020	29/10/2020	30/10/2020
Hb (g/dl)	12.7	12.4	11.6
TLC (c/mm)	7800	7700	9000
DLC (%) N/L/M/E/B	85/10/5/-/-	89/05/06/-/-	89/06/05/-/-
Platelet count (cu/mm)	60000	62000	14000
PT/INR	-	-	17.6/1.28

Date	28/10/20	29/10/20	30/10/20
Urea/Creat (mg/dl)	35/1.07	44/1.09	62/2.28
S.Bilirubin (mg/dl)	1.5	1.8	3.8
SGOT (IU/L)	28	44	92
SGPT (IU/L)	32	40	75
Alkaline phosphatase	70	85	81

Date	28/10/20	29/10/20	30/10/20
PCV	-	-	35.8
MCV (fl)	87.5	87.5	87.5
MCH (pg)	28.6	28.6	28.6
MCHC (pg)	32.8	32.8	32.8
RBC (million/ml)	4.4	4.4	4.4
PBS for hemolysis	No hemolysis seen	No hemolysis seen	-
PBS for Malaria		Negative	

Date	30/10/20
pANCA	4.19, Positive
Gram Stain	Polymorphonuclear cells seen
Urine RE/ME	Proteinuria

On further investigations, Serum electrolytes, serum protein and C-reactive protein levels were within normal limits. Blood culture, BAL culture, Dengue serology, HbsAg, HIV, COVID-19 Antigen & antibodies, Gene expert for MTB, Antibodies against GBM/ds-DNA/IgM Leptospira/ANA/Factor Hep2 showed negative results.

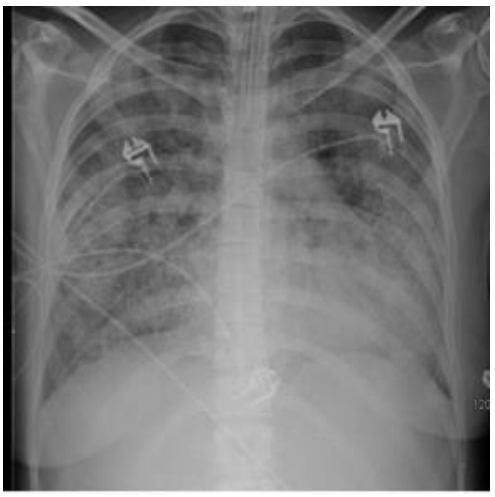

Figure 1 : X Ray shows bilateral fluffy opacities

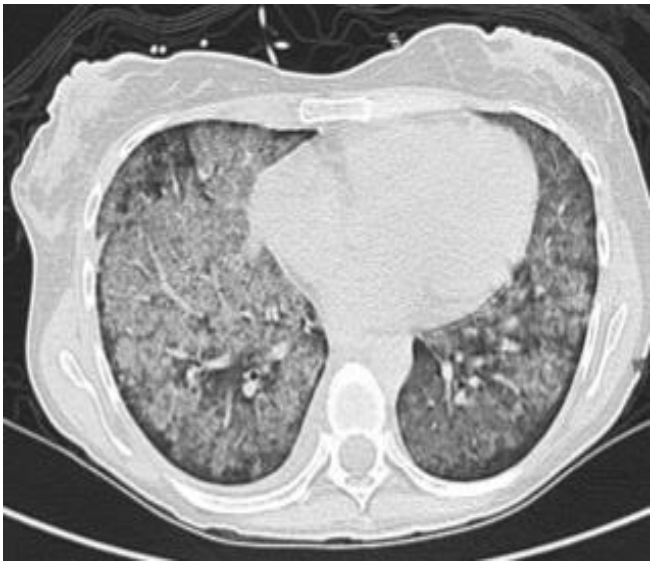

Figure 2 CT scan shows multiple nodular densities.

Patient was taken for autopsy and on external/ gross examination of organs, there was visible hepatomegaly, kidney showed multiple petechial hemorrhages and both lungs were boggy.

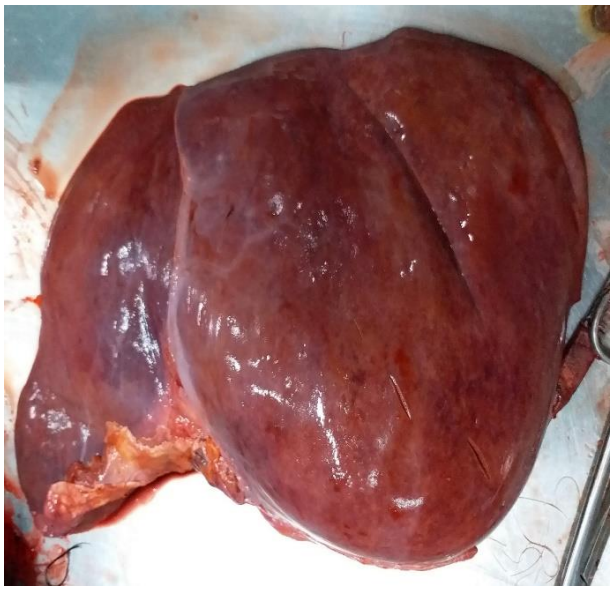

Figure 3 Hepatomegaly , liver weighing 2300gm

Figure 4: Petechial hemorrhages over both kidneys

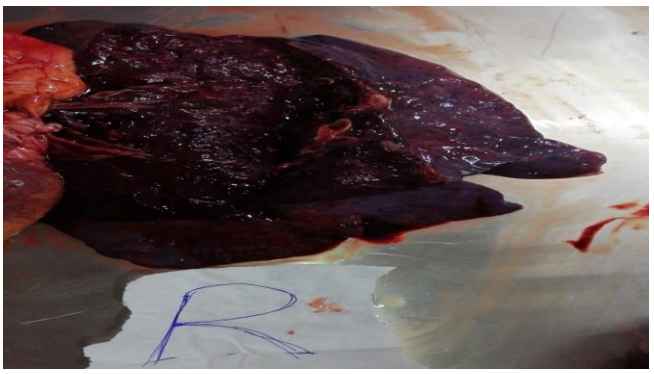

Figure 5 Bilateral lungs are boggy and heavy

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.

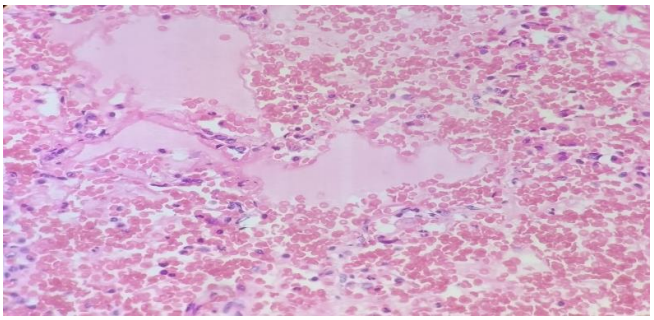

Figure 6 Section shows interstitial and intra-alveolar hemorrhage with denuded alveolar lining epithelium

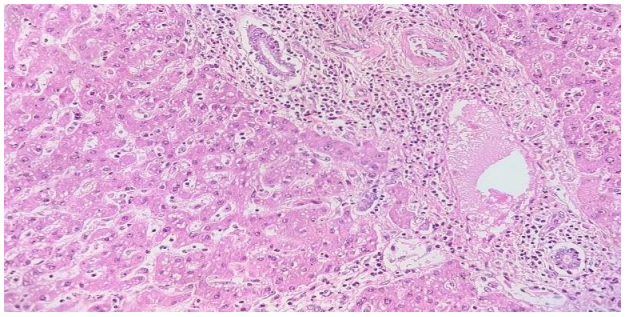

Figure 7 Section shows dilated vessels with peri-portal inflammation and dilatation of sinusoids.

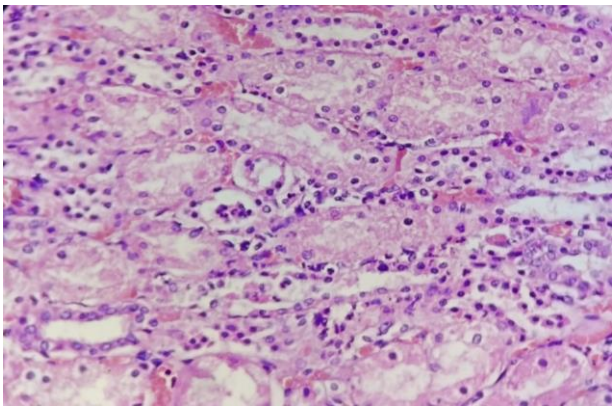

Figure 8 Section shows congested blood vessels and tubular nephritis with small interstitial capillary injury

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.T he 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 to 0.22units/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. Early diagnosis and medical intervention is extremely important to improve long term survival. Hence, any case of vasculitis should be approached in following manner as mentioned in below table:

Rule out secondary causes	Access extent of vasculitis	Confirm diagnosis of vasculitis	Identify the specific type of vasculitis	Differentiate between c- ANCA & p-ANCA
Blood cultures Echocardiogram Hepatitis screen (B&C) HIV test Anti-glomerular basement membrane antibody Antiphospholipid antibodies Antinuclear antibody	Urine dipstick and microscopy Chest radiography Nerve conduction studies Electromyography CK	Biopsy Angiogram	ANCA Cryoglobulin Complement level Eosinophil counts/IgE levels Specific findings on biopsy	Immunofluoresence ELISA

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