Case report

Diffuse alveolar hemorrhage revealing systemic lupus erythematosus

Abstract :

Alveolarhemorrhageis an uncommon complication of systemic lupus erythematosus, rarelybeing the initial symptom. This condition is a medical emergency, as it can quicklyprogress to life-threateningrespiratoryfailure. The characteristicsymptomsincludecoughing up blood, anemia, and lung inflammation on imaging. Earlytreatmentwithcorticosteroids and immunosuppressants can significantlyimproveoutcomes. Wepresent a case of a youngwomanwithalveolarhemorrhagethatled to the diagnosis of severesystemic lupus erythematosus. Sherespondedfavorably to immediatetreatment.

Introduction :

Systemic lupus erythematosusis one of the major systemicautoimmunediseases. This condition ischaracterized by significantclinicalpolymorphism, markedessentially by rheumatological, dermatological, hematological, nephrological, cardiac, and pulmonarysigns, which are exceptionallycomplicated by intra-alveolarhemorrhage. We report a case of intra-alveolarhemorrhagerevealingsystemic lupus erythematosus.

Case presentation :

Observation:

This is a 17-year-old female patient with no significantpastmedicalhistory. One monthprior to admission, shepresentedwithinflammatoryarthralgiaaffecting large, medium, and small joints, associatedwithlowerextremityedema and dyspnea on exertion. Subsequently, hersymptomsworsenedwith the acute onset of respiratorydistress and massive hemoptysis. Shewasadmitted to the internalmedicinedepartment.

On admission, the patient wasafebrile, tachycardic at 120 beats per minute, with an oxygen saturation of 89% on room air, pittingedema of the lowerextremities, and non-scarringalopecia. Shewasinitiallyplaced on oxygentherapy. Laboratory investigations

revealed a hypochromicmicrocyticanemia at 2.9 g/L secondary to massive hemoptysis, thrombocytopenia at 75,000, and lymphopenia at 640, with a positive Coombs test and a C-reactiveprotein of 60 mg/L. Renal and liverfunction tests were normal.

A chest CT scan showed multiple poorlydefined, confluent, diffuse hyperdense lesionsaffecting all lung lobes with a ground-glass appearance, suggestive of alveolarhemorrhage. Bronchoalveolar lavage confirmedalveolarhemorrhagewithuniformlyhemorrhagicfluid.(Fig1)

The patient receivedthreeunits of packedredbloodcells. Afterstabilization, an extensive workupwasperformed. Infectiousworkup, including HIV, tuberculosis, Aspergillus, and Pneumocystis jiroveci, wasnegative. Cytobacteriologicalexamination of sputumwasnegative.

Immunologicalworkuprevealed positive antinuclearantibodies at a titer of 320 with a speckled pattern, positive anti-DNA antibodies, and lowcomplement C3 and C4 levels, rheumatoid factor, and anti-neutrophilcytoplasmicantibodieswerenegative. Given the edema, a renalworkupwasperformed, revealingnephrotic syndrome withhypoalbuminemia at 25 g/L, 24-hour proteinuria of 2.40 g, and hematuria on urine sediment. A renalbiopsyshowed class 4-5 lupus nephritis.(Fig 2)

Afterruling out infection, vasculitis, and otherautoimmunediseases, a diagnosis of systemic lupus erythematosuswas made based on the 2019 ACR/EULAR criteria, supported by positive immunologicalfindings, inflammatoryarthralgia, hemolyticanemia, thrombocytopenia, class 4-5 lupus nephritis, and alveolarhemorrhage.

Treatmentincluded oral corticosteroids at 1 mg/kg/day, alongwith four monthly pulses of cyclophosphamide,followed by maintenance therapywith azathioprine 150 mg daily. After four months of treatment(Fig 3), the patient showedsignificantclinical, biological, and radiologicalimprovementwithout complications or relapse.

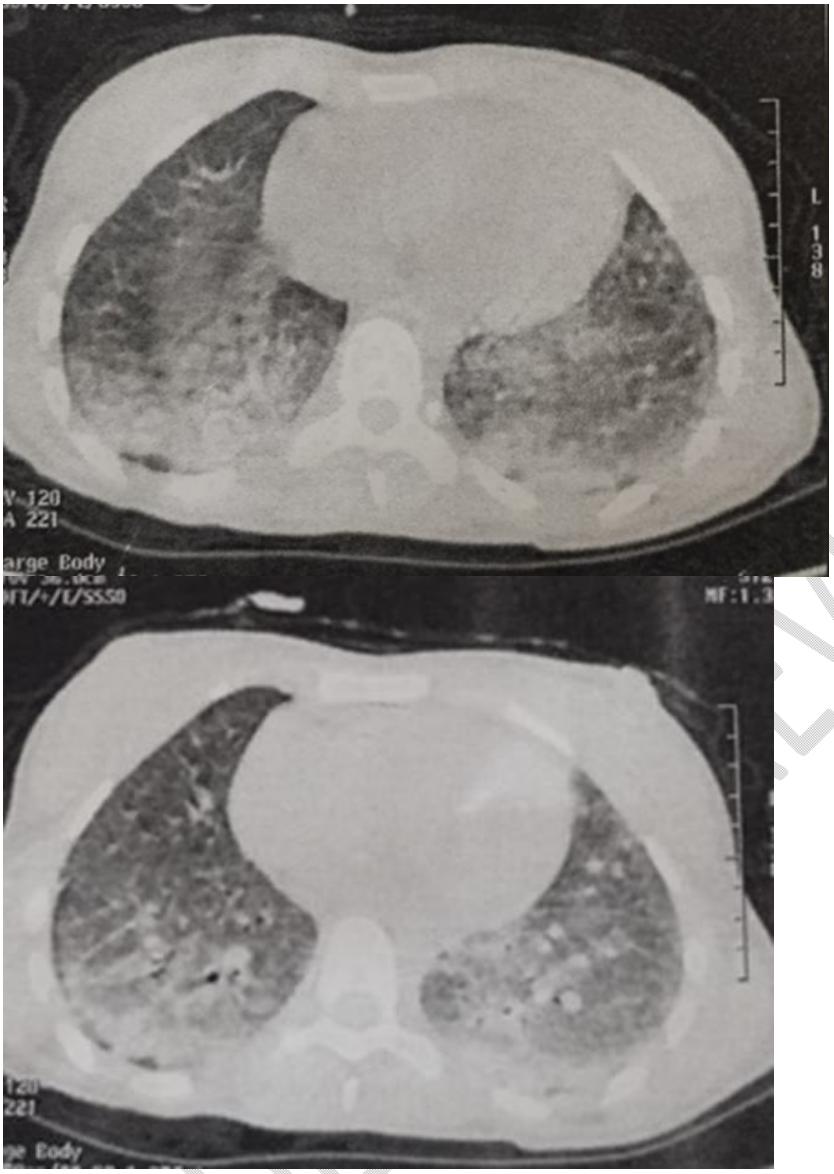

Fig 1 : Chest CT scan showed multiple poorlydefined, confluent, and diffuse hyperdense lesionswith a ground-glass appearance**.**

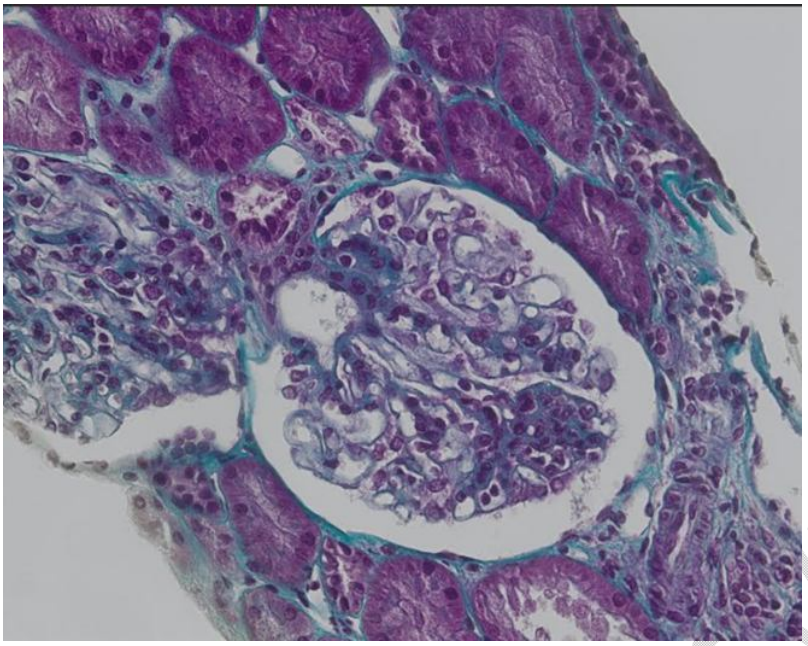

Fig 2 : renalbiopsy showed class 4-5 lupus nephritis

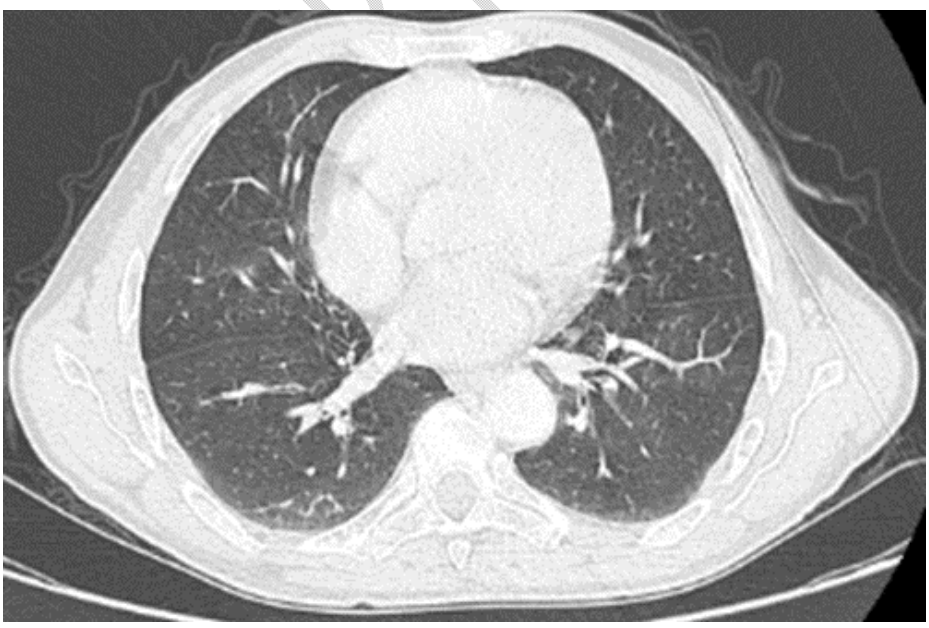

Fig 3 : Chest CT scan afterfour months of treatment.

Discussion:

Pleuropulmonary manifestations are common in Systemic Lupus Erythematosus (SLE) and can beclassifiedinto five main groups[1]based on anatomy: pleural involvement, infiltrative lungdisease, airwayinvolvement (upper and lowerairways), vascularinvolvement (pulmonary hypertension, alveolarhemorrhage), and muscle and diaphragmaticinvolvement. [2]

Alveolarhemorrhageis a rare but serious manifestation[3], defined by the leakage of bloodinto the pulmonaryalveolusfollowing damage to the alveolar-capillarybarrierresultingfromvariousmechanisms[4]. Itspathogenesisinvolvesgenetic, environmental, and immunologicalfactors, with a complement-dependentcytotoxicreaction of autoantibodies, a reaction to circulating immune complexes, and B-lymphocyte hyperactivity [5]. The occurrence of alveolarhemorrhage in lupus isrelated to vascularlesionssuch as capillaritis or thoseinduced by immunoglobulindeposits, leading to fibrinoidnecrosis of the capillary [6]. The clinicalpresentation of alveolarhemorrhageistypicallycharacterized by a triad of hemoptysis, dyspnea, and pulmonaryinfiltrates on imaging [7]. This complication ismostoftenseen in women in whom the diagnosis of SLE has already been made[8], althoughitis rare for alveolarhemorrhage to be the first manifestation of lupus, necessitatingearly management[9]. The treatment goals are rapid control of bleeding and prevention of progression to irreversible damage, such as chronicrespiratoryfailure[10]. Treatmentisbased on high-dose corticosteroids, plasmapheresis, and cyclophosphamide[11]. Immunosuppressive therapyshouldbeinitiatedearly, combiningcorticosteroids and oftenintravenous cyclophosphamide[12]. Plasma exchange isindicated in poorlycontrolled SLE or in case of failure of high-dose corticosteroidtherapy. In refractory cases, the use of biologic agents such as rituximab and belimumabisnecessary, allowing for a reduction in comorbidities and disease relapses and improving long-termprognosis[13][14]

Conclusion:

Intra-alveolarhemorrhage in systemic lupus erythematosusis a rare but serious complication, oftenassociatedwithothersystemic manifestations. It istypicallysecondary to alveolarcapillaritis, the exact cause of whichremainsunclear. This complication significantly impacts the patient'sprognosis and requires prompt and well-definedtreatment. Despite new diagnostic tools and therapies, systemic lupus erythematosusassociated diffuse alveolarhemorrhageremains a diagnostic and therapeutic challenge, with high mortality, sotrying to identifyfactorsassociatedwithmortality, and the measures and strategies to diminish or avoidit, are essential tasks. Controlledstudies, cohortseries, and case reports are important to evaluate the potentialbenefit of currenttherapies, whichincludebiologicaldrugs.

CONSENT :

As per international standards or university standards, patient(s) written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL :

Ethicalapprovalwasexempted by the EthicalCommittee at Ibn Roch universityhospital for reportingthis case.

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Details of the AI usage are givenbelow:

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