

Plasma cell leukemia occurring six months after covid-19 is associated with a paraneoplastic limbic encephalitis

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Abstract

A 73-year-old obese individual was diagnosed with an acute COVID-19 infection. The patient's condition improved, but six months later a new deterioration ensued. Following the appearance of pancytopenia, plasmacytosis involved the peripheral blood (35%), as well as the bone marrow (31%), disclosing an osteoclastic pattern. An encephalitis which prevailed at that stage was of the limbic type. It displayed anti-GABA B-antibodies. Therefore, the condition being neither an infection, nor a metabolic disorder, nor a vascular lesion, is most consistent with an autoimmune reaction. The clinical features were consistent with a plasma cell myeloma, associated with a paraneoplastic syndrome. The diagnosis favored the POEMS syndrome, even though a few criteria were missing. It was sustained by a fluid collection surrounding an enlarged spleen, as well as filling up both pleural gutters. This POEMS case was related with COVID-19, both via the original acute episode, as well as with a late possible subacute COVID-19 pulmonary disorder, assessed by extensive radiologic ground glass changes. In this instance, the patient developed hypoxemic respiratory insufficiency and he died of respiratory failure, in addition to due to the plasma cell leukemia. Thus, we have reported the simultaneous occurrence of a POEMS syndrome, induced by a plasma cell leukemia, and confounded by a pulmonary COVID-19, perhaps with long COVID features.

Keywords: COVID-19, Plasma cell leukemia, Paraneoplastic syndrome, Encephalitis, POEMS syndrome, Hypoxemic respiratory failure

Case Presentation

This elderly man was overweight, hypertensive, and was treated for dyslipidemia. Culminating six months prior to the present occurrence, an acute COVID-19 episode terminated with normal respiratory function tests.

The patient's status deterioration included several components: an acute neurological episode, with marked confusion and cognitive alterations developed together with pancytopenia, highlighting a plasmacytosis of 35% in the peripheral blood, and of 31% in a bone marrow biopsy, the absolute peripheral blood plasma cell count being $> 2.0 \times 10^9 /L$, accompanied by osteosclerosis.

A brain MRI imaging demonstrated an intense DWI-FLAIR signal without signs of hemorrhage or enrichment in the limbic system, more pronounced on the left lobe. These findings were interpreted as a paraneoplastic syndrome, exhibited as autoimmune encephalitis, most probably initiated by a plasma cell neoplasm, notably by

a lambda monotypic plasma cell leukemia. Splenomegaly and lymphadenopathy were evident. Creatinine was 0.80 mg/L [1-3].

An attempt was made to classify the above syndrome. The syndrome's components encompassed a paraneoplastic syndrome [4], in the form of encephalitis; monoclonal gammopathy and osteoclastic bone lesions. Moreover, a flow analysis disclosed 98.2% cytoplasmic lambda light chains, which sustained the diagnosis of POEMS. Of note, fluid collections were exhibited in both pleural gutters and surrounded an enlarged spleen.

A further critical aspect of this case, consisted of a hypoxemic respiratory failure, highlighted by an excess of ground glass opacities in both lungs. This, in turn, may evoke an association with respiratory COVID-19. Of note, the patient had suffered six months prior to the present syndrome of this infectious disease. The theory of a long COVID has been raised and should perhaps be retained [5].

Regarding the cause of death, although respiratory failure is a major candidate, the plasma cell neoplasm might have contributed significantly. Untreated POEMS patients are reported to die of the associated plasma cell neoplasm [6-8].

Minor additional features include hematophagocytosis, and focal and scarce expression of HHV-8, both of unknown relevance to the course of the present disease.

Conclusion

Our patient is proposed to have developed POEMS syndrome, six months after having restored to health from an acute episode of COVID-19 infection. The unusual aspects, underlining this report, include the nature of the plasma cell infiltrates. Thirty-five% plasma cells in the peripheral blood, disclosing clonality, are diagnostic of plasma cell leukemia. Splenomegaly, lymphadenopathy, and lytic bone lesions lend partial confirmation to the primary diagnosis. Together with the limbic encephalitis, the plasma cell myeloma variant, confirms the diagnosis of POEMS syndrome.

The other outstanding features consist of an ongoing association with COVID-19, first as the acute episode of the infectious disease, and later, in the context of a ruthless respiratory involvement with multiple ground glass opacities (GGO), compatible with severe pulmonary COVID-19 infection, highlighted by the hypoxemic respiratory failure, a major contribution to the patient's death. The radiological evidence of GGO raises a differential diagnosis which includes several lung disorders. To establish a more precise diagnosis one should determine if the disease is acute, sub-acute or chronic, and whether the malady involves the air spaces, the interstitial regions, or causes alveolar collapse. A clinical-radiological correlation becomes necessary. Moreover, the COVID-19 pneumonia being a major radiological imitator, one should account for the situation of the pandemic in the patient's neighborhood.

To our knowledge, we may have presently reported an association between POEMS syndrome and a severe aspect of pulmonary COVID-19, possibly a long COVID-19 complication.

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Conflicts of Interest

The authors declare 'No conflicts of interest exist'.

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