An autopsy case report of intravascular large B-cell lymphoma with initial neurologic presentation

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Intravascular large B-cell lymphoma is defined as a rare type of extranodal large B-cell lymphoma characterized by the selective growth of lymphoma cells within the lumina of blood vessels, in particular capillaries, and with the exception of larger arteries and veins. Here, I present an autopsy case of intravascular large B-cell lymphoma in a 60 years old Chinese male who initially presented with neurologic deficit.

Case presentation

The deceased was a 60-year-old Chinese male with a history of diabetes, hyperlipidemia, right intertuberous osteoarthritis and tuberculosis lung abscesses. He was initially admitted to the medical unit for low back pain and bilateral lower limb weakness. He developed acute retention of urine, increasing lower limb weakness, lower limb areflexia and confusion later. Initial blood test showed anemia, hypotension, reduced thyroxine (T4) and triiodothyronine (T3) and markedly elevated lactate dehydrogenase (LDH) (1341 U/L), ferritin (10722 pmol/L), C-reactive protein (CRP) (110 mg/L) and erythrocyte sedimentation rate (ESR)(102 mm/hr). Initial neurological workups including CT EEG were unremarkable. MRI spine was performed and showed nerve root enhancement at cauda equina and Guillain-Barre syndrome was suspected. MRI brain showed small old infarcts in right parietal lobe and right basal ganglia only. Lumbar puncture was performed and showed elevated protein level, white cell count (lymphocytic predominant) and normal glucose level. Nerve conduction test and electromyography showed evidence of sensory and motor axonal polyneuropathy with active denervation. Cytology study on cerebrospinal fluid was negative. Extensive microbiological, virological and metabolic investigations were unremarkable. Autoimmune antibodies and screening for multiple myeloma were all negative. The working diagnosis was Guillain-Barre syndrome with Bickerstaff encephalitis. He was treated with IVIG with partial improvement of mental status but persistent neurologic deficits. Subsequently the patient developed high swinging fever with sputum culture yielding methicillin-resistant Staphylococcus aureus and patchy haziness on chest X-ray. Antibiotics were given. The patient eventually passed away two months after admission. The case was referred to the Coroner’s court due to uncertain cause of death and the family of the deceased agreed for autopsy.

Microscopic findings

The most significant finding was the presence of large malignant lymphoma cells with irregular nuclei and occasional cytoplasmic extrusion in the small blood vessels of cauda equina, spinal cord, brain, pituitary gland, thyroid gland, adrenal glands, lungs, epicardium, spleen, kidneys and urinary bladder.

Diagnosis and follow-up

The cause of death was stated as pneumonia with intravascular large B-cell lymphoma as the underlying cause and diabetes mellitus as a contributing factor.

Discussion

Intravascular large B-cell lymphoma is a rare type of extranodal large B-cell lymphoma occurring in adults. The median age is 67 years but it can occur over a wide age range from 13 to 90 years. No significant male or female predominance is observed.

Pathologically, the lymphoma cells are found in small blood vessels, typically capillaries and post-capillary venules. Medium-sized blood vessels and sinusoids can also be affected, but not large arteries, large veins or lymphatics. The lymphoma cells are typically large with vesicular nuclei, distinct nucleoli and brisk mitosis. Occasionally irregular nuclei and smaller cell size may be encountered. Haemophagocytosis may be seen. Immunohistochemically, the lymphoma cells are positive for CD20, BCL2 and often show a non-GCB phenotype by Hans algorithm, with MUM1 positivity, CD10 and BCL6 negativity. The prognosis of intravascular B-cell lymphoma is in general poor but has improved significantly with the addition of rituximab to anthracycline-based chemotherapy also appears to improve the outcome of patients.