Cerebral venous thrombosis due to high-altitude polycythemia

Case report

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A case of cerebral venous thrombosis due to polycythemia secondary to adaptation to a high altitude is reported. A 27-year-old previously healthy man developed severe neurological symptoms after climbing 8511 m. Computerized tomography and cerebral angiography suggested hemorrhagic infarction or intratumoral hemorrhage, and a craniotomy was performed. Pathological examination confirmed the diagnosis of hemorrhagic infarction secondary to cortical venous thrombosis. The etiology and incidence of cerebral venous thrombosis secondary to polycythemia are discussed.

KEY WORDS • cerebral venous thrombosis • high-altitude sickness • polycythemia • infarction

Cerebral venous thrombosis due to polycythemia has rarely been reported. It is well known that cardiopulmonary collapse, cerebral edema, and thrombophlebitis of the extremities are common in climbers at high altitudes. However, no pathologically confirmed case of cerebral venous thrombosis has previously been reported in this group of individuals. We report a case of cerebral venous thrombosis as a complication of polycythemia secondary to adaptation to a high altitude.

Case Report

This 27-year-old previously healthy man climbed from Kathmandu, Nepal (at 1300 m above sea level), to a base camp 4050 m above his starting point. Twenty days later, after climbing another 3161 m, he developed headaches and subsequently became confused. High-altitude sickness and dehydration were diagnosed. A hemoglobin titer of 21.5 gm/dl was confirmed, and corticosteroids were administered intravenously. He did not improve and was admitted to the Neurosurgical Service at the Tokyo Metropolitan Komagome Hospital.

Examination. At the time of admission, the patient was disoriented. Funduscoppy revealed papilledema and retinal hemorrhages. Pertinent laboratory data showed a high hemoglobin titer (16.5 gm/dl), mild hepatic dysfunction, slightly prolonged prothrombin time, and activated partial thromboplastin time. Computerized tomography demonstrated a high-density area with surrounding low-density in the left posterior temporal region. Irregular enhancement occurred in this region on administration of contrast material (Fig. 1). Cerebral angiography showed slow circulation over the left hemisphere; the vein of Labbé could not be seen on that side, although small corkscrew-like collateral veins were seen emptying into the superior sagittal sinus (Fig. 2). Hemorrhagic infarction or intratumoral hemorrhage was suspected.

Operation. The patient continued to worsen neurologically while receiving steroids, and he subsequently underwent a left temporoparietal craniotomy. The vein of Labbé and some of the Sylvian veins were found to be thrombosed and the underlying cortex was necrotic and hemorrhagic. The thrombosed vein was excised and the devitalized brain and subcortical hematoma were evacuated.

Postoperative Course. The patient improved dramatically and had no residual neurological difficulty. Pathological evaluation revealed dilated cortical veins
High-altitude polycythemic cerebral venous thrombosis

FIG. 1. Computerized tomography scans obtained on admission. Upper: Plain scans showing a high-density area and surrounding irregular low-density area in the left posterior temporal lobe. Lower: Scans obtained after administration of contrast material showing irregular enhancement surrounding the high-density area.

filled with partially organized thrombi (Fig. 3). The parenchyma was necrotic and bloody, confirming the diagnosis of hemorrhagic infarction secondary to cortical venous thrombosis.

Discussion

Various causes of cerebral venous thrombosis have been reported, including infection,~15,18,19 oral contraceptives,~18,19 postpartum complications,~15,16,18 cancer,~7,19 and idiopathy.~18 Although cerebral arterial thrombosis has been associated with polycythemia vera~2,20 and a high hemoglobin titer,~1,17,21 only one case of cerebral venous thrombosis due to polycythemia has been previously documented.~19

Altitude illness may manifest itself clinically with pulmonary and cerebral edema, and retinal hemorrhage.~5,6,8,9,11 Venous thrombosis of the extremities and pulmonary embolism have also been associated with mountain climbing.~10 Yet, there have been no previous

FIG. 2. Angiograms showing corkscrew-like collateral veins flowing toward the superior sagittal sinus. The vein of Labbé is not visible.

FIG. 3. Photomicrograph of surgical specimen showing a dilated cortical vein filled with organized thrombus and adjacent capillaries. Lymphocytes, macrophages, and hemosiderin deposits are also seen. H & E, × 15.
pathologically confirmed cases of cerebral venous thrombosis associated with high altitudes. The etiology of this entity is a low oxygen tension-induced erythropoiesis, with hematocrit levels sometimes exceeding 70%.[12,14] Subsequently, blood viscosity increases with a hematocrit over 60%, resulting in decreased cerebral blood flow and oxygen transport.[13,22] In addition, increased blood viscosity induced by polycythemia damages the vascular endothelium and activates platelets, thereby accelerating the thrombotic process. Hypoxia and low temperatures can also have the same effect.[3]

Individuals who live at high altitudes are typically polycythemic, yet they are not particularly susceptible to thrombotic events.[12,14] In our patient multiple factors, such as secondary polycythemia, cold, hypoxia, and alternating cardiovascular stress during the climb, might have resulted in the cerebral venous thrombosis and subsequent hemorrhagic infarction.

Fatal altitude sickness has been attributed to acute brain swelling[11] or cardiopulmonary failure. Autopsies performed on such individuals have documented diffuse cerebral swelling with multiple hemorrhages.[11] Our experience with this condition suggests that the neurological deficits in this patient were secondary to cerebral venous thrombosis due to the high altitude.

References

Manuscript received April 25, 1985.
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