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Case Report

A patient with acute myeloid leukemia presented with a superior sagittal sinus thrombosis as the first manifestation of Trousseau syndrome

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This paper reports a case of Trousseau syndrome with intracranial venous sinus thrombosis as the first manifestation, which is relatively rare in the clinic. A 44-year-old female patient presented with a blurred vision of the visual substance for 2 months, and the condition was aggravated with a headache for 10 days. The final diagnosis was intracranial venous sinus thrombosis and acute myeloid leukemia subtype M2. Anticoagulant + intra-arterial regimen (cytarabine + igdabistar) was given, and the patient's headache and blurred vision were gradually restored. After 2 courses of chemotherapy, acute myeloid leukemia subtype M2 was in complete remission. After 6 months of follow-up, headache and the blurred vision disappeared, leukemia did not recur, limb vascular ultrasound was screened regularly, and no new vascular embolism disease occurred.

Keywords

Trousseau syndrome; Acute myeloid leukemia subtype M2; Intracranial venous sinus thrombosis; Thrombosis; Cerebrovascular disease; Neurology

1. Introduction

Superior sagittal sinus thrombosis is a rare and life-threatening complication in leukemia, especially in acute myeloid leukemia [1]. Cerebral venous sinus thrombosis (CVST) is a particular type of cerebrovascular disease. The incidence of CVST is less than 1% of all strokes [2]. It mainly includes transverse sinus, sigmoid sinus, superior sagittal sinus, inferior sagittal sinus and cerebral venous thrombosis. Its clinical manifestations are closely related to the involved venous sinus. Recent studies have shown that the possible cause of superior sagittal sinus thrombosis is its use with corticosteroids [1]. In this study, a patient with acute myeloid leukemia presented with a superior sagittal sinus thrombosis as the first manifestation is reported in this paper.

2. Case presentation

A 44-year-old female patient was admitted to our hospital because of blurred vision for 2 months and aggravation of headaches for 10 days. Two months ago, there was no obvious inducement for the patient's visual acuity to decline. The patient's sense of light became dark. She had transient amaurosis. The symptoms gradually worsened until she could not distinguish the objects in front of her. The sense of light was like night. Ten days before she was admitted to the hospital, the patient had headaches, which was whole brain pain, accompanied by nausea and vomiting-previous physical fitness. The institutional review board of the Harrison International Peace Hospital approved this study, code 2018-1-004. A written informed consent for publication of case details and pictures was obtained from the patient prior to the study. There was no similar history in the family. No history of drug and food allergy. Physical examination showed that vital signs were stable, anemic, clear and fluent. The pupils of both sides are perfectly round and sensitive to light reflection. The light of both eyes is like the night, and their abduction is limited. Bilateral frontal lines and nasolabial sulcus did not become shallow-limb muscle strength level 5. Double Babinski syndrome was negative.

No abnormality was found in liver and kidney function and coagulation routine. The D- two polymers were 2.6 mg/L. The blood routine showed that the platelets were 87 $\times$ 109/L. The fundus examination showed papilloedema. The head computed tomographic (CT) showed that the right frontal sulcus lobes' density was increased (Fig. 1A), and hemorrhage was considered. The head magnetic resonance imaging (MRI) + susceptibility weighted imaging (SWI) showed hemorrhage in the right frontal lobe, right cerebellum, right tentorium and posterior longitudinal fissure cistern (Fig. 1B,C,D,F,G,H). The head magnetic resonance venography (MRV) showed the posterior occipital segment of the sagittal sinus, hypoplasia of the sigmoid sinus, venous embolism, increased and enlargement of venous collateral circulation (Fig. 1E).

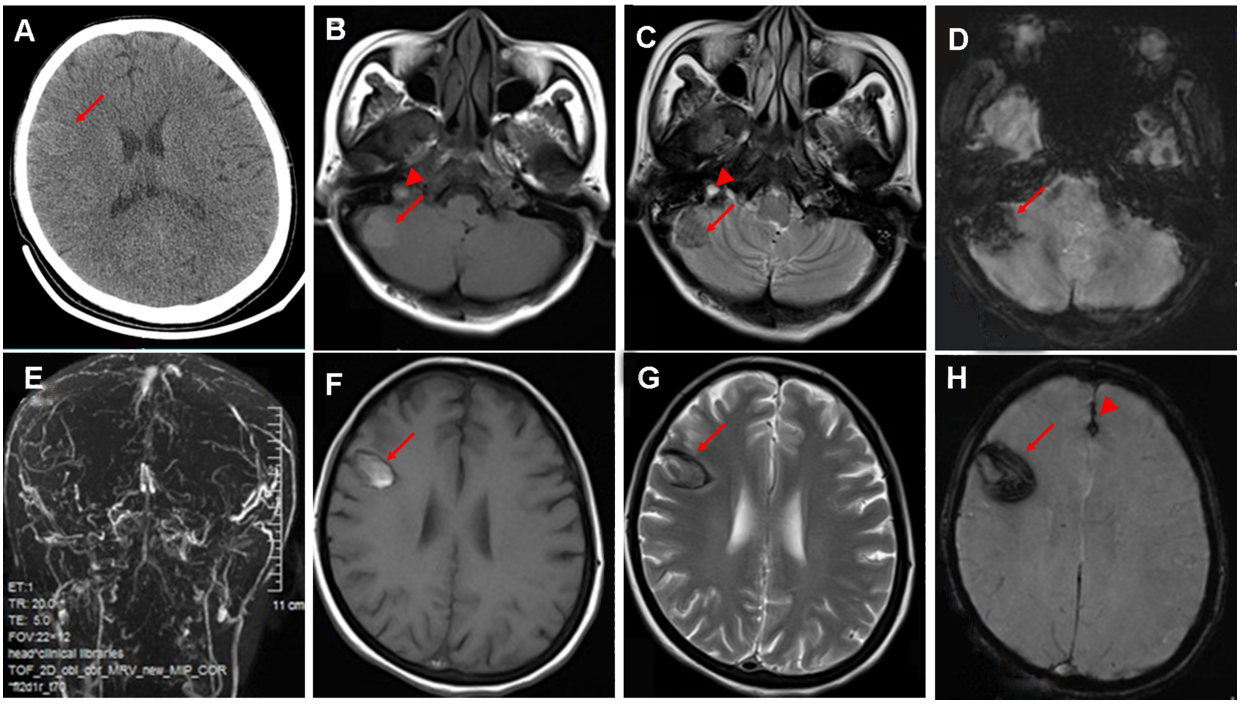

Fig. 1. A head computed tomographic (CT) showed right frontal brain hemorrhage (red arrow). B, C, D, head magnetic resonance imaging (MRI) showed short T1 and short T2 signal in the right cerebellum, right cerebellar susceptibility weighted imaging (SWI) showed low signal, hemorrhage (red arrow) was considered. B, C, head MRI showed short T1 signal, long T2 signal on the right side of the internal jugular vein region, thrombosis was considered (red arrowheads). F, G, H, head MRI showed short T1 and short T2 signal in the right frontal lobe, SWI showed low signal in the right frontal lobe, hemorrhage was considered (red arrow). H, SWI showed a low signal of anterior superior sagittal sinus, thrombosis was considered (red arrowheads). E, magnetic resonance venography (MRV) showed partial venous embolism in the sagittal posterior occipital segment, bilateral transverse and sigmoid sinus. The collateral circulation was increased and thickened.

Bone marrow imaging report showed that bone marrow hyperplasia was II-III grade; the granulocyte line accounted for 98%. The primary cells in the granulocyte line accounted for 81%. The proportion of promyelocytes was high. Erythrocyte proliferation was significantly inhibited, no nucleated cells were found, and mature red blood cells varied in size and shape. No megakaryocyte and fewer platelets were found. The histochemical analysis showed POX-72%, +21%, ++7%; NSE-56%, +33%, ++11%; NaF-62%, +30%, ++8%.

The number of white blood cells increased significantly, and the primordial cells accounted for 79%. The morphology of white blood cells was the same as that of bone marrow. The ratio of the early, middle and late granulocytes was higher. Mature red blood cells vary in size. Low platelet count. Flow cytology reported that 76.21% cells expressed CD117, CD33, CD13, CD38, cMPOdim, CD123 and CD11c, partly CD34, HLA-DR, CD64, but not CD7, cCD3, cCD79a, CD10, CD19, CD14, CD300e, CD56, CD138, which were abnormal phenotype of immature myeloid cells. The final diagnosis was intracranial venous sinus thrombosis, acute myeloid leukemia subtype M2 (AML-M2) was confirmed. The anticoagulant + intra-arterial (IA) regimen (cytarabine + igdabistar) was given, the headache and blurred vision gradually recovered, and AML-M2 was completely relieved after 2 courses of chemotherapy. After 6 months of follow-up, headache and blurred vision disappeared, no relapse and new thrombosis occurred in leukemia.

3. Discussion

In recent years, it has been found that patients with hematologic malignancies have similar or even higher thrombotic complications than those with solid tumors, mainly including multiple myeloma, acute leukemia and central nervous system lymphoma [3].

Intracranial venous and venous sinus thrombosis is rare in the clinic. It is often misdiagnosed or missed because of its various forms and clinical manifestations, with a high disability rate and mortality.

Headache and blurred visual substance were the first manifestations of this patient. The early symptoms of acute leukemia such as fever, hemorrhage and lymph node enlargement were absent, making the first diagnosis more difficult. Head CT showed multiple cerebral hemorrhages in unusual locations. There was no direct or indirect sign of venous sinus thrombosis, which was considered to be related to the low platelet level's abnormal coagulation mechanism. In contrast, the formation of intracranial venous sinus thrombosis was not considered. However, the location and amount of bleeding could not explain the limitation of ball abduction and blurred vision. The examination of the ocular fundus

showed bilateral optic papillae edema. Headache, optic papillae edema combined with binocular abduction nerve limitation indicated that patients' intracranial pressure increased, which was not consistent with the amount of bleeding shown by head CT, which should arouse the alertness of clinicians. The possibility of venous sinus thrombosis should be considered in patients with subcortical and marginal irregular multiple hemorrhages. This patient has multiple intracranial hemorrhages close to the venous sinus and cerebral cortex, which is highly suggestive of intracranial venous sinus thrombosis, and elevated D-dimer also supported this diagnosis. The diagnosis of intracranial venous sinus thrombosis was confirmed by further MRI and MRV examination.

The etiology of intracranial venous sinus thrombosis mainly included hypercoagulable state (heredity and acquired), infection, tumor, autoimmune disease, etc. The patient's blood routine showed increased leukocyte and thrombocytopenia, suggesting the possibility of hematological diseases, and finally, acute myelocytic leukemia was confirmed by bone marrow puncture. Trousseau syndrome is a variety of clinical events of thromboembolism associated with abnormal coagulation and fibrinolysis mechanism in cancer patients. It mainly includes migratory phlebitis, deep venous thrombosis, myocardial infarction, cerebral infarction [4], etc. At present, Trousseau syndrome is mostly characterized by deep vein thrombosis, which is more common in the extremities and less common in the cerebral venous system. As we know, there are only reports about Trousseau syndrome involving the internal jugular vein [5]. The patient had intracranial venous sinus thrombosis with acute myelocytic leukemia, which was consistent with the diagnosis of Trousseau syndrome, and venous sinus thrombosis was mainly associated with a hypercoagulable state caused by acute myeloid leukemia.

Unlike this article, there have been reports of rare thrombotic complications, superior sagittal sinus thrombosis, and hypercoagulable conditions secondary to the combination of L-asparaginase [6] and corticosteroids. The patients with tumor-associated with venous thrombosis should be treated mainly by anticancer + anticoagulant therapy, and anticoagulant therapy can improve the survival period of cancer patients [7]. The patient was given IA chemotherapy and low molecular weight heparin anticoagulation during hospitalization, and warfarin anticoagulation was given to the patient after discharge. The patient's headache symptoms gradually alleviated, and the leukemia was resolved entirely after 2 courses of chemotherapy. After 6 months of follow-up, headache and the blurred vision disappeared, leukemia did not recur, limb vascular ultrasound was screened regularly, and no new vascular embolism disease occurred.

In the context of leukemia, most cases of superior sagittal sinus thrombosis seem to be caused by the cytotoxic drug L-asparaginase [1, 4] but not in this paper. In this paper, the relationship between CVST and acute myeloid leukemia is related to the venous thromboembolism accompanied by acute myeloid leukemia, known as Trousseau syndrome.

Author contributions

Y.B. and J.Z.H. analyzed and interpreted the patient data, N.W. and Y.L. do the literature research, Y.B., N.W. and J.Z.H. prepared the manuscript. All authors read and approved the final manuscript.

Ethics approval and consent to participate

The institutional review board of the Harrison International Peace Hospital approved this study, code 2018-1-004. A written informed consent for publication of case details and pictures was obtained from the patient prior to the study.

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Conflict of interest

The author promises that there is no conflict of interest in the article.

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