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Essential thrombocythemia presenting with spontaneous vertebral artery dissection: Case report and review of the literature

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Abstract

Background and purpose: Arterial dissection is a common cause of young adults' stroke, usually associated with cervical strain, either due to infection or trauma or occurring spontaneously. Myeloproliferative Neoplasms (MPNs) are vascular events that occur due to hyperviscosity but are rarely linked to arterial dissection in literature.

Clinical presentation: A 33-year-old male with no significant medical or family history presented with a right occipital headache, radiating ipsilaterally, oppressive, with pain intensity of 6 in a scale of 0 to 10. During the pain peak, he reported monocular visual disturbances, described as a "white bright spot" lasting over 60 minutes. He denied trauma or physical exertion. The visual disturbances resolved within 24 hours, and the headache also improved during this time, progressively getting better and fully resolving within one week. A brain CT was normal. CT angiography revealed right vertebral artery dissection of the terminal portion of segment V2. Laboratory findings showed hemoglobin 17.5 g/L, hematocrit 51.2%, and $734x10^9$/L platelets. Karyotype was normal. Bone marrow biopsy showed megakaryocytic hyperplasia, and the myelogram revealed mild dysplasia, likely secondary to hydroxyurea. The JAK2 V617F mutation was confirmed. He was referred to Neurology and Hematology follow-up.

Discussion/Conclusion: In our case, thrombocytosis was the only identified risk factor, with hyperviscosity likely contributing to endothelial damage and vascular dysfunction, increasing susceptibility to dissection.

Introduction

Cervical artery dissection is a significant cause of ischemic stroke in individuals under 50 years, accounting for approximately 20–25% of cases in this age group [1]. Dissection involves a tear in the arterial wall that leads to intramural hematoma or aneurysmal dilatation, causing either ischemia via embolization or hemodynamic compromise. The vertebral and internal carotid arteries are most frequently involved. Known triggers include blunt trauma, cervical manipulation, or sudden neck movements. However, spontaneous dissections without identifiable precipitating factors are well documented, suggesting underlying structural or systemic predispositions.

Myeloproliferative Neoplasms (MPNs) such as Polycythemia Vera (PV) and Essential Thrombocythemia (ET) are hematologic malignancies driven in most cases by somatic JAK2 V617F mutations. Essential thrombocythemia is characterized by sustained thrombocytosis and clonal megakaryocytic proliferation [2]. While thrombotic events, particularly ischemic stroke, are well-recognized complications of ET, arterial dissection is an exceedingly rare and likely underdiagnosed manifestation. However, mounting evidence suggests that MPN-induced vascular inflammation and endothelial dysfunction may compromise arterial integrity and facilitate dissection even in the absence of traditional cardiovascular risk factors [3]. Moreover, hyperviscosity may exert mechanical stress on the arterial wall, especially in high-shear zones such as the vertebral arteries during head movements, even minor ones.

This case underscores the importance of broadening the differential diagnosis in spontaneous arterial dissection, particularly in younger patients with hematologic abnormalities. Early recognition of myeloproliferative neoplasms in such contexts is essential for reducing long-term vascular complications.

Written informed consent was obtained from the patient. The literature review was conducted through searches in PubMed and Science Direct between November 2024 and May 2025. We included peer-reviewed articles written in English and available in full-text format. This case report aims to contribute to the growing yet sparse body of evidence suggesting that myeloproliferative neoplasms may predispose to arterial dissection through complex hemodynamic and inflammatory mechanisms. It also highlights the need for vigilance among clinicians evaluating stroke-like symptoms in young individuals, prompting consideration of hematologic investigations even in the absence of classic thrombotic events.

Case Presentation

A 33-year-old male, employed as a firefighter, with merely a history of vascular surgery for varicose veins at 24 years. He had no history of chronic medication use, allergies, toxic substance exposure, or significant family medical history.

The patient experienced a right occipital headache upon awakening, which radiated ipsilaterally and was described as oppressive with intensity variations (3/10 on a 0/10 scale), but 6/10 at the beginning. Concurrently, he reported monocular visual disturbances in the right eye, described as a “white bright monochromatic scotoma,” lasting over 60 minutes. These visual symptoms were reported at the peak

headache intensity and resolved as the headache subsided. The patient took over-the-counter painkillers every 8 hours, achieving partial relief. He denied any recent cough, vomiting, physical exertion, or sudden neck movements, and he reported no previous history of similar headaches.

Due to the visual changes, he visited the Ophthalmology department, where no abnormalities were detected on eye examination.

A series of imaging and laboratory assessments were conducted. CT and CTA revealed stenosis and irregularity in the terminal portion of segment V2 of the right vertebral artery. MRI and MRA performed a few days later confirmed a right vertebral artery dissection with partial recanalization. Laboratory tests revealed a hemoglobin level of 17.5 g/dL, hematocrit of 51.2%, platelet count of 734×10^9/L and an Erythropoietin (EPO) level of 1.8 mIU/mL. These findings supported the diagnosis of a right vertebral dissection, with MRI indicating a healing phase and no evidence of ischemic lesions.

The patient started on an antiplatelet regimen of 150 mg of acetylsalicylic acid daily to mitigate thrombotic risks. To address the hyperviscosity state, two phlebotomy procedures were performed during hospitalization to reduce hematocrit levels. After interdisciplinary consultation with the Clinical Hematology team, hydroxyurea therapy was initiated at a dose of 1000 mg/day.

Due to the analytical abnormalities, further investigation was carried out, including bone marrow biopsy, karyotype analysis, genetic test and myelogram**.** The bone marrow biopsy revealed marked megakaryocytic hyperplasia with preserved morphology and no significant abnormalities in erythroid or myeloid lineages. No fibrosis was identified, and a slight interstitial plasmocytosis was noted. Cytogenetic analysis showed a normal male karyotype (46,XY[20]) with no chromosomal abnormalities. The myelogram demonstrated mild dysplasia, interpreted as likely secondary to hydroxyurea exposure, megakaryocytes in normal number with pleomorphism (hypolobulated/normolobulated forms), granular forms and hypernucleated cells and myeloid series was increased with megaloblastoid elements and hypersegmented neutrophils. The genetic test confirmed the presence of a JAK2 V617F mutation. Considering all investigations performed, the most likely etiology is essential thrombocythemia.

The patient remained under follow-up in both Neurology and Clinical Hematology consultations, remained asymptomatic with no new neurological symptoms and with hematocrit and platelet levels within normal limits, and continued the prescribed therapy.

Discussion

We report the case of a young man who experienced a transient headache and visual changes, with no vascular risk factors, history of exertion, trauma, or migraines, where the only identified trigger for arterial dissection was elevated hematocrit and thrombocytosis.

The differential diagnosis of headache with visual aura typically includes primary migraine, but in this case, the vascular etiology was evident. Arterial dissections can provoke headache, often occipital in

vertebral dissections, and may be accompanied by transient visual symptoms [4]. Cervical artery dissection has been associated with migraine-like syndromes, suggesting overlapping vascular mechanisms [5].

Neurological symptoms such as headache, blurred vision, dizziness, and tinnitus are frequently reported in ET and are attributed to microvascular disturbances caused by platelet activation and abnormal vasomotor reactivity, affecting up to 55% of patients [6]. Vasomotor manifestations, including headaches and transient visual disturbances, are among the most common microvascular symptoms and often improve with low-dose aspirine [7]. Although arterial dissection is not a classical feature of ET, rare case reports have described this complication. Raza et al. (2020) reported bilateral vertebral artery dissection in a patient with ET, managed with antiplatelet therapy and stenting, suggesting that chronic platelet activation, endothelial dysfunction, and abnormal shear stress may contribute to arterial wall injury even in the absence of conventional cardiovascular risk factors [8]. In our case, the coexistence of sustained thrombocytosis and a JAK2 mutation, together with the occurrence of a spontaneous arterial dissection, reinforces the hypothesis of a causal link between ET-related vascular pathology and arterial wall fragility.

Other myeloproliferative neoplasms like polycythemia may independently contribute to headache and aura. Headaches are prevalent in polycythemia vera and often attributed to increased viscosity and altered cerebral perfusion [9]. Regensburger et al. (2022) described retinal artery occlusions causing transient visual symptoms in early-stage polycythemia, echoing our patient's experience [10].

The underlying pathophysiology suggests that hyperviscosity from elevated hematocrit impairs blood flow, especially in smaller vessels, leading to localized ischemia and increased shear stress. This mechanical strain, compounded by pro-inflammatory cytokine release and leukocyte-endothelium interactions, creates a hostile vascular microenvironment [8,11]. Myeloproliferative disorders, particularly those with JAK2 mutations, show enhanced neutrophil activation, contributing to endothelial damage via oxidative stress and protease release, such as elastase [12].

Additionally, arterial dissection and migraine-like phenomena may share mechanistic overlaps. The release of vasoactive peptides and impaired autoregulation from endothelial dysfunction could explain the transient neurologic symptoms. While standard imaging revealed no ischemic lesions, microvascular dysfunction or small-vessel dissections, beyond current radiologic resolution, may also be contributory [4]. High-resolution MRI techniques might help identify such subtle changes in future investigations.

Emerging literature also links myeloproliferative neoplasms with dissection across various arterial beds. Yasin et al. (2024) reported spontaneous coronary artery dissections in JAK2-positive patients, while Censori et al. (2004) and D'Ambrosio et al. (2008) identified similar patterns in carotid and aortic territories, suggesting systemic vascular involvement [3,13,14]. Thrombocytosis may further destabilize vascular walls through platelet-derived growth factor and proteolytic enzyme activity [15].

This case demonstrates how symptoms commonly attributed to migraine or benign headache may, in the appropriate clinical context, signal a significant vascular pathology. It reinforces the importance of considering hematologic disorders in the differential diagnosis of unexplained dissections, especially in

young adults without cardiovascular risk factors.

Conclusion

This case adds to the limited literature linking myeloproliferative neoplasms like essential thrombocytemia to arterial dissection. It may be considered as an unrecognized risk factor for arterial dissection, especially in those patients with increased hematocrit and platelet count. This case also highlights the need for physicians to consider transient symptoms for example visual disturbances or headaches, as possible manifestations of hyperviscosity in these patients. More investigations are required to determine how myeloproliferative neoplasms are related to vascular diseases and to devise specific measures to avoid and treat such complications.

Clinical recommendations and future directions

The literature highlights the necessity for early diagnosis and intervention in polycythemia and thrombocytosis to prevent complications such as arterial dissection. Recommended interventions include:

- Regular monitoring of hematocrit and platelet levels
- Antiplatelet therapy, reducing thrombotic risk, particularly in patients with elevated platelet counts and JAK2 mutations
- Interdisciplinary approach: collaboration between hematologists and neurologists is essential for managing patients with MPNs who present with vascular symptoms, given the heightened risk of cerebrovascular and cardiovascular events.

Physicians should consider myeloproliferative neoplasms in patients with unexplained dissection or transient neurological symptoms and assess for underlying hyperviscosity syndromes. Early recognition and interdisciplinary management may prevent severe cerebrovascular events.

Key clinical message

Occipital headache with transient monocular visual symptoms may be the only neurological manifestation of vertebral artery dissection. In young patients without trauma or vascular risk factors, the presence of thrombocytosis or elevated hematocrit should prompt consideration of an underlying myeloproliferative neoplasm as a potential precipitating factor.

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