

Non-specific skin purpura

Shalihin SE, Harun Z, Osman IF

Shalihin SE, Harun Z, Osman IF. Non-specific skin purpura. *Malays Fam Physician*. 2018;13(2):39–41.

Keywords:

Purpura, Thrombocythemia, Essential.

Authors:

Mohd Shaiful Ehsan Shalihin

(Corresponding author)

MBBS UIAM

Family Medicine Trainee Lecturer

DU51p, Department of Family

Medicine, International Islamic

University of Malaysia, Kuantan

Malaysia.

Email: shaifulehsan@iium.edu.my

Zulkifli Harun

MD UKM

Department of Family Medicine

Universiti Kebangsaan Malaysia

Malaysia.

Iskandar Firzada Osman

MMed Family Medicine USM

Klinik Kesihatan Jaya Gading

Kuantan, Malaysia.

Abstract

Essential thrombocythemia is one of the myeloproliferative neoplasms. Palpable purpura is a rare manifestation that may delay diagnosis and treatment. We report a case of essential thrombocythemia in a 50-year-old man, who presented with recurrent thigh pain for the past one year with nonspecific localized purpura. His full blood count revealed isolated thrombocytosis of 880,000/ μ L with an impression of myeloproliferative disorder from peripheral blood film. He was referred urgently to the hematology team, which proceeded with a venesection. His condition improved with hydroxyurea. This was a rare case of chronic presentation of myeloproliferative neoplasm detected at a primary care clinic.

Introduction

Essential thrombocythemia is a nonreactive, chronic myeloproliferative disorder in which sustained megakaryocyte proliferation leads to an increase in the number of circulating platelets. Its incidence is less than 3 per 100,000 in population per year, and it occurs mainly in older age groups. The majority of patients are asymptomatic until an incidental blood count finding or presenting with a thrombotic event. It is characterized by a persistently elevated platelet count of greater than 450,000/ μ L, megakaryocytic hyperplasia, and the presence of splenomegaly. Morbidity includes large-vessel or microvascular thrombosis and bleeding. A localized, palpable skin purpura is, in fact, a rare manifestation for which other differential diagnoses need to be considered at the first visit.

Case Report

A 50-year-old man, with an underlying, reducible internal hemorrhoid under surgical follow-up, presented to our clinic at Jaya Gading with recurrent symptoms of right thigh pain for the past one year. The pain had been increasing in severity for the past six months, with skin changes on the right thigh. The pain had no specific aggravating or relieving factors and was radiating to his toes. Physical examination revealed mild hepatosplenomegaly. No other mass or lymph nodes were palpable. There were no neurological deficits or musculoskeletal

deformities detected. However, there was a localized, palpable purpura skin lesion over his right thigh; it was non-blanching, non-tender, and was not warm on palpation. Peripheral pulses were palpable and equal. He denied any similar skin lesions or pain elsewhere. He denied any history of falls, atopy, recurrent fever, or bleeding tendencies.



Figure 1: Non-specific, palpable purpura skin lesion on patient's thigh during patient's clinic visit.

An urgent full blood count revealed isolated thrombocytosis of 880,000/ μ L. His hemoglobin and leukocyte counts were normal. Peripheral blood film showed thrombocytosis with the presence of megakaryocytes with an impression of myeloproliferative disorder. He was referred and admitted to the hospital immediately after the blood film result came out. The pre-venesection platelet count was 943,000/ μ L, and the post-venesection count was 798,000/ μ L after removing 450 ml of blood. He was started on aspirin and hydroxyurea. His next platelet count during regular follow-up was 501,000/ μ L. He was otherwise asymptomatic, and his localized skin lesion had improved. He is positive for the JAK 2 mutation, which is evidence that supports the presence of the disease.

Discussion

Purpura is a non-blanchable discoloration of the skin or mucous membrane which is related to disorders of small blood vessels or a disorder of their intravascular components.¹ Palpable purpura is a sign of inflammation of underlying blood vessels in which the purpura can be felt. During the first encounter, differential diagnoses related to platelet disorders, such as vascular disorders or infections, need to be ruled out.^{2,3} Our patient did not present with any history suggestive of hematological malignancy, such as bleeding tendencies, recurrent fever, or anaemic symptoms. There were no symptoms of autoimmune diseases, such as multiple joint pain or back pain. He denied any neck swelling or masses elsewhere. There was no relation to drugs. Absence of those histories would cause difficulty in narrowing down the possible diagnosis.

In fact, our patient presented with the nonspecific symptom of localized thigh pain, which may cause the treating doctor to mistakenly attribute the disease to localized inflammation or trauma. The pain our patient suffered differed from thrombotic pain as he denied any intermittent claudication induced by activity or heat. There was no distal numbness. He described the pain as dull aching and not disturbing his quality of life. This sinister disease would have been definitely missed if the treating physician had not inspected and examined the thigh thoroughly or had assumed it to be a non-related benign soft-tissue injury, such as a muscle strain.

Another issue is the appearance of the skin lesion, which is not typical of the flat brownish-red spots of purpura. His skin lesions were raised, with papular- to nodular- like features, but they still had a similar brownish-red background color, although not to the extent of being red-purple. This could be due to the long duration of the presentation. Nevertheless, they were non-blanchable, as expected in purpura. Overall, these skin lesions, described as “palpable purpura,” are more common in vasculitis than in myeloproliferative disorders, such as essential thrombocytopenia.^{4,5,6}

Essential thrombocythemia is one of four myeloproliferative disorders manifested by overproduction of platelets by the megakaryocytes in the bone marrow.^{7,8} Even though it is rare, it may develop into acute myeloid leukemia or myelofibrosis. Most

patients are asymptomatic and might present incidentally with isolated thrombocytosis during a blood investigation for another disease. Meanwhile, in a symptomatic patient, the most common presentation is with thrombotic features that cause burning pain, numbness, or dusky discoloration of the extremities. The pain is typically aggravated by exposure to heat and improves with cold exposure.^{7,8,9} Patients may also present with neurological disorders, such as migraine headaches, dizziness, or even transient ischemic attacks. However, none of these features are associated with our case, except the intermittent localized pain.

At a primary care level, a thorough history and examination are indeed helpful initial assessment tools when investigating any symptoms that come to attention. These items can provide an essential list of probable diagnoses, including alarming diseases that usually present with red flags.² In our case, other than the nonspecific history, the finding of mild splenomegaly did provide us with an important clue that an underlying hematological disease needed to be ruled out. His splenomegaly could have been missed if we had not elicited for Traube's space dullness during the abdominal examination.

A *full blood* count is not only essential for an initial investigation of a chronic skin presentation, but it is also useful as an adjunctive supporting tool for patients presenting with chronic symptoms with any degree of splenomegaly. In our case, the patient presented with nonspecific thigh pain, which was unusual as it had been occurring in the same area for up to a year. The presence of purpura, in fact, added further to the need for performing a full blood count to rule out any bleeding disorder, specifically low platelets. Surprisingly, an extremely high level of platelets was detected.

Essential thrombocythemia is characterized by the following criteria: 1) persistent thrombocytosis greater than 450,000/ μ L from a full blood count, 2) megakaryocytic hyperplasia, 3) splenomegaly, and 4) a thrombotic or hemorrhagic event. Our case presented with all of these related features.^{4,7,8} His positive JAK2 mutation indicates a poor prognosis for his disease, for which early treatment is needed. He benefited from low-dose aspirin and hydroxyurea and was symptom-free during follow-up, with his purpura resolved. Hydroxyurea is generally

considered to be the first-line drug for cytoreductive therapy in essential thrombocythemia.

Conclusion

This case has indeed proven that a focused and proper history taking, a directed and appropriate physical examination with a high index of suspicion, as well

as a selective and simple supportive investigation commonly practiced in a primary care setting play an important role in identifying uncommon presentations of uncommon medical conditions, which, in this case, led to a timely intervention. Chronic palpable purpura may signify hematological malignancies rather than vasculitis, especially in males and those aged 40-years-old and above.

How does this paper make a difference to general practice?

- Though platelet disorders usually present with flat, red-purplish purpura, they can also present with palpable, raised skin lesions, which reflect chronicity, especially in hematological malignancy. Therefore, primary care providers need to differentiate palpable purpura from chronic urticaria, which may also present with similar skin lesions, although they are usually generalized.
- This case report shows that detailed history taking and a complete, thorough examination can identify the possible diagnosis, even without an investigation, at the primary care level. Traube's space dullness needs to be elicited in any abdominal examination whenever the spleen is not palpable in order to not to miss mild splenomegaly, which provides an important clue towards an underlying suspected hematological disease.
- This case report proved that a simple full blood count can provide an impressive amount of information for primary care practitioners, even without a peripheral blood film, in chronic skin lesions.
- Skin lesions requires a proper examination, including inspection of the surrounding area to look for alarming signs. Differential diagnosis can be further divided into blanchable or non-blanchable skin erythema.
- This case report showed that pain is one of the alarming symptoms in any skin disorder. Further characterization of the pain needs to be conducted by the primary care provider to rule out other, non-related diseases.

References

1. Kelly, Robert. (2017). "Approach to the Patient with Retiform (Angulated) Purpura." <http://www.uptodate.com> Retrieved 21 April, 2017.
2. Murtagh, J. and J. Rosenblatt. (2011). *Murtagh's General Practice, Fifth Edition*, McGraw-Hill Medical.
3. Longmore, M., et al. (2010). *Oxford Handbook of Clinical Medicine Oxford*.
4. Sehkar, M. (2015). *London Cancer Essential Thrombocythemia Guidelines 2015*.
5. Gotta, Carmen. (2017). "Overview of Cutaneous Small Vessel Vasculitis." <http://www.uptodate.com> Retrieved 20 April, 2017.
6. Fett, Nicole. (2017). "Evaluation of Adults with Cutaneous Lesions of Vasculitis." <http://www.uptodate.com> Retrieved 21 November, 2017.
7. Lal, A. (2016). "Essential Thrombocytosis." *Medscape*. Retrieved 27 November, 2016.
8. Tefferi A. (2016). "Diagnosis and Clinical Manifestations of Essential Thrombocythemia." <http://www.uptodate.com> Retrieved 20 November, 2016.
9. Aster, John C. and M. Richard. (2017). "Clinical Manifestations and Diagnosis of the Myelodysplastic Syndromes." <http://www.uptodate.com> Retrieved 20 April, 2017.