Rosacea Associated with Polycythemia Vera

— *Skin Lesions Improved with Phlebotomy* —

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We report a case of rosacea in a 65 year old female with a 14 year history of polycythemia vera. The patient suffered from several constitutional symptoms and signs suggestive of polycythemia vera. Six years prior to our initial examination, erythematous lesions were first noted on the center of the face. These lesions exhibited periodic improvement and exacerbation without specific treatment.

Histopathologic examination of the facial lesions showed nodular infiltration of lymphocytes and histiocytes and dilation of blood vessels. On the basis of laboratory examination, the patient was diagnosed as polycythemia vera.

The patient has received metronidazole, tetracycline, and topical steroids to control rosacea-like facial lesions. The treatment results were not significant. However, phlebotomy markedly improved the skin lesion. To our knowledge, there are no reports describing the clinical course of rosacea lesions following the treatment with phlebotomy.
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Key Words: Phlebotomy, Polycythemia vera, Rosacea

Polycythemia vera(PV) is one of the myeloproliferative disorders characterized by splenomegaly and an absolute increase in the number of circulating red blood cells, leukocytes, and platelets[1].

Cutaneous changes are characteristic, and are mainly attributed to increased blood viscosity and vascular distension. These changes include rubor of the skin and mucous membranes, rosacea, and spider nevi[1-3].

We describe a case of rosacea associated with polycythemia vera, which showed significant improvement of skin lesions after phlebotomy.

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REPORT OF A CASE

A 65 year old female patient was referred to the Department of Dermatology due to erythematous lesions on the face for 6 years. Her medical history included PV 14 years previously. Family history was not contributory.

Since the development of PV she suffered from a generalized itching sensation, easy fatigability, and frequent facial flushing. Erythematous skin lesions were first noted on the central portion of the face 6 years prior to our examination, follwed by a period of improvement and exacerbation without specific treatment. Occasionally, cyanotic rubor, purpura, and hemosiderosis were observed on the face and extremities. She also complained of an itching sensation and discomfort due to skin problems. Recently, she also suffered from epigastric distress due to a greatly enlarged spleen. She has received irregular treatments with phlebotomy at a local clinic to control polycythemia.

On physical examination she had a chronically ill appearance with a massive protruded abdomen caused by splenomegaly. The skin color of the hand was more cyanotic, or bluish hue, than that of a normal person. Slight scaly erythematous patches, papules, and palques with telangiectasia were largely located on the center of the face, such as the nose, periocular, and perioral areas (Fig. 1). She also had a conjunctival confusion and blepharitis.

Laboratory studies showed marked erythrocytosis, greatly elevated hemoglobin concentration, hematocrit volume, and leukocytosis with a left shift of differential counts(Table). A chest x-ray showed cardiomegaly and a prominent aortic arch. The results of urinalysis, liver function test, and electrocardiography were negative, or within normal limits. Direct smears for fungi and Demodex folliculorum from the facial lesions were negative. Bone marrow examination revealed either erythroid hyperplasia or panhyperplasia without distinctive morphologic features. Evidence of secondary polycythemia, such as chronic pulmonary or cardiac disorder, was lacking. Histopathologic study of the facial papular lesions revealed nodular lymphohistiocytic infiltrations and marked dilation of blood vessels in the entire dermis(Fig. 2).

Based on clinical manifestations, laboratory data, and histologic indicators we diagnosed this case as rosacea resulting from increased blood viscosity in PV.

The patient has received metronidazole, tetracycline, and topical steroids to control the facial lesions during about 6 months. Although the skin lesions showed mild to moderate improvement, we and the patient were not satisfied(Fig. 3). However, when the patient received phlebotomy of approximately 480 ml of whold blood at the Department of Internal Medicine, she experienced a significant improvement of the facial lesions 2 weeks after phlebotomy(Fig. 4). At that time, hemoglobin concentration, hematocrit volume, and white blood cell count were 12.7 gm%, 44 vol. %, and 23,000 per square millimeter, respectively. These results showed a marked improvement compared with the levels prior to phlebotomy. However, the improvement of skin lesions was temporary. New lesions started to reappear on the face about 2 weeks after phlebotomy. At the time of reappearence hemoglobin concentration, hematocrit volume, and leukocyte count were increasing compared to the levels two weeks earlier(Table 1).

Table 1. Comparison of laboratory data and skin lesions relating to phlebotomy

	Before phlebotomy	After Phlebotomy	
		2 weeks	4 weeks
RBC(10^6/mm^3)	8.19	4.86	5.50
Hgb(gm %)	19.2	12.7	13.6
Hct(vol.%)	67	44	48
WBC(/mm^3)	32,000	23,000	23,200
Skin lesion	+++	-	+

+++ : severe rosacea + : mild rosacea - : improved

DISCUSSION

Polycythemia vera is characterized by splenomegaly and an increased production of all myeloid elements[1]. PV begins in late middle life and is slightly more common in males[1].

PV patients typically experience headache, vertigo, weakness, dyspnea, and epigastric distress. Cutaneous changes include rosacea, spider nevi, hemangiomas, hemorrhage, ecchymosis, acne urticata, and erythroderma. These constitutional symptoms and cutaneous signs are chiefly caused by increased blood volume and viscosity[2,3].

Our patient suffered from several constitutional symptoms suggesting PV. On admission, we observed cyanotic rubor, and ecchymotic and hemosiderotic patches on the extremities. Moreover, she had had rosacea like skin lesion on the center of the face for 6 years.

Rosacea is a chronic skin disorder affecting the center of the face[4,5]. It is usually responsive to tetracycline or metronidazole[5], but our patient did not showed significant clearing of the lesions from these drugs. Rather, phlebotomy caused the facial lesions to improve significantly.

Although many factors have been considered in the etiology of rosacea, none of them has been shown to be the direct cause of the disease[5]. However, it has been suggested that rosacea in PV is attributed to increased blood viscosity[1]. Even though there were reports that a reduction of the hematocrit and blood volume by phlebotomy could lead to improve constitutional symptoms[6,7],

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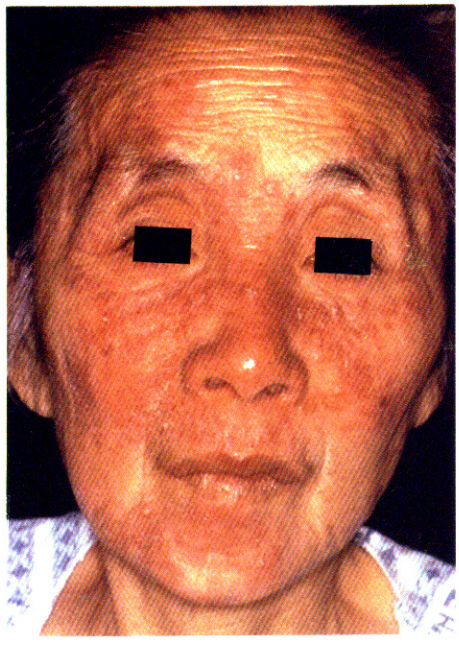

Fig. 1. Erythematous patches, papules, and plaques on the central areas of the face, such as the nose, periocular, and perioral areas.

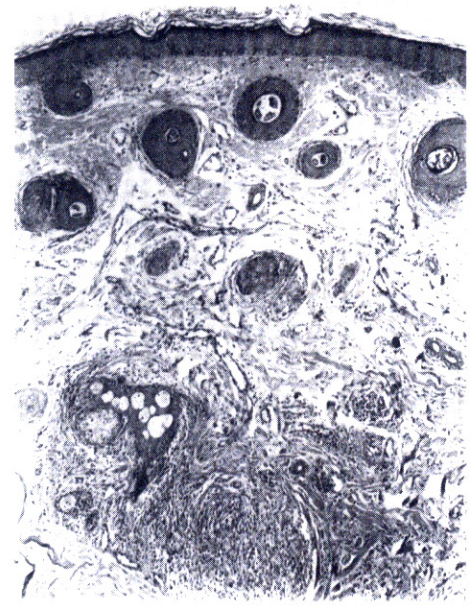

Fig. 2. Nodular infiltration of lymphohistiocytes and marked dilation of blood vessels in the entire dermis from the histopathological study of the papular skin lesions(H & E stain, × 40).

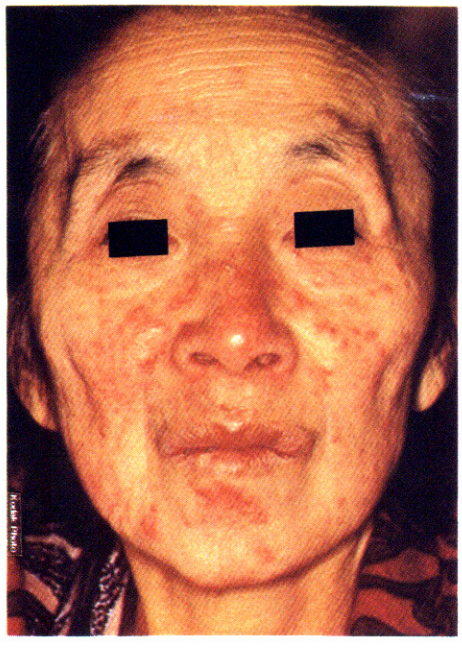

Fig. 3. Moderate improvement in facial lesions 4 weeks after metronidazole treatment.

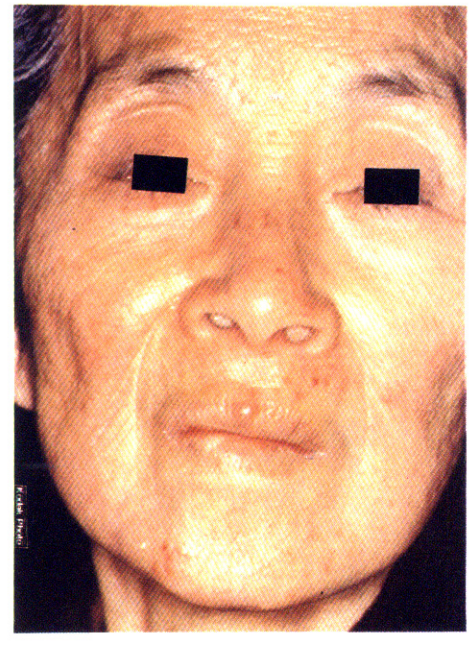

Fig. 4. Significantly improved facial lesions 2 weeks after phlebotomy.

we could not find any reports relating improvement of rosacea to phlebotomy. When we analyzed the relationship between the clinical course of the skin lesions and laboratory data for several weeks, we interpreted that the skin manifestations of our patient might be influenced by changes in the red blood cell number and concentration (Table 1). We repeatedly observed this pattern of improvement and exacerbation of facial lesions after phlebotomy, though we have not described this in detail in the case report. But, we could not explain why the facial lesions reappeared 2 weeks after phlebotomy, despite complete blood counts being within normal range(Table). So, we could not completely exclude the possibilities of other factors the skin lesions to reappear at that time, in addition to the change of blood concentration. In conclusion, we think a further pathogenetic understanding of rosacea in PV would need a more and detail observation of rosacea patients with PV and long-term follow up studies.

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