FURTHER STUDIES OF PLATELET ROSETTES AROUND GRANULOCYTES IN BEHCET’S SYNDROME

G.E. EHRLICH, M. KAJANI, I.R. SCHWARTZ and R.F. McALACK
Sections of Rheumatology (Arthritis Center) and Hematology and the Renal Transplantation Laboratory, Albert Einstein Medical Center, Philadelphia, Pennsylvania

Abstract—Rosettes of platelets around granulocytes—platelet satellitism—previously described in Behcet’s syndrome led to the discovery of a case that may have been 80 years in duration. A strong relationship between calcium ions and the phenomenon was suggested by its specificity in edetic-acid-anticoagulated blood, and the subsequent migration of platelets on supravital preparation from around neutrophils upon addition of 0.2 M calcium chloride to heparinized EDTA-treated blood. Plasma from the patient was able to cause the phenomenon with donor granulocytes and platelets. Platelet agglutinins were also demonstrable. Specificity in Behcet’s syndrome is possible, but remains unproved.

INTRODUCTION

Rosettes of platelets clustering around granulocytes had previously been described only in a patient with thrombocythemia (1), when Prchal and Blakeley discovered a similar phenomenon in a patient who had Behcet’s syndrome (2). Their letter to the editor led to the discovery of another case of Behcet’s syndrome and further studies of the phenomenon.

CASE REPORT

An 83-year-old white woman of remote English extraction, whose family had long lived in Maryland, was admitted for extraction of a cataract from the right eye. She had been deaf and dumb since the age of 3, following an attack of “meningitis”. The history as recorded by the attending physician and house staff was otherwise quite unremarkable. When the differential count was done preoperatively, the technician noted that all the platelets were clustered in rosettes around granulocytes. She drew this to the attention of an attending
hematologist, who could offer no ready explanation, as the platelet count had not shown an excess of thrombocytes. The discussion was overheard by one of us (I.R.S.) and remembered a few days later when the next issue of the *New England Journal of Medicine* contained a letter to the editor, with appropriate illustration (2). Knowing of the interest of one of the co-authors (G.E.E.) in Behcet's syndrome, this phenomenon was drawn to his attention. The study of the hospital chart was unrevealing; the cataract extraction had meanwhile been performed, and a vitreous rupture had been repaired with vitrectomy on November 15, 1973.

The patient was interviewed, with her daughter acting as interpreter, using sign language. The daughter and granddaughter were able to add further details of the history.

The details of meningitis at age 3 are vague, but it is known that the patient never spoke or heard thereafter. Recurrent crops of aphthae began shortly thereafter and continued throughout her life. These ulcers involved the labial and buccal mucosa and the tongue and soft palate, were painful enough to interfere with eating, and lasted for up to 3 wk each time, followed by an asymptomatic interval of up to 3 months. At age 18, an inflammation of the eyes developed, requiring surgery to the left eye. Ophthalmologic examination at this time suggested that this operation was a paracentesis of the anterior chamber, an old form of treatment for hypopyon iritis. Shortly thereafter, migratory arthralgias began, with occasional monarticular arthritis involving a knee, although other large joints were sometimes afflicted. Arthritic involvement was characterized by synovitis of a transient nature, lasting 2–3 wk. Painless vulvar ulcers had been discovered by the granddaughter while washing the disabled grandmother; they, too, came in recurrent crops, although the length of time during which they have recurred is unknown. There was no history of skin lesions. Atrophic papillae of the tongue had been noted on several occasions during her life. There were recurrent episodes of peripheral thrombophlebitis, both in the upper and lower extremities throughout her life. There was a history of intermittent vaginal bleeding, whose source was never discovered. Chronic obstipation was present, and roentgenograms revealed a megacolon. In the past, there had been recurrent rectal bleeding the source of which was never discovered.

The patient's 8 children and many grandchildren are all alive and well, with no history suggestive of similar symptoms. However, the patient's sister has several grandchildren who have hearing loss, supposedly from infection. The patient's mother died of intracranial hemorrhage at age 38, and the father of carcinoma of the stomach at age 50.

Examination on transfer of the patient to the Arthritis Center confirmed aphthous lesions of the labial mucosa, throughout the oral cavity, and many clusters in the posterior pharynx. Vulvar ulcers were present, but no cervical ulcers were found. Bilateral cataracts and bilateral corneal dystrophy were recorded.

A complete laboratory work-up concentrated on historical abnormalities. Erythrocyte sedimentation rate was 70 mm/h (Westergren). There were 124,000 platelets/mm$^3$. Hemogram was otherwise normal, as were urinalysis, blood chemistries with the exception of potassium (3.0 mg/100 ml), and protein electrophoresis. Antinuclear antibody, anti-DNA antibody, latex fixation test for rheumatoid factor, and LE preparation were negative. VDRL was weakly reactive, but FTA was negative. C'3 (beta 1C) complement component was 145 (normal: 120–185). Roentgenograms and electrocardiogram were unremarkable, with the exception of diverticulosis demonstrated on barium enema after relief of the fecal impaction.

A plasma transfusion was given, and within 2 days, the oral and vulvar ulcers had almost disappeared. After discharge, these ulcers recurred 6 wk later, and a further plasma transfusion was given, again with ready response. Aspiration of the right knee at this time yielded translucent joint fluid of normal viscosity, containing 100 WBC, no crystals, and