Primary systemic amyloidosis with malignant pinch purpura: A case report.

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ABSTRACT:

SASTARARUJI D, KOOTIRATRAKARN T, PRATCHYAPRUIT W. PRIMARY SYSTEMIC AMYLOIDOSIS WITH MALIGNANT PINCH PURPURA: A CASE REPORT.

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Bilateral periorbital purpura (raccoon's eyes) has been associated with many causes such as traumatic (basal skull fracture and rhinoplasty), neuroblastoma, lymphoma, kaposi's sarcoma and primary systemic amyloidosis. We report 88-year-old man presented with a 6 months history of extensive periorbital hemorrhagic nodular plaques forming gross distortion which disturbed his eye-fields and numerous translucent and hyperpigment papules over trunk and back. Histopathologic study from the skin lesions at periorbital region and back showed amorphous eosinophilic material deposit in superficial to deep dermis and around blood vessel wall. After Congo red staining, the sample showed a red-green birefringence and dichroism under polarized light microscopy. Further work-up revealed free light chain (kappa) on urine protein electrophoresis and serum immunofixation electrophoresis showed IgG kappa monoclonal gammopathy. According to these findings, the diagnosis of primary systemic amyloidosis

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associated with plasma cell dyscrasia was made. In conclusion, this case presented with massive periorbital hemorrhagic nodular mass and numerous papular eruptions which were rarely seen in systemic amyloidosis. Malignant pinch purpura was shown as a crucial clue in the diagnosis of systemic amyloidosis in our patient who has no other systemic complaints.

Key words: pinch purpura, primary systemic amyloidosis

บทคัดย่อ :

ดาดนุช ศาสตระรุจิ, ธนวัฒน์ คูถิรตระการ, วลัยอร ปรัชญพฤทธิ์ รายงานผู้ป่วยที่มีจ้ำเลือดออกรุนแรงรอบตาในโรคอะมัยลอยโดสิส วารสารโรคผิวหนัง 2560; 33: 209-214.

สถาบันโรคผิวหนัง กรมการแพทย์ กระทรวงสาธารณสุข

ภาวะจ้ำเลือดออกรุนแรงรอบตาสามารถพบได้จากอุบัติเหตุ, มะเร็งต่อมน้ำเหลือง, มะเร็งต่อมหมวกไต (neuroblastoma), มะเร็งคาโปซี (kaposi's sarcoma) และโรคอะมัยลอยโดสิส (primary systemic amyloidosis) รายงานฉบับนี้เป็นการนำเสนอผู้ป่วยชายอายุ 88 ปี ที่มีจ้ำเลือดออกขนาดใหญ่ รอบตา จนรบกวนการมองเห็น และมีตุ่มนูนสีใสและสีดำตามลำตัว หลัง ปริมาณมาก มานาน 6 เดือน ลักษณะทางพยาธิวิทยาและย้อมพิเศษเพิ่มเติมของจ้ำเลือดออกรอบตาและตุ่มนูนสีดำที่หลัง เข้าได้กับโรคอะมัยลอยโดสิส ทำการสืบค้นเพิ่มเติม พบมีการเพิ่มขึ้นของอิมมูโนโกลบูลิน (monoclonal gammopathy) ทั้งในเลือดและปัสสาวะ เข้าได้กับ โรคอะมัยลอยโดสิส ที่มี plasma cell dyscrasia ผู้ป่วยรายนี้มาด้วยผื่นจ้ำเลือดออกขนาดใหญ่รอบตา และตุ่มนูนตามลำตัวปริมาณมาก ลักษณะผื่นดังกล่าวพบน้อยในโรคอะมัยลอยโดสิส ในผู้ป่วยของเราที่ไม่มีอาการทางระบบ ผื่นจ้ำเลือดออกรุนแรงรอบตา เป็นสิ่งชี้นำสำคัญในการสืบค้นเพิ่มเติมโรคอื่นต่อไป

คำสำคัญ: ภาวะจ้ำเลือดออกรุนแรงรอบตา, โรคอะมัยลอยโดสิส

Case report

A 88-year-old man presented with a 6 months history of marked periorbital swelling with hemorrhagic component which disturbed his eye-fields and extensive purpura of trunk and all extremities. No bleeding in the other parts of the body was reported. Besides benign prostatic hyperplasia, his general health seemed to be all right.

On physical examinations, the patient was a thin, elderly man with good consciousness. His sight was blocked by bilateral eyelid swelling and hemorrhagic nodular plaques (Figure 1). Mild ectropion was also noted. The extensive purpuric patches were seen on the face, trunk, and all extremities. Multiple translucent papules, hyperpigment papules, comedones and skin erosion over the trunk and back (Figure 2). There

was large hemorrhagic flaccid bulla located on a noninflammtory purpuric base on right leg (Figure 3). No macroglossia was detected. The remainder of the examination was unremarkable.

Figure 1 Bilateral massive periorbital purpura nodules causing visual impairment.

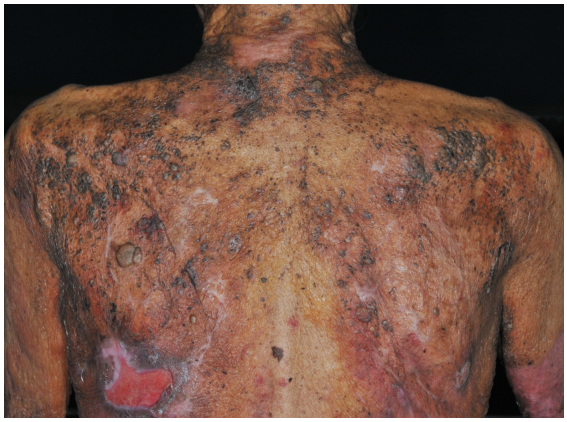

Figure 2 Multiple translucent papules, hyperpigment papules, comedones, scars and skin erosion over the back.

Figure 3 Large hemorrhagic flaccid bulla on a noninflammtory purpuric base on right leg.

On blood analysis, anemia (hemoglobin 9.4 g/dl), renal impairment (creatinine 1.5 mg/dL), hypercalcemia (calcium 10.9 mg/dL), hypoalbuminemia (albumin 2.3 g/dl) and proteinuria (protein 2+) were detected. A skin biopsy was performed at hemorrhagic plaque on periorbital region and hyperpigment papule on back and both revealed thinning of epidermis and amorphous eosinophilic material deposit in superficial and deep dermis (Figure 4) and around blood vessel wall (Figure 5). After Congo red staining, the sample showed a red-green birefringence and dichroism under polarized light microscopy consistent with systemic amyloidosis (Figure 6).

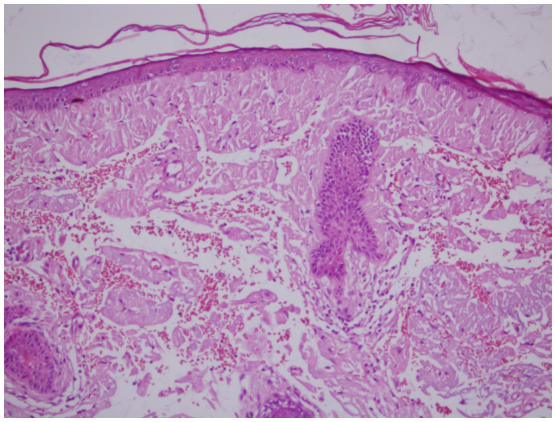

Figure 4 Eosinophilic materials deposit in superficial to deep dermis and periadnexal (Hematoxylin-eosin stain, original magnification ×20).

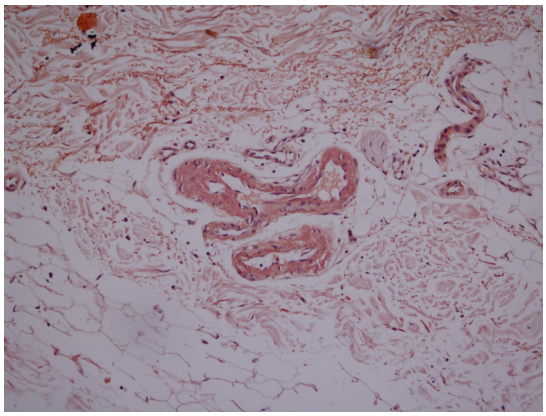

Figure 5 Congo red staining reveals the presence of amyloid around blood vessel wall (Congo red stain, original magnification ×10).

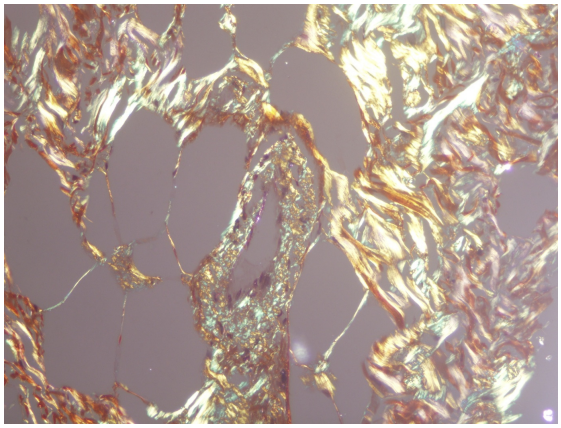

Figure 6 Polarized microscopy of section stain with Congo red demonstrated apple green birefringence around blood vessel wall and fat lobules (original magnification ×20).

After establishing the diagnosis of amyloidosis in the skin lesion, the patient underwent further investigations. Serum free light chain (SFLC) assay on urine protein found an increase in free lambda chain (kappa) at 747 mg/dL (reference, 3.3-19.4) and serum immunofixation electrophoresis showed IgG kappa monoclonal gammopathy. Unfortunately, the patient refused for further investigations, thus; prothrombin time and activated partial prothrombin test, echocardiogram, electrocardiogram, ultrasonography of abdomen, bone marrow aspiration biopsy and subcutaneous fat aspirate were not done. The patient died 5 months after the diagnosis of primary systemic amyloidosis with plasma cell dyscrasia was done.

Discussion

Primary systemic amyloidosis (AL) is a rare disease with an incidence in Sweden was 3.2 per million inhabitants per year[1]. However, it is not well documented in Thailand. It is a serious problem of malignant or benign monoclonal gammopathy. Up to 10–20% of patients with primary systemic amyloidosis have an underlying multiple myeloma[2]. It occurs most commonly in elderly men with mean age of onset is 65 years[3]. About one-third of patients with primary systemic amyloidosis have mucocutaneous manifestation which is a hint to early diagnosis of existence of an underlying plasma cell dyscrasia[4]. It usually presents as purpuric lesions on the eyelids and the flexural regions[4] which develop suddenly or after minor trauma, vomiting, the Valsava maneuver, coughing[5] result from capillary fragility associated with amyloid deposit in dermal and sub cutis blood vessel walls[6]. However, massive swelling of the eyelids and hemorrhagic eyelid mass causing visual impairment, as in our case, are rarely seen in primary systemic amyloidosis[7]. Although, basal skull fracture and rhinoplasty, neuroblastoma, lymphoma and kaposi's sarcoma[8] can be causes of periorbital purpura (raccoon's eyes). In addition, waxy yellowish translucent papules may be found on the face, flexural and the buccal mucosa, these are uncommon[9]. Our patient had multiple shinny yellowish translucent and hyperpigment papules on trunk and back. A biopsy from skin lesions over back was consistent with the diagnosis of amyloidosis. Other less common clinical presentation included bullous lesions, pachydermia of the scalp, alopecia, subungual striations and anonychia[10]. Other from this, amyloid can deposit in many organs such as renal which presents with nephrotic syndrome, hypertension and severe renal failure, cardiac that can progress to congestive heart failure, peripheral neuropathy and autonomic neuropathy.

A majority of patients dying within 13 month without therapy. In Two major trials, a combination with melphalan and prednisolone is more effective, compare with no therapy and therapy with colchicine alone, but survival rate were still short.[11,12]

In conclusion, we present a rare case of primary systemic amyloidosis who presented with massive bilateral periorbital hemorrhagic nodular plaque causing visual impairment and multiple papular eruptions which are rarely seen in primary systemic amyloidosis. Our patient had no other systemic complaints, and unique periorbital purpura (raccoon's eyes) was the only diagnostic clue for underlying disease. This case demonstrates uncommon type of skin manifestations in primary systemic amyloidosis and importance of an initial skin manifestation

that leads to the diagnosis of underlying systemic diseases.

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