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Case report

Isolated Cutaneous Glomeruloid Hemangioma: A Case Report in the Absence of POEMS Syndrome and Castleman Disease

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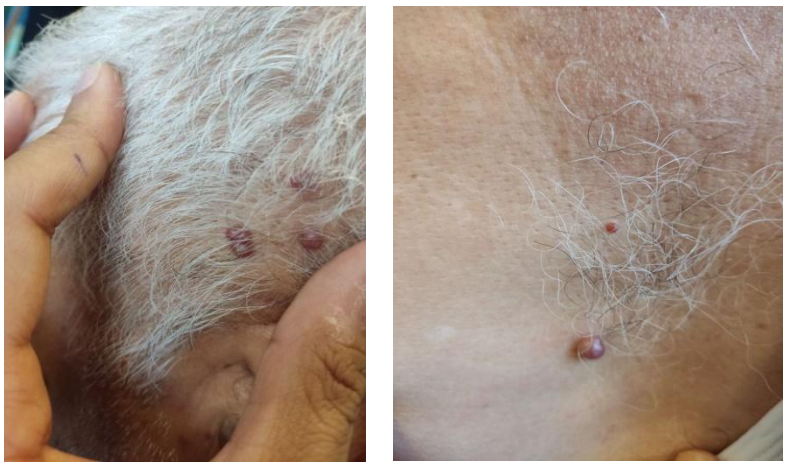
Abstract
Glomeruloid hemangioma is a rare, benign vascular proliferation of the skin, clinically manifests as multiple red-purple papules or nodules, predominantly affecting the chest and upper extremities. Histopathologically, it is defined by dermal capillaries arranged within vascular spaces, closely resembling a renal glomerulus. The presence of glomeruloid hemangioma is considered a diagnostic clue for systemic disorders such as polyneuropathy, organomegaly, endocrinopathy, M-protein skin abnormality syndrome (POEMS), and Castleman disease, all of which share a similar proinflammatory cytokine profile. We report the case of a 64-year-old male who presented with multiple painless, slowly progressive erythematous papules and nodules over more than one year, involving the face, trunk, and upper extremities. An excisional biopsy revealed characteristic histopathological features consistent with glomeruloid hemangioma. Comprehensive clinical and laboratory evaluations revealed no systemic involvement, suggesting an isolated cutaneous presentation. Although glomeruloid hemangioma is strongly associated with POEMS syndrome and other systemic diseases, it may occasionally occur without systemic manifestations.
Keywords: Glomeruloid Hemangioma, POEMS Syndrome, Castleman Disease, Case report

Introduction

Glomeruloid hemangioma (GH) is a rare, benign vascular proliferation of the skin, first characterized by Chan et al. in 1990. Clinically, it presents as multiple red-purple papules or nodules, predominantly affecting the chest and upper extremities. Histopathologically, it is defined by dermal capillaries arranged within vascular spaces, closely resembling a renal glomerulus [1]. The rapid appearance of lesions within a few days or weeks is often associated with POEMS syndrome, a paraneoplastic disorder resulting from a plasma cell dyscrasia. The term, introduced by Bardwick in 1980, encompasses polyradiculoneuropathy, organomegaly, endocrinopathy, monoclonal plasma cell dyscrasia, and dermatological alterations, along with Castleman disease, all of which share a similar proinflammatory cytokine profile [2]. On the other hand, lesions that develop more gradually may occur in individuals without any related symptoms. Similar isolated cases of GH have been reported without systemic involvement, highlighting that GH can occur independently, particularly with a gradual onset.

Case presentation

a 64-year-old male with painless, slowly progressing erythematous papules and nodules over the past year, predominantly affecting the face, chest, and trunk. Physical examination revealed seven non-tender, firm, compressible, sessile, red-to-violaceous dome-shaped papules and nodules measuring 1–2 cm in diameter (Figure 1).



(a) **(b)**

Figure 1. (a) Red-to-violaceous dome-shaped papules and nodules on the scalp; (b) Similar nodules found on the chest.

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Systemic examination, including a neurological assessment, showed no abnormalities. Blood and urine tests, as well as a CT scan, were unremarkable.

The differential diagnosis included Kaposi sarcoma, Masson tumor, angiosarcoma, and intravascular pyogenic granuloma. An excisional skin biopsy and histopathological examination revealed circumscribed dermal ectatic vascular spaces lined by swollen endothelial cells. Larger central vessels were formed by sinusoidal endothelial cells, while smaller projections into the vascular spaces resembled renal glomeruli, composed of capillary endothelial cells interspersed with stromal cells. These findings were periodic acid-Schiff (PAS)-negative (Figure 2). Based on these distinctive histopathological features, a diagnosis of glomeruloid hemangioma was made.

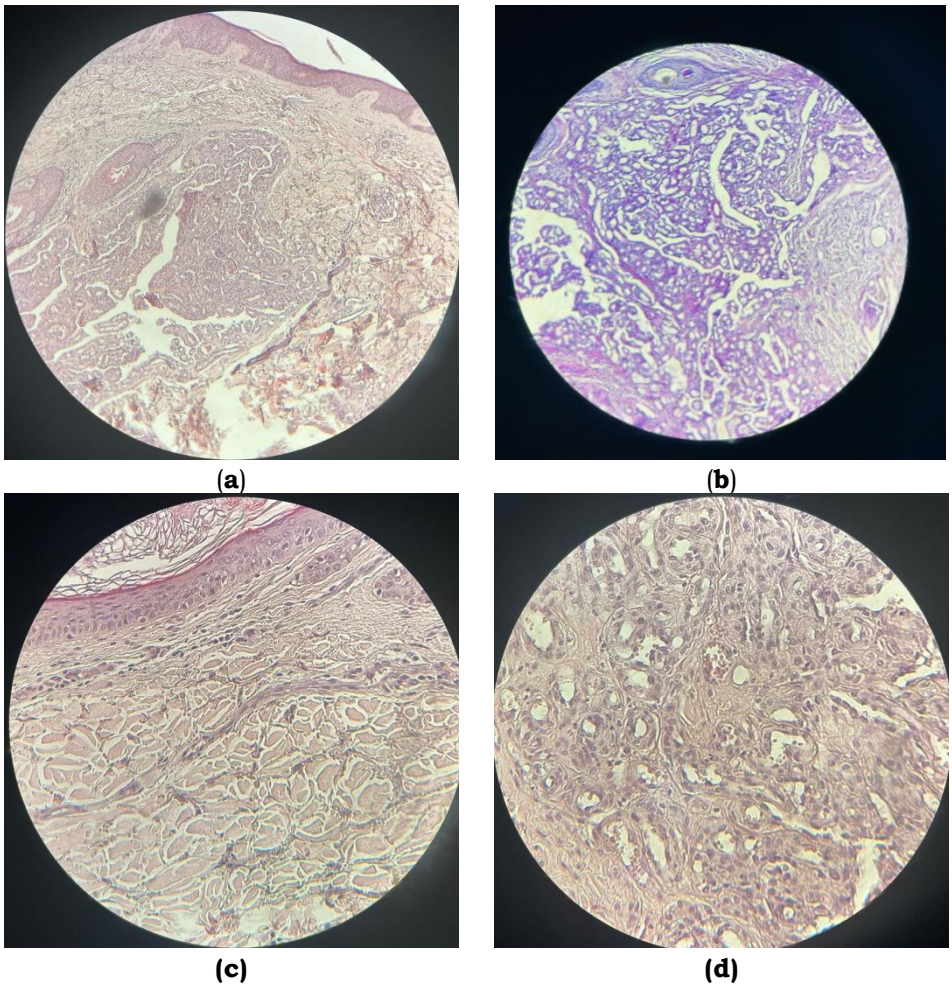

Figure 2. (a) Photomicrograph showing glomeruloid aggregates vascular structures in the dermis resembling renal glomeruli (H&E X 10); (b) periodic acid-Schiff stain X 10 demonstrating negative results; (c) projections extending into the vascular spaces composed of capillary endothelial cells interwoven with stromal cells (H&E stain X 40); (d) Larger central vessels are formed by sinusoidal endothelial cells containing red blood cells (Hematoxylin and eosin stain X 40).

Discussion

Chan et al. first described GH in 1990 as cutaneous hemangiomas discovered in two patients with Castleman's disease and POEMS syndrome. These lesions were associated with distinct renal glomerular-like endothelial features, giving rise to the name glomeruloid hemangiomas. Clinically, GH presents as firm or dome-shaped red papules, typically found on the trunk or upper limbs, although they can occasionally occur on the face or other areas of the body. Their size can vary from a few millimeters to several centimeters, and they may appear papulonodular, sessile, pedunculated, or cerebriform [1]. Until recently, these lesions were strongly associated with POEMS syndrome and, less commonly, with Castleman disease, and their presence prompted a full systemic investigation. However, more cases have emerged without association with systemic findings [3,4].

Although various hypotheses have been proposed, the precise pathogenesis of GH remains unclear. Several researchers suggest that lesion formation could be influenced by an excess of angiogenic cytokines and signaling molecules. Alternative hypotheses propose that increased immunoglobulin (Ig) deposition within endothelial cells may lead to enhanced endothelial proliferation in glomeruloid patterns, heightened estrogen

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levels, or infection with human herpesvirus-8 among patients with Castleman disease [4,5].
POEMS syndrome is a paraneoplastic disorder resulting from plasma cell dyscrasia. The term, introduced by Bardwick in 1980, includes polyradiculoneuropathy, organomegaly, endocrinopathy, monoclonal plasma cell dyscrasia, and dermatological alterations [2]. Motor impairment and a progressive onset of polyneuropathy are hallmarks of POEMS syndrome. This condition is driven by hypergammaglobulinemia, particularly M-protein monoclonal gammopathy, leading to the overproduction of myeloma protein, an aberrant monoclonal antibody produced by precancerous or malignant plasma cells. It is usually recognized by the presence of sclerotic bone lesions and IgA or IgG monoclonal gammopathy. About 50% of patients develop hepatomegaly, and up to 67% experience endocrine problems, such as diabetes mellitus and hypothyroidism. Additionally, GH is one of the cutaneous signs, along with hyperpigmentation, hypertrichosis, acrocyanosis, sclerodermoid changes, and leukonychia. GH, which occurs in 26% to 44% of affected individuals, is especially suggestive of POEMS syndrome, although it is not present in every patient and is considered a minor criterion in the diagnostic framework [6].
Lastly, POEMS syndrome is regarded as a malignancy, and treatments include autologous hematopoietic stem cell transplantation, systemic corticosteroids, radiation therapy, and alkylating agents [7]. However, GH is benign, and interventional therapy is generally unnecessary. Depending on individual patient needs, surgical excision, intralesional triamcinolone, oral prednisone, laser therapy, and thalidomide may be considered as alternatives. If glomeruloid hemangiomas are unresponsive to medical treatment or are aesthetically concerning, surgical removal may be performed [8].

Conclusion

Glomeruloid hemangioma is a benign condition that develops slowly and usually does not require further treatment. Patients diagnosed with GH should be evaluated for potential indicators of undiagnosed POEMS syndrome at the time of diagnosis and monitored over several years for the emergence of this syndrome.

Conflicts of Interest

The authors declare no conflicts of interest

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الملخص

الورم الوعائي الكبيبي هو تكاثر وعائي حميد نادر في الجلد، يتجلى سريريًا على شكل حطاطات أو عقيدات متعددة حمراء-أرجوانية، تؤثر بشكل رئيسي على الصدر والأطراف العلوية. من الناحية النسيجية، يتم تعريفه من خلال الشعيرات الدموية الجلدية المرتبة داخل الفراغات الوعائية، والتي تشبه إلى حد كبير الكبيبة الكلوية. يُعد وجود الورم الوعائي الكبيبي دليلاً تشخيصيًا للاضطرابات الجهازية مثل اعتلال الأعصاب المتعدد، وتضخم الأعضاء، واعتلال الغدد الصماء، ومتلازمة شذوذ الجلد البروتيني(POEMS) M ، ومرض كاسلمان، والتي تشترك جميعها في ملف السيتوكين المؤيد للالتهابات. نبلغ عن حالة رجل يبلغ من العمر 64 عامًا حضر مصابًا بحطاطات وعقيدات حمراء متعددة غير مؤلمة، تتقدم ببطء، على مدى أكثر من عام، وتشمل الوجه والجذع والأطراف العلوية. كشفت الخزعة الاستئصالية عن سمات نسيجية مرضية مميزة تتوافق مع الورم الوعائي الكبيبي. أظهرت التقييمات السريرية والمخبرية الشاملة عدم وجود أي تأثر جهازي، مما يشير إلى وجود عرض جلدي معزول. على الرغم من أن الورم الوعائي الكبيبي يرتبط ارتباطًا وثيقًا بمتلازمة POEMS وأمراض جهازية أخرى، إلا أنه قد يحدث أحيانًا دون أعراض جهازية.

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