# An unusual association of recurrent pyogenic granuloma on nevus flammeus in a patient with Von Recklinghausen's disease

*Hend M. Alotaibi,* MD.


## ABSTRACT

الورم الحبيبي المتقيح هو أحد الأورام الحميدة الشائعة الحدوث بالجلد والأغشية المخاطية. تقارير طبية قليلة ربطت بين هذه الأورام ووحمة الصباغ الخمري ( Port wine stain )، ولكن لا توجد دراسات وصفية طبية تشير بوضوح إلى ارتباطها بداء الأورام الليفية العصبية من النوع الأول ( Neurofiromatosis type 1 ). نستعرض في هذا التقرير حالة مريض سعودي يبلغ من العمر ٢٩ عاما ومصاب بداء الأورام الليفية العصبية النوع الأول، ويشتكي من ورم حبيبي متقيح متكرر الحدوث فوق وحمة الصباغ الخمري على رقبته. تمت معالجته بجراحة جلدية سطحية مع كي كهربائي وخلال خمس سنوات من المتابعة لم يعاود الورم الظهور.

Pyogenic granuloma is a common benign vascular lesion of the skin and mucosa. There are a few reports on the rare association between it and port wine stain, but there is no clear description of an association with neurofibromatosis type 1 in the literature. This report presents a 29-year-old Saudi male with Von Recklinghausen's disease with recurrent pyogenic granuloma on the nevus flammeus over his neck. He was treated with shave excision and electrocautery with clearance and no recurrence of pyogenic granuloma for the last 5 years follow-up.


*Saudi Med J 2009; Vol. 30 (6): 844-846*

*From the Department of Dermatology, College of Medicine, King Saud University, Riyadh, Kingdom of Saudi Arabia.*

*Received 21st February 2009. Accepted 29th April 2009.*

*Address correspondence and reprint request to: Dr. Hend M. Alotaibi, Acting Consultant Dermatologist, Department of Dermatology, King Saud University, PO Box 62690, Riyadh 11595, Kingdom of Saudi Arabia. Tel. +966 (1) 4691426. Fax. +966 (1) 4691432. E-mail: dr_halotaibi@yahoo.com*

Pyogenic granuloma (PG) is a benign, acquired, proliferative vascular lesion of the skin and mucous membrane whose exact cause is unknown. The lesion usually occurs as a solitary glistening red papule or nodule that is prone to bleeding and ulceration. It typically evolves rapidly over a few weeks, most often on the head, neck, extremities, and upper trunk.[1] Therapeutic varieties include curettage, excision, electrocautery, sclerotherapy, radiotherapy, and the use of lasers.[1-5] Neurofibromatosis (NF) type 1 or Von Recklinghausen's disease is a multisystem genetic disorder that is commonly associated with cutaneous, neurologic, and orthopedic manifestations Clinical diagnosis requires the presence of at least 2 of 7 criteria to confirm the presence of NF type 1.[6] This report present a case of an adult with the defined criteria for NF type 1, with a congenital vascular lesion, nevus flammeus, and an acquired vascular one, recurrent PG, an unusual association not clearly described in literature.

**Case Report.** A 29-year-old Saudi male, presented to the dermatology clinic at King Khalid University Hospital in Riyadh with a recurrent tumor like skin lesion over the left side of his neck at the site of a birth mark (the nevus flammeus). This lesion was recurrent more than 6 times over the previous 2 years, with a history of bleeding occasionally. There was no history of preceding trauma. There was a history of previous surgical treatments in other hospitals. The nevus flammeus presented at birth, and become thicker and darker in color with time. There was no family history of similar red skin lesions, but his 2 daughters were diagnosed with NF type 1 and were being followed by pediatric physicians. On examination, the skin showed a 6x8 cm well-circumscribed red to purple plaque over the left side of face and neck involving the ear (the nevus flammeus). A solitary 1x3 cm red polypoid nodule was also noted over the preceding nevus (**Figure 1**). Examination of the trunk and extremities revealed numerous well-defined regular oval uniformly light to dark brown macules and patches (*café-au-lait macules*) varying in size from 2 mm to 4 cm, over most of the body area, except the palms and soles, with multiple axillary freckles (**Figure 2**). There were a few

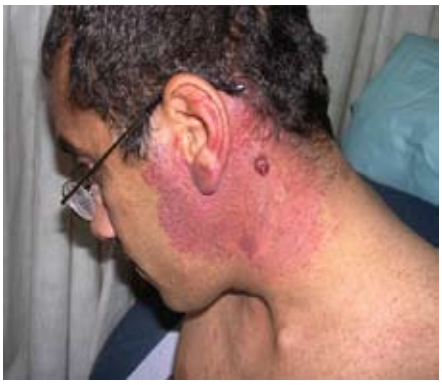

**Figure 1 -** Pyogenic granuloma over a port wine stain before treatment.

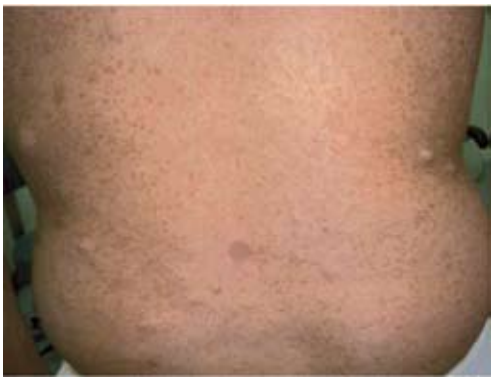

**Figure 3 -** Neurofibromas over the back.

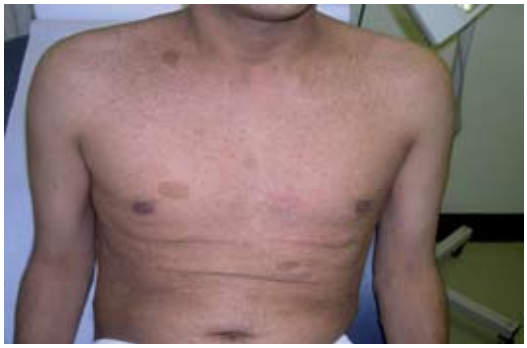

**Figure 2 -** Café-au-lait macules over the chest.

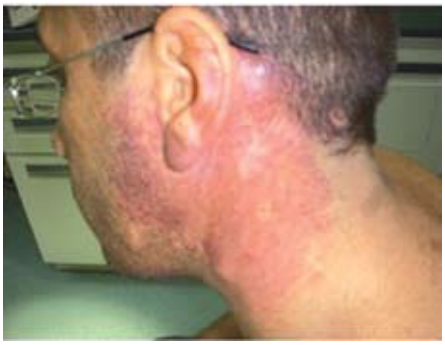

**Figure 4 -** Pyogenic granuloma cleared after treatment.

skin colored to brown soft nodules (neurofibromas) on the trunk (**Figure 3**). Ophthalmologic examination by a specialist, showed the presence of multiple brownish-grey nodules, greater than 2 mm, irregularly distributed all over the surface of both irises, compatible with Lisch nodules. Blood pressure, neurologic, and orthopedic examination detected no abnormalities. An MRI of the brain was normal. He gave consent for the procedures and photographs, and then he was started on cryotherapy treatment for one session, one month later he was treated with shave excision and electrocautery with a good response as clearance and a small hypopigmented macule with no recurrence for the last 5 years follow up (**Figure 4**). He was also referred to a specialist for the vascular laser of the nevus flammeus, and showed fair improvement after 6 sessions.

**Discussion.** There are few reports on the rare association between PG and port wine stain (PWS), which are either de novo or following trauma, such as laser therapy, and cryotherapy.[7-10] This association could be explained by the PG, which is considered a reactive lesion that tends to develop at highly vascularized areas such as the fingers, hands, face, tongue, and PWS, as these sites are associated with microscopic arteriovenous anastomoses.[7,10] Askar et al[10] described a PG in PWS on the posterior cervical area, and suggested the role of continuous trauma by the collar of the patient's shirt, since that area is not as highly vascular as acral sites. The patient discussed earlier had many laser sessions for his nevus flammeus lesion, which may explain the recurrence of PG so many times.

The association between recurrent PG on the nevus flammeus and Von Recklinghausen's disease or NF type 1 has not been described clearly in the medical literature before. However, Ruiz Villaverde et al,[11] described a 10-year-old male with giant nevus flammeus, nevus spilus, asymmetry in the development of both lower limbs, and Lisch nodules. They believe that this case can be classified as phakomatosis pigmentovascularis type IIb, associated with Lisch nodules as part of segmental NF or NF type V, which is a controversial clinical form of NF. Several authors, including Riccardi[13] believe that this term has occasionally been applied to giant nevus spilus without meeting the criteria for NF.

In conclusion, dermatologists, as well as neurologists should have knowledge of and be aware of this rare association.

## References


1. Mirshams M, Daneshpazhooh M, Mirshekari A, Taheri A, Mansoori P, Hekmat S. Cryotherapy in the treatment of pyogenic granuloma. ***J Eur Acad Dermatol Venereol*** 2006; 20: 788-790.
2. González S, Vibhagool C, Falo LD Jr, Momtaz KT, Grevelink J, González E. Treatment of pyogenic granulomas with the 585 nm pulsed dye laser. ***J Am Acad Dermatol*** 1996; 35 (3 Pt 1): 428-431.
3. Quitkin HM, Rosenwasser MP, Strauch RJ. The efficacy of silver nitrate cauterization for pyogenic granuloma of the hand. ***J Hand Surg*** 2003; 28: 435-438.
4. Raulin C, Greve B, Hammes S. The combined continuous wave/pulsed carbon dioxide laser for treatment of pyogenic granuloma. ***Arch Dermatol*** 2002; 138: 33-37.
5. Matsumoto K, Nakanishi H, Seike T, Koizumi Y, Mihara K, Kubo Y. Treatment of pyogenic granuloma with a sclerosing agent. ***Dermatol Surg*** 2001; 27: 521-523.
6. Pivnick EK, Riccardi VM. Neurofibromatoses. In: Freedberg IM, Eisen AZ, Wolf K, Austen KF, Goldsmin LA, Katz SI, editors. Fitzpatrick's dermatology in general medicine. 6th ed. New York (NY): McGraw-Hill; 2003. p. 1825-1833.
7. Aghaei S. Pyogenic granuloma arising in port-wine stain after cryotherapy. ***Dermatol Online J*** 2003; 9: 16.
8. Castanedo-Cazares JP, Lepe V, Moncada B. Pyogenic granuloma within port-wine stains. ***Eur J Dermatol*** 2002; 12: 616.
9. Sheehan DJ, Lesher JL Jr. Pyogenic granuloma arising within a port-wine stains. ***Cutis*** 2004; 73: 175-180.
10. Askar I, Kilinc N, Yucetas A. Pyogenic granuloma appearing on port-wine stain: a case report. ***Acta Chir Plast*** 2003; 45: 52-54.
11. Ruiz Villaverde R, Viera Ramirez A, Linares Solano J, Naranjo Sintes R, Gutierrez Salmerón MT. Phakomatosis pigmentovascularis and Lisch nodules. Relationship between Von Recklinghausen and phakomatosis pigmentovascularis? ***Eur J Dermatol*** 2003; 17: 53-55.


### Related topics

Kayaoglu CR, Sengul G, Aydin IH. Multiple schwannomas of cauda equina in the absence of von Recklinghausen's disease. ***Saudi Med J*** 2007; 28: 1907-1909.

Tillawi IS. Amyloid-depositing plasmacytoma of cervical spine masquerades as a granulomatous inflammatory reaction. ***Saudi Med J*** 2007; 28: 147-149.

Al-Maghrabi JA, Sawan AS, Kanaan HD. Hodgkin's lymphoma with exuberant granulomatous reaction. ***Saudi Med J*** 2006; 27: 1905-1907.

Safioleas MC, Stamatakos MC, Diab AI, Michael OP, Safioleas KM, Liapis GC. Granuloma of the appendiceal stump. An unusual cause of right hemicolectomy. ***Saudi Med J*** 2006; 27: 1230-1231.

Wani MM, Reshi AR, Banday KA, Najar MS. von Recklinghausen's neurofibromatosis associated with membranous glomerulonephritis. ***Saudi Med J*** 2006; 27: 534-535.