Case Reports

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

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ABSTRACT

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.


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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. Therapeutic varieties include curettage, excision, electrocautery, sclerotherapy, radiotherapy, and the use of lasers.

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. This report presents 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 birthmark (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 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
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 al10 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 Riccardi13 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.
Pyogenic granuloma, port wine stain, and nevus flammeus ... Al-Otaibi

References


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