

Glomangioma: Rare Localization of a Rare Skin Tumor

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Abstract

Observations: Diagnosis of some of the skin tumors might be difficult as they have no typical clinical appearance. However, subjective complaints such as the painful character of the lesion could be a guide in the differential diagnosis. The extreme pin point pain is a characteristic of glomus tumor and alongside this extreme pain it can be easily diagnosed. Glomangioma is a highly vascularised variant of glomus tumor and is a rare, slow growing benign tumor of dermis or subcutaneous tissue. The most common site of occurrence is in the hands, especially in the distal phalanx and extra digital localization of glomus tumor is rare. Hence, it cannot be easily diagnosed in atypical localizations, thus sometimes causing misdiagnosis. Differential diagnosis is critical for proper interventions in atypical localization. Here, we report a 64 years old male patient suffering from localized pain in the leg for 22 years without diagnosis. Even ultrasonographic evaluation was very painful, it revealed a subcutaneous hypo echoic mass with increased vascularization which was also detected at Doppler US. The patient had great benefit from surgical excision, with a histopathologically confirmed diagnosis of glomangioma. In cases of unexplained point hyperesthesia and pain, atypically located glomus tumors should be encountered in differential diagnosis. Ultrasonography should be the method of first choice due to its high diagnostic value, as well as being economic and easy.

Introduction

Diagnosis of some of the skin tumors might be difficult as they have no typical clinical appearance. However, the painful character of the lesion could be a guide in the differential diagnosis. Glomus tumors should be considered in addition to the differential diagnosis with leiomyoma, eccrine spiradenoma, neuroma, dermatofibroma, angioliipoma, neurilemmoma, endometrioma and granular cell tumor [1]. Glomangioma or glomus tumor is a rare, slow growing benign tumor of dermis or subcutaneous tissue. It originates from en-

domyoarterial corpuscles in the glomus body [2]. The most common site of occurrence is the hands, especially the distal phalanx. Characteristic of this tumour, alongside extreme pain it can be easily diagnosed. Although extradigital localization of glomus tumor is rare, in atypical localizations it cannot be easily diagnosed thus sometimes causing misdiagnosis [3]. Differential diagnosis is critical for proper interventions in atypical localization. Here, we report a 64 years old male patient suffering from pain in the leg for 22 years without diagnosis.

Case Report

A sixty-four year-old male patient was admitted to neurosurgery department with right leg pain for 22 years. This serious pain was localized in the outer part of his left leg and was aggravated with touch. He was seen by different doctors for this pain, but did not receive any diagnosis. Since 2 years, the leg pain increased and became resistant to analgesics and intolerant to his clothes. His motor and sensory examination was found to be normal. His Straight Leg Raise (SLR) was negative. There was "point hyperesthesia" in a localized area. His physical examination was not compatible with the pathology in his spine. Dermatology consultation was required. The dermatological examination revealed normal appearance of skin over the allocated area (**Figure 1**). Palpitation was extremely painful and a subcutaneous mass was suspected. An ultrasonography (US) revealed a subcutaneous hypoechoic mass measuring 7x7 mm, and increased vascularization of mass was also detected at Doppler US. Surgical excision was performed by regional anesthesia. Macroscopically tumor was observed as a well circumscribed nodular lesion of brownish color. The pain completely disappeared after the operation. Histopathologically, dilated vascular spaces, some of them thick-walled, surrounded by uniform glomus cells were observed. Glomus cells had round nuclei and scant cytoplasm. Mitotic activity or cellular atypia were not present (**Figure 2**). The histopathological diagnosis was compatible with glomangioma. There were no complications during or after the operation.

Discussion

A glomangioma is typically a small, painful and cold-sensitive lesion occurring in the finger tips usually under the nail. It measures



Figure 1. Normal clinical appearance of the painful skin

usually 1 cm or less. The lesion was first described in 1812 as a painful subcutaneous nodule which worsens by changes in temperature and is cured by surgical removal [4]. These tumors arise from the glomus body, which is responsible for thermoregulatory function and is composed of glomus cells, which are closely related to the smooth muscle cell. Nerve fibers are usually present in the glomus tumor but do not correlate with painful or nonpainful symptomatology [5]. The solitary form is more common than the multiple type, representing 90% of all cases. A solitary glomus tumour is a pink or purple nodule with a classic triad of pain, cold sensitivity and point tenderness.

The glomangiomas are rare tumors. They are localized predominantly in the hands, particularly the fingers [6]. Extra digital localized glomus tumors are very rarely seen. The unusual locations have also been reported such as in the ankle, foot, knee, thigh hip, stomach, tongue, lung, sella, trachea, and vagina [7, 8, 9, 10, 11]. Glomangioma accounts for 1–2% of all soft tissue tumors [5]. Clinically, differential diagnosis of painful tumors of the skin such as eccrine spiradenoma and neuroma should be estimated. Histopathological examination has much value and regarded as gold standart in such lesions. Additionally, in histopathological examination, differential diagnosis should include glomangiosarcoma. Glomangiosarcomas are reported to have a size greater than 2 cm. In our case the maximum diameter was 0,7 cm. They are usually located subfacially or at visceral tissues. They contain mitotic figures, some of them atypical showing nuclear atypia. In our case, lesion was located

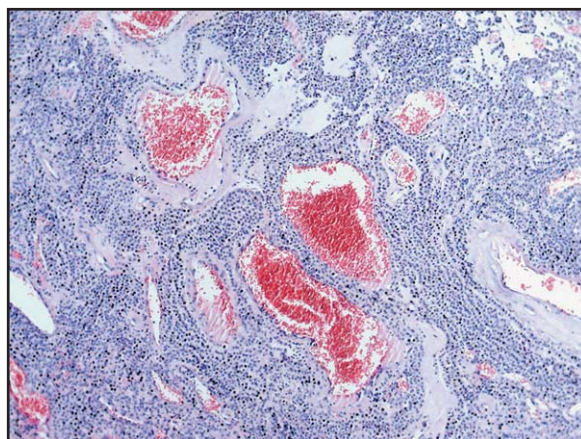


Figure 2. Uniform glomus cells surrounding dilated vascular spaces (H+E, x 100)

subcutaneously and we could not observe mitotic activity or nuclear atypia [12].

A glomangioma usually manifests as a nonspecific, solid, hypoechoic mass at US. The high-velocity flow in intratumoral shunt vessels causes this lesion to be hypervascular at color Doppler. This is a specific finding for the diagnosis of subungual glomangioma and in our case the tumor also showed the similar US pattern.

Magnetic Resonance Imaging (MRI) could also help in differential diagnosis. MRI features that are considered as diagnostic for glomus tumor include intermediate or low signal intensity on T1-weighted images, marked hyperintensity on T2-weighted images, and strong enhancement after the injection of gadolinium-based contrast material. Although MRI is reported to be definitive in subungual glomangioma, it does not supply specific evidence for extradigital locations [13]. MRI was not performed in our case as the Doppler US was strongly suggestive of a glomus tumor showing specific features and relevant with clinical findings.

This patient had diagnostic difficulties due to his inappropriate admissions to different clinics. Because of the rare location of the tumor, a subcutaneous tumor was not suspected and the patient had been treated as with lumbar nerve entrapment syndromes.

In cases of unexplained point hyperesthesia and pain, ultrasound should be the method of first choice due to its high diagnostic value, as well as being economic and easy.

Patients may have great benefits from surgical treatment and relief of this very discomforting situation. Other treatment options for glomangiomas include sclerotherapy, gamma-knife, radiotherapy, lasers and periodic observation of asymptomatic lesions [14, 15]. Our patient had great benefit from surgical treatment by the relief of leg pain after 22 years.

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