Extradigital Glomus Tumor: A Case Report of Pain on Back Over 10 Years

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ABSTRACT:
Glomus tumors are benign neoplasms of the glomus body, commonly found in the subungual area of digits. Occasionally, these tumors occur outside the digital region, making diagnosis more challenging. We report a case of 62-year-old woman who experienced pain on her back for 10 years. A careful examination revealed a small erythematous papule on the interscapular area. Dermoscopic findings showed homogenous, unstructured purplish area. A punch excision was performed, and histological and immunohistochemistry analysis of the tissue was consistent with a glomus tumor. The patient reported no pain following the procedure. This case report highlights the clinical, dermoscopic, and histological features of extradigital glomus tumors. It also emphasizes the importance of including these tumors in the differential diagnosis of painful nodules and demonstrates the effectiveness of complete surgical excision in achieving excellent outcomes.
Key words: Glomus tumor, Extradigital glomus tumor, Painful cutaneous tumor

Introduction

Glomus tumors are uncommon benign neoplasms that arise from the thermoregulatory glomus body. They typically present as painful subcutaneous nodules in the subungual area of digits, which is where these tumors commonly found. However, in about 34% of cases, these tumors occur in extradigital locations such as the trunk, extremities, and visceral organs, often leading to diagnostic delays due to their rarity and atypical presentation[1,2]. This case report aims to highlight the clinical, dermoscopic findings, and histological features of extradigital glomus tumors. Understanding the unique features of these tumors is crucial for preventing misdiagnosis and improving patient care.

Case

A 62-year-old Thai female visited to the outpatient clinic due to a complaint of pain on her back lasting for about ten years. She noted that the pain began after minor trauma to her back and was usually aggravated by digital pressure and heavy lifting. She had underlying medical conditions of type 2 diabetes mellitus and dyslipidemia, for which she was treated with 850 mg of metformin daily, 20 mg of rosuvastatin daily, and 10 mg of ezetimibe daily. Her family history was unremarkable. Physical examination revealed a tiny erythematous papule with tenderness on the interscapular area (Figure 1A). Dermoscopy revealed a homogenous, unstructured purplish area (Figure 1B). A punch excision was performed, and histological examination of the lesion revealed an intradermal, non-encapsulated and well-circumscribed nodule composed of uniform round cells with central nuclei and heterogeneous chromatin (Figure 2A). Small blood vessels were observed within the tumor.

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No cellular atypia was noted. Immunohistochemical analysis revealed that these neoplastic cells stained positive for smooth muscle actin (Figure 2B). CD31 and CD34 were positive for blood vessels, and S100 was positive for dendritic cells within the tumor (Figure 2 C,D). Pan-cytokeratin was not expressed. These findings were consistent with a glomus tumor. After punch excision, there were no complications, and the patient reported no pain.

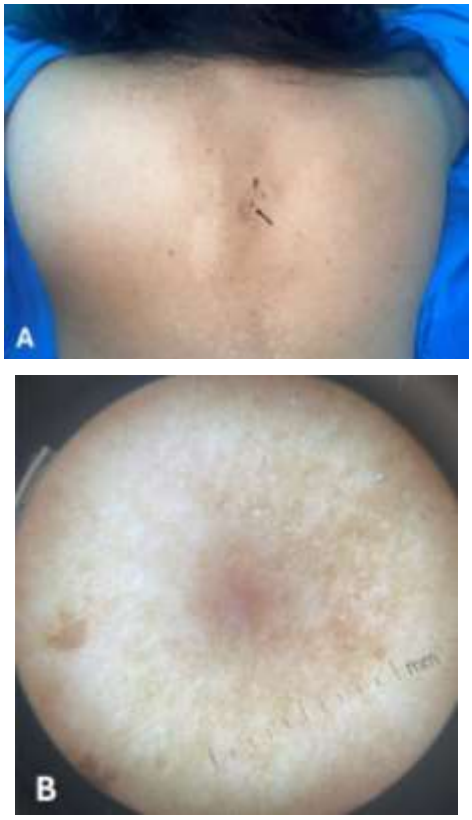

Figure 1 A tiny erythematous papules on interscapular area, arrow (A). Polarized, non-contrast dermoscopy showed a homogeneous, unstructured purplish area (B)

Discussion

The glomus body is the specialized contractile neuromyoarterial structure involved in the regulation of skin temperature, particularly found in digits. Glomus tumors arise from glomus bodies, with the majority developing in the digital region, especially in the subungual area. In addition, they can occur in the trunk, visceral organs, and extremities. Glomus tumors represent 2% of all soft tissue tumors, while extradigital glomus tumors comprising approximately 34% of all glomus tumor cases. This condition typically affects individuals aged 30 to 50, although it can manifest at any age. There is a higher incidence of glomus tumors in females, whereas extradigital variants are more commonly found in men and tend to occur at an older age[1].

Extradigital glomus tumors are present with a painful nodule that may be purplish or bluish; they are often accompanied by pain that can be aggravated by pressure, but they are not usually aggravated by cold temperatures, unlike the glomus tumor in the digital area. The typical size range of extradigital glomus tumors is from 0.1 cm to 9.5 cm, with a mean size of approximately 1 cm.[1] Previous research indicates that TRPM8 and TRPA1 may play a role in the pathogenesis of pain and cold sensitivity in glomus tumors[3]. However, additional research is necessary to evaluate the expression of these channels in extradigital groups. According to available data, there may be an association between the glomus tumor and regional tissue irritation or trauma[1]. A series of glomus tumors cases showed that the diagnosis of extradigital glomus tumors is often delayed, with delays ranging from 5 to 25 years[4]. Dermoscopy plays a crucial role in enhancing diagnostic accuracy. The dermoscopic features of glomus tumors and extradigital glomus tumors may show a similar appearance but extradigital glomus tumors can also present with peripheral telangiectasias. In this case, the dermoscopic findings revealed a homogenous, unstructured purplish area. In a study involving 39 cases of extradigital glomus tumors reported dermoscopic findings similar to this case, which revealed linear unfocused vessels and a

structureless purplish-white to reddish-white homogeneous area. This pattern is highly specific (95.7%) for extradigital glomus tumors. Dermoscopy of an extradigital glomus tumor can be differentiated from other vascular tumors, such as hemangiomas and pyogenic granulomas by the absence of lacunae. Furthermore, they can also be distinguished from malignant skin tumors, as extradigital glomus tumors do not exhibit any signs of ulceration[5].

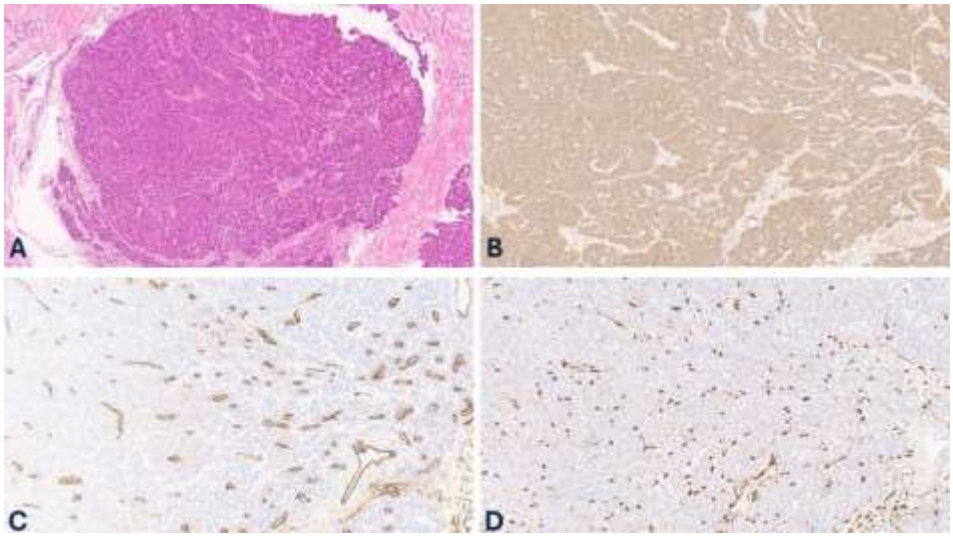

Figure 2 Histological examination revealed intradermal nodules of uniform round cells with central nuclei and heterogeneous chromatin, Hematoxylin and Eosin X10 (A). Immunohistochemical analysis showed tumor cells stained positive for smooth muscle actin, X20 (B). CD34 was positive for blood vessels (C), and S100 was positive for dendritic cells in tumor (D), X20

Imaging plays a crucial role in facilitating accurate diagnosis and differentiating extradigital glomus tumors from similar conditions. On ultrasound imaging, extradigital glomus tumors typically appear as circumscribed, hypoechoic oval nodules located in the subcutaneous layer. These lesions may exhibit internal vascularization, which can be assessed using Doppler ultrasound to reveal both venous and arterial flow. The "vascular stalk sign" may also be observed in some cases, indicating a connection to surrounding vessels. While ultrasound findings are not specific, they are essential for early diagnosis and provide valuable information to guide surgical excision[6]. Magnetic resonance imaging (MRI) is considered the gold standard, offering approximately 90% sensitivity and 50% specificity for glomus tumor, and provides essential information regarding tumor characteristics, size, and location[7]. The typical finding in MRI is a well-defined lesion that appears brighter on T2-weighted images and shows increased enhancement following gadolinium contrast administration. MRI can help distinguish glomus tumors from several other conditions by highlighting specific imaging characteristics. For instance, hemangiomas typically appear more heterogeneous with less defined margins compared to glomus tumors. Angiomyomas exhibit different signal characteristics on MRI, while arteriovenous malformations present as ill-defined lesions with characteristic shunting, and venous malformations are usually poorly demarcated and demonstrate only venous flow[6].

However, in this case, an MRI was not performed because the lesion was small, and the clinical clues, particularly the presence of pain aggravating by digital pressure, are consistent with an extradigital glomus tumor. In addition, given the very small size of the lesion, there was a possibility that the MRI could yield a false negative result.

Regarding the glomus tumors often present with painful nodules, dermatofibroma, leiomyoma, neurofibroma, and eccrine spiradenoma should be considered as differential diagnosis, and a biopsy is required to rule out these tumors[1,6]. The histology of glomus tumors is characterized by well-circumscribed, non-encapsulated nodules. These nodules consist of glomus cells, which are round with central nuclei and heterogeneous chromatin[6,8]. Immunohistochemical staining reveals positive results for smooth muscle actin, vimentin, type IV collagen, and Bcl-2[9].

Surgical excision remains the most effective treatment for extradigital glomus tumors. It is essential to perform a complete resection to prevent any chance of recurrence. Due to the small size of lesion in this case, punch excision was sufficient to completely remove the tumor. At the follow-up visit, the patient reported no pain and no residual tumor was observed. The prognosis for patients with extradigital glomus tumors is generally excellent following complete surgical excision. Recurrence rates are low, and malignant transformation is rare[1,2,10].

In conclusion, this case report focuses on the diagnosis and treatment of an extradigital glomus tumor, which manifested as a painful nodule on the back for ten years. The tumor was identified through a detailed clinical examination, and dermoscopic finding along with histological analysis confirmed the diagnosis of the extradigital glomus tumor. Surgical excision resulted in complete removal of the tumor, leading to the resolve of the pain. This case demonstrates the importance of including extradigital glomus tumors in differential diagnoses of painful nodules and emphasizes the effectiveness of complete surgical excision in achieving excellent outcomes.

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