Int J Clin Exp Med 2014;7(1):323-326
www.ijcem.com /ISSN:1940-5901/IJCEM1311007

Case Report

Soft tissue perineurioma with peripheral lymphoid cuff of the tongue: a case report and literature review

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Received November 9, 2013; Accepted December 11, 2013; Epub January 15, 2014; Published January 30, 2014

Abstract: Perineurioma are rare tumors, derived from nerve sheath perineurial cells. Soft tissue perineurioma are extraordinarily rare in the tongue, with only one previous report in a child to our knowledge. Herein, we describe the first case of an adult patient who had a soft tissue perineurioma localized to the tongue, with emphasis on the clinic pathologic and immunohistochemical features of this tumor, and review the previously reported soft tissue cases. Besides, we first describe the histologic feature of peripheral lymphoid cuff in perineurioma.

Keywords: Soft tissue perineurioma, tongue, nerve sheath neoplasms, oral cavity

Introduction

Perineurioma are rare benign peripheral nerve sheath tumors composed of perineurial cells [1]. Two distinct subtypes are recognized and include intraneural perineurioma and soft tissue perineurioma. Soft tissue perineurioma are generally not associated with an identifiable nerve and found mainly in the subcutis and dermis [2]; furthermore, some rare locations have been reported [3]. The aim of this study is to further investigate the clinicopathologic characteristics of this rare tumor.

Case report

A 47-year-old woman presented in April 2011 with seven years' history of a painless lump in the tongue. She was previously healthy without signs of neurofibromatosis, and denied trauma, or any history of tongue tumors. Grossly, the tumor was unassociated with the nerve and measured 1.5 cm in size and weighted 7.5 g. On sectioning, it was well-circumscribed, solid, pale pink-white and rubbery. Surgery biopsies were fixed in 10% neutral buffered formalin then subjected to routine processing and paraffin embedding. Sections were stained with hematoxylin and eosin. Histologically, the tumor located in the muscle layer of the tongue and surrounded by peripheral lymphoid cuff with germinal centers, which was often presented in schwannoma (**Figure 1A**). It was unencapsulated and well-circumscribed in most areas, but in some areas, it seemed to infiltrate the muscle layer focally. The tumor was composed predominantly of uniform bland spindle cells, which formed wavy fascicles in some areas, with ovoid nuclei and delicate eosinophilic to vacuolated cytoplasm. The stroma was myxoid in some areas. No significant necrosis, nuclear pleomorphism or mitotic activity was noted. To obtain an exact pathological diagnosis, immunohistochemical staining was performed to detect expressions of epithelial membrane antigen (EMA), collagen IV, CD34, CD117, S-100 protein (S-100) and alpha-smooth muscle actin (SMA). Immunohistochemical stains were performed on paraffin embedded tissues, sections of 3 μm thickness from the block were stained using an automated stainer (Leica Instruments, Nussloch, Germany) along with appropriate positive and negative controls. Immunohistochemical staining showed EMA was diffuse positive in the cytoplasm of tumor cells (**Figure 1B**), collagen IV was positive in the cell membrane (**Figure 1C**); while CD34, S-100, SMA and CD117 were negative in tumor cells (**Figure 1D**). There was no evidence of recurrence 26 months after surgical resection. Informed consent was obtained from the patient.

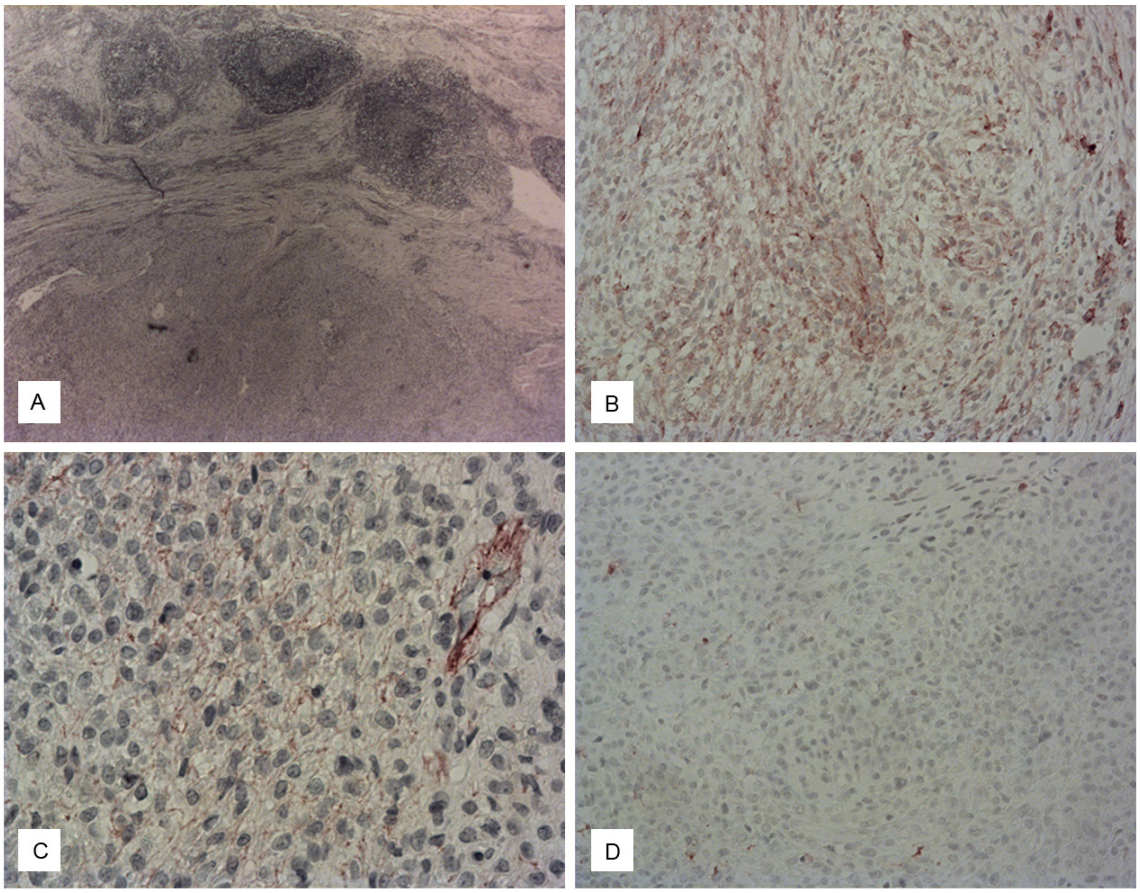

Figure 1. The imaging and histopathological features of the soft tissue perineurioma. (A) The tumor is surrounded by peripheral lymphoid cuff with germinal centers (hematoxylin-eosin staining; original magnifications: A, 25 ×;). (B-D) The tumor cells were diffusely positive for EMA (B), membranous positive for collagen IV (C), and focal positive for S-100 (D) (immunohistochemistry; original magnifications, D, 200 ×; C, 400 ×).

Discussion

Perineurioma are uncommon benign peripheral nerve sheath tumors that include two subtypes: intraneural perineurioma and soft tissue perineurioma [1]. Whereas intraneural perineurioma are found around major peripheral nerves and occurred predominantly in the nerve of upper limb, soft tissue perineurioma are generally not associated with an identifiable nerve and found mainly in the subcutis and dermis, most commonly in the limbs and trunk [2]. Furthermore, soft tissue perineurioma have been reported in the stomach [4], intestinal tract [5], kidney [6], lip [7], maxillary sinus [7], mandible [8] and cardiac ventricle [9] et al. They are more often found in the superficial soft tissues (70%) than in deep tissues [2]. Here, we present a case of soft tissue perineurioma of the tongue.

Soft tissue perineurioma are seen in adults of all ages, with a female predilection. They are usually present, as in this case, with local painless, slowly growing and solid lumps measured 0.3-20 cm (mean 4.1 cm) [2], some may have compression symptoms, such as abdominal pain [10] or respiratory infection [11]. They are typically well circumscribed, often having a fibrous pseudocapsule; however, they may have infiltrative margins focally, such as several cases described before and this report [3, 12]. The most common morphological finding from soft tissue perineurioma is the presence of uniform bland spindle or epithelioid neoplastic cells; they are arranged in vague fascicular, storiform or whorl-forming patterns, with ovoid to elongated nuclei and inconspicuous nucleoli. Stroma may be markedly myxoid, collagenous or hyalinized, but mitotic figures are rare [2]. On the frozen section and HE stained slides of this

case, the features of uniform spindle cells with vacuolated cytoplasm, wavy or fascicular patterns, proliferative lymphoid tissue around tumor, as well as myxoid stroma all led us to make a diagnosis of benign neurogenic tumor. Then, immunohistochemistry played a significant role in accurate recognition of this tumor; the result from our case consisted of findings that typically occur in soft tissue perineurioma, i.e. strong expression of EMA with no expression of S-100 protein. Peripheral lymphoid cuff with germinal centers is a typical histologic feature that assists in recognition of schwannoma [13], but it has not been described in perineurioma hitherto, we found this feature can also assists in soft tissue perineurioma, so we think it maybe a feature of all peripheral nerve sheath tumors. Furthermore, recently demonstrated findings that perineurioma express claudin-1, a tight junction-associated protein, and Glut-1, an erythrocyte-type glucose transporter protein; membranous staining for laminin and collagen IV may also reinforce evidence for diagnosing perineurioma [3]. The most important differential diagnoses are with neurofibroma and schwannoma. Immunohistochemical analysis can easily separate perineurioma from them, since the former are positive for EMA protein and others are positive for S-100. Granular cell tumor is another tumor most commonly occur in the tongue, but it has characteristic granular cytoplasm and expresses S-100. Moreover, we performed SMA and CD34 staining to exclude smooth muscle tumor and solitary fibrous tumors. It has been suggested that the diagnosis of perineurioma also requires ultrastructural confirmation; ultrastructural features of perineurial cells include long thin cytoplasmic processes with numerous pinocytotic vesicles, abundant collagenous stroma, in-continuous basal lamina and rudimentary intercellular junctions [14, 15].

Soft tissue perineurioma probably have a benign clinical course, and surgical resection with clean margins is considered curative [16]. Atypical cellular features such as scattered pleomorphic or multinucleated cells, hypercellular areas, or focal infiltrative margins are not uncommon in soft tissue perineurioma, and many investigators believe these features to be a degenerative change and therefore to have no clinical or prognostic significance [2, 17]. The criteria used for the diagnosis of malignant perineurioma (also known as malignant peripheral nerve sheath tumors (MPNST)) are infiltrative growth pattern, significant cytologic atypia, presence of necrosis, and high mitotic activity [17]. None of these features were identified in this case.

While perineurioma are extraordinarily rare, the case presented in this paper represents a soft tissue perineurioma of the tongue. Histopathological analysis with HE stained slides, immunohistochemical stains and ultrastructural finds are important in accurate diagnosis of this tumor.

Disclosure of conflict of interest

None.

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