Plexiform Schwannoma: A Case Report

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Abstract
Plexiform schwannoma is a rare benign neoplasm of the neural sheath characterized by a multinodular plexiform growth pattern. About 5% of schwannomas grow in a plexiform or multinodular growth patterns. Extracranially, 25% of them are located in the head and neck region while only 1% is present in intra oral origin. The intra oral lesions show predilection in the tongue, followed by palate, buccal mucosa, lip and gingivae. Microscopic examination is necessary to confirm the diagnosis. Characteristic histological features are spindle shaped cells with wavy nuclei showing abundant eosinophilic cytoplasm that arrange into multiple lobules. Here a case presented as a soft circumscribed lump on upper lip in a 20 year old female patient clinically similar to fibroepithelial polyp is described. This case report highlights the important of histopathological analysis of each and every nodule present in the human body.

Key words: Plexiform schwannoma, upper lip

Introduction
Schwannoma is a benign tumour arising from sheath of myelinated nerve fibers that may occur in any part of the body. Plexiform schwannoma is a rare variant of schwannoma that account for only 5% of all schwannomas, and typically shows a plexiform or multinodular growth pattern. It was first described by Harkin and Reed in 1978, and a very few cases have been reported since. Plexiform schwannomas usually develop in the dermis or subcutaneous tissue and it is uncommon for schwannoma to develop in deep seated nerves. It is a benign sporadic and is characterized by spindle shaped cells with abundant cytoplasm, arranged into multiple nodular pattern. This rare tumour is worthy of recognition because it can be misdiagnosed as plexiform neurofibroma. Differentiation from plexiform neurofibroma is important because the latter is pathognomonic of Von Recklinghausen disease and carries a significant risk of malignant transformation. Plexiform schwannoma is in contrast, not necessarily associated with Von Recklinghausen disease (Neurofibromatosis type 1) and malignant transformation has never been observed.

Case Report
A 20 year old female presented with a one year history of a soft circumscribed painless lump on left side upper labial mucosa. The medical
history was non contributory. On intra oral examination, there was a smooth, non fluctuant round lump on upper lip. It was covered with normal appearing mucosa and measuring approximately 4 x 4 mm in size (Figure 1). Differential diagnosis included fibro epithelial polyp, neurofibroma, and mucocele. Excisional biopsy was performed under local anesthesia. The sample sent for histopathological investigations. Microscopically there was a mucosal nodule covered by unremarkable parakeratinized stratified squamous epithelium. The corium comprised spindle shaped cells arranged into multiple lobules (Figure 2a, 2b). Although not prominent, Verueae body formation was also observed in the section. Immunohistochemical stains for S-100 protein was strongly positive in most of the spindle cells (Figure 3). Therefore the final diagnosis was Plexiform Schwannoma. The patient is on regular review and there are no signs of recurrence or new lesions even after eight months.

Figure 1. Clinical presentation of the lesion

Figure 2a. Multinodular arrangement of Schwann cells

Figure 2b. higher magnification shows spindle shaped cells with occasional Verueae bodies

Figure 3. Positive expression of S-100 protein by immunohistochemistry
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Discussion
Plexiform schwannoma is a rare variant of Schwann cell tumor. Occurring in either conventional or cellular type, they are characterized either grossly or histologically by a plexiform pattern of intraneural growth often with multinodularity. There are 11 cases of this condition in upper lip reported so far in English medical literature. Embryologically, Schwann cells arise during the fourth week of development from a specialized population of ectomesenchymal cells of neural crest.

The main types of schwannoma are conventional (common, solitary), cellular, plexiform, ancient forms, and melanotic schwannoma. Recently a form of plexiform schwannoma consisting of a proliferation of epithelioid and spindled cells positive for S-100 was described, Melan-A, and HMB-45 but no obvious melanin pigmentation, called plexiform melanocytic schwannoma. Other rare variants are malignant epithelioid schwannoma without neurofibromatosis, and cutaneous pseudoglandular schwannoma containing glandlike structures formed by neoplastic Schwann cells and containing mucinous material.

Typically plexiform schwannomas arise in superficial soft tissues and show a predilection for the head and neck region. Infrequent examples arise in the setting of neurofibromatosis type 2 or schwannomatosis. Solitary plexiform schwannoma is characterized by thin capsules, fascicles and multiple interconnecting nodules with presence of Antoni A and focal or absent Antoni B areas.

Most reports suggest that the majority of tumours are present between the ages of 10 and 40 years and are equally distributed between the two sexes. It is usually found on the trunk, head, neck and upper extremities most often involve the skin and subcutaneous tissue. 25% to 45% of all schwannaomas are located in the head and neck regions and 1% is intra oral.

It is usually asymptomatic, commonly appears as a single, slow growing encapsulated nodule, but sometimes can cause displacement and compression of the surrounding normal nerves tissue. Occasionally there may be pain and paraesthesia (commonly affected VIII acoustic cranial nerve). In the oral cavity, the lesion is usually presented in soft tissue, more commonly the tongue, followed by hard and soft palate, buccal mucosa, lip and gingivae. The lesion may have clinical features similar to other benign lesions like fibroma, lipoma, mucocele, fibroepithelial hyperplasia and benign salivary tumour. Unlike plexiform neurofibroma, which are considered nearly pathognomonic of Neurofibromatosis type I, the association of plexiform schwannoma with Neurofibromatosis type-1 or Neurofibromatosis-2 is considerably weaker. Approximately 50 cases had been report in the literature and only a few cases had been associated with Neurofibromatosis type-1 or Neurofibromatosis type-2.

The differential diagnosis for this describing lesion includes plexiform neurofibroma, palisaded encapsulated neuroma (PEN), and neurotropic melanoma. Plexiform neurofibromas are often far less cellular, have a relatively mucin-rich matrix, and are less S-100 protein immunoreactive. Unlike plexiform schwannoma, PEN contains axons which are demonstrable on both silver impregnation (Bielschowsky and Bodian stain) and neurofilament immunostain. Plexiform schwannoma and neurotropic melanoma both affect skin and subcutaneous tissue. Given the marked difference in their prognoses, it is of great importance to distinguish between the two. Neurotropic melanoma is a rare variant of desmoplastic melanoma which gives strong positivity for s100 protein. Melanin markers such as Melan A, HMB45, LEU7 and NKCI3 are also positive focally which is completely negative in schwannoma.
Solitary plexiform schwannoma is characterized by a thin capsule, fascicles and multiple interconnecting nodules with the presence of Antoni A areas. There may be focal or absent Antoni B areas. Immunohistochemically, diffuse and strong positivity for S-100 a marker for neural tissue (neural crest origin tissue) is diagnostic. Further, laminin and collagen type IV are also positive.

It is important to differentiate plexiform schwannoma from plexiform neurofibromas and malignant peripheral nerve sheath tumours because plexiform schwannoma follows a benign clinical course, with complete surgical excision being curative. The treatment of choice is the complete surgical removal of the lesion. Ultrasonography, CT and MRI may be helpful in planning the treatment depending on the site of the lesion. Recurrences and malignant transformation are rare events.

**Conclusion**

The plexiform schwannoma represents a lesion not often encountered in clinical practice. The submucosal forms of this lesion are usually indistinguishable from other benign neoplasms that are usually seen in the same region. The final diagnosis should be done after histopathological examination with immunohistochemical analysis. This case report highlights the important of histopathological analysis of each and every nodules presented in the human body.

**References**

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