CASE 17

Patient: A 45-year-old man from Rayong

Chief Complaint: Multiple papules on chin for 22 years

Present Illness: The patient presented with an asymptomatic group of slowly enlarging skin-colored papules on his chin since he was 23-year-old.

Past History: No underlying disease. No history of hearing loss.

Family History: None of the family members experienced similar skin lesions.

Physical Examination: Complete neurologic, ophthalmologic and ear, nose, throat examination did not reveal any abnormality.

Dermatological Examination (Figure 17.1-2)**:** Skin lesion showed a group of skin-colored papules, soft to firm consistency which distributed in linear configuration and extended from lower lip to left side of chin.

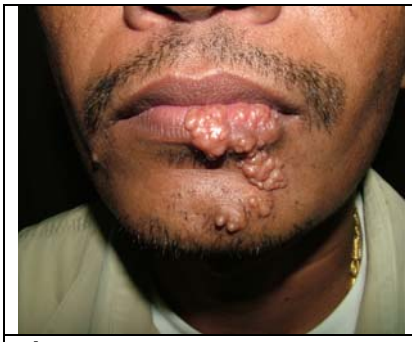

Figure 17.1

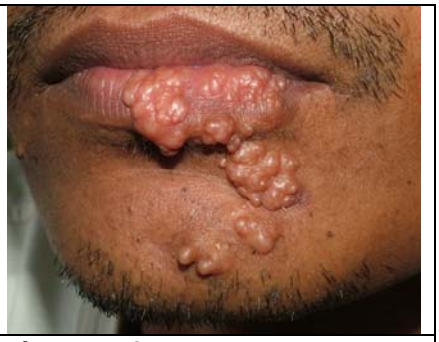

Figure 17.2

Histopathology (S09-17130) (Figure 17.3-4)**:** Multiple plexiform aggregates of oval and spindle cells some arranged in interlacing fascicles and some tendency for palisading nuclei, throughout the dermis

Immunohistochemistry (Figure 17.5): positive S100

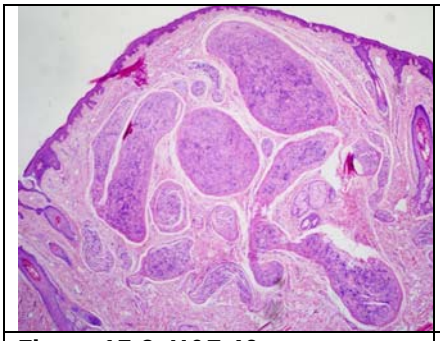

Figure 17.3, H&E 40x

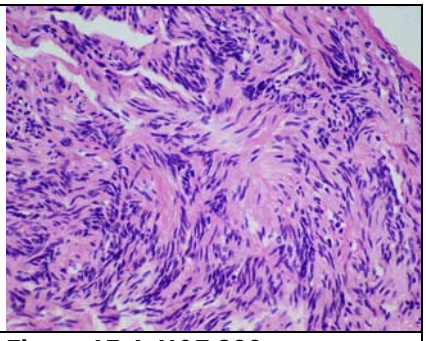

Figure 17.4, H&E 200x

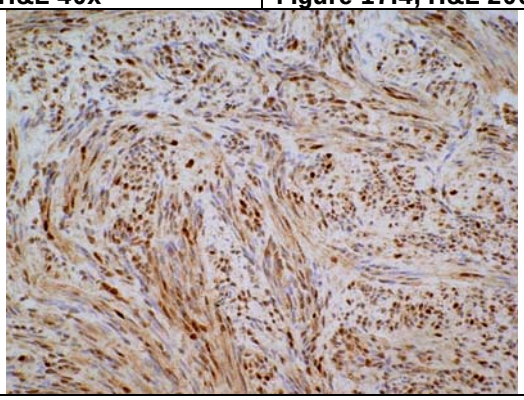

Figure 17.5, S100 200x

Diagnosis: Plexiform schwannoma

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Consultant: Vasanop Vachiramon

Discussion:

Plexiform schawannoma was first described in 1978 by Harkin et al. as an unusual form of benign peripheral nerve sheath tumor. Subsequently, In 1985 Erlandson classified Schwannomas into seven subtypes: classical (Verocay), cranial nerve, cellular, plexiform (multinodular), ancient (degenerated), melanotic, and granular cell schwannoma.[1] Plexiform subtype is a rare distinct variant constituting only 5% of case and tend to occur in adolescents and young adults of either sex.[2]

In the majority of cases, schwannomas present as a slowly growing asymptomatic solitary or multiple nodules, rarely it is painful or tender. In contrast to conventional schwannomas (which most frequently arise on the flexor aspect of the limbs), the trunk, head, and neck region are the most common sites for the plexiform variant.[2,3]Ordinary as well as plexiform schwannoma typically locates in superficial soft tissues (in the dermis and subcutaneous tissue) with only rare presence in non-cutaneous regions having been reported.[4] Of the 98 documented cases, 77 tumors (79%) arise in superficial tissues, 5 (5%) originated in the mucosa, 3 (3%) in visceral organs, and 5 (5%) were of deep soft-tissue origin.[5,6] Particularly uncommon sites involvement compose of the oral cavity, larynx, vulva, testes, breast, deep soft tissue, small intestine and colon.[7,8]

Histologically, this variant is similar to those of a conventional schwannoma other than its plexiform growth pattern.

It is characterized by multiple encapsulated nodules that vary in size and shape and are separated from one another by hyalinized fibrous stroma.[9] Most of the tumor consists of Antony type A while Antoni B areas are sparse,

except in particularly large nodules.[2,3] Immunohistochemically, the tumor cells are strongly positive for S-100 protein.

Plexiform schwannoma should be differentiated from plexiform neurofibroma by the latter is usually hypocellular and primarily composed of a disorganized array of Schwann cells, fibroblasts and axons in prominent myxoid matrix.[7]It is worthy of recognition because plexiform neurofibroma is a pathognomonic feature of neurofibromatosis and has a propensity for malignant potential where as most reported cases of plexiform schwannoma have not been associated with neurofibromatosis as well as malignant transformation.[4,7]Additionally, other differential diagnostic considerations include plexiform malignant peripheral nerve sheath tumor, plexiform granular cell tumor and traumatic neuroma.

Generally, surgical resection is the treatment of choice, although local recurrence can occurs as a result of incomplete resection of tumor.[7]

References

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3. Pinto RGW, Mandreker S, Nadkarni N, et al. Plexiform (multinodular) schwannoma in a child: a rare variant of schwannoma. J Indian Med Assoc 1995; 93(8): 317-8

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5 Tomita K, Mori K, Miyajima Y, Nakashima T. Plexiform schwannoma of the neck extending deeply to the mediastinum. Acta Otolaryngol Suppl 1998; 539: 106–109.

6 Shishiba T, Niimura M, Ohtsuka F, Tsuru N. Multiple cutaneous neurilemmomas as a skin manifestation of neurilemmomatosis. J Am Acad Dermatol 1984; 10: 744–754.
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9. TerasaKi K, Mera Y, Uchimiya H, Katahira Y, Kanzaki T. Plexiform schwannoma. Clin Exp Dermatol 2003; 28: 371–374.