Nerve sheath myxoma of the upper lip: a case report

Üst dudağın sinir kılıfı miksoması: Olgu sunumu

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Nerve sheath myxoma is a benign tumor that rarely occurs in the lip area. In this article, a 28-year-old woman admitted with a complaint of painless swelling in the upper lip is presented. The lesion was excised, and histopathological and immunohistochemical examinations were performed. There are only five reported cases in the available literature. Although very rare, nerve sheath myxoma should be considered in the differential diagnosis of lip mass and widely excised to avoid local recurrence. The patient is under clinical control with no signs of recurrence after 16 months.

Key Words: Lip; neoplasm; nerve sheath myxoma.

Sinir kılıfı miksoması dudak bölgesinde nadiren ortaya çıkan benign bir tümördür. Bu yazıda üst dudakta ağrısız şişlik yakınması ile başvuran 28 yaşında kadın olgu sunuldu. Lezyon eksize edildi ve histopatolojik ve immünohistokimyasal incelemeler yapıldı. Ulaşılabilir literatürde bildirilmiş sadece beş olgu vardır. Sinir kılıfı miksoması dudak kitleleri her ne kadar nadir olsa da ayırıcı tanıda akılda bulundurulmalı ve yerel nükslerden kaçınmak için geniş bir şekilde çıkarılmalıdır. Hasta 16 ay sonra nüks belirtisi olmaksızın klinik kontrol altındadır.

Anahtar Sözcükler: Dudak; tümör; sinir kılıfı miksoması.

Nerve sheath myxoma (NSM) also known as neurothekeoma, is a benign nerve sheath tumor. This tumor, initially described as a benign cutaneous lesion showing neural growing pattern, was initially reported NSM and later reported as neurothekeoma.[1] Nerve sheath myxoma often occurs on the head and neck but rarely in the lip. To our knowledge, only five cases of this entity are documented in the English literature.[2-6] In this article we report a female patient with a painless swelling in the upper lip.

CASE REPORT

A 28-year-old female admitted of our clinic with a 10-year history of painless mass in the upper lip. Her family and medical histories were unremarkable. On physical examination a 1x1 cm solid, mobile, painless mass was noted in the upper lip. The laboratory tests were normal. A surgical exploration was performed under sedation with local anesthesia. The mass was completely excised by intraoral surgical approach. It was dissected from orbicularis oris muscle by blunt dissection. On macroscopic examination the specimen consisted of 0.8x0.8x0.3 cm oval, elastic, solid nodule. Histopathological examination demonstrated multiple myxoid nodules. The nodules were clear intercellular matrix positive for periodic acid-Schiff alcian blue staining. The intercellular myxoid stroma was highly positive with alcian blue (Figure 1).

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Figure 1. *Histopathological examination showing proliferating myxoid nodules (H-E x 40).*

Figure 2. *The immunostaining of the neoplasm showing S100 positivity (H-E x 100).*

Immunhistochemistry studies showed the tumor cells to be diffusely positive for S100 while negative for actin, GFAB (glial fibrillary acidic protein), cd117, desmin and EMA (epithelial membran antigen; Figure 2). The lesion was diagnosed as nerve sheath myxoma.

DISCUSSION

Nerve sheath myxoma is an uncommon tumor that arises from any peripheral nerve. In our case that derived from the infraorbital nerve. They have a wide distribution, but about 14% occur on the head and neck area.[1] The first NSM of the lip was described by Mason et al.[3] To our knowledge, five cases are documented in the literature (Table 1).[2-6] All of the cases localized in the lower lip and oral cavity. Our case localized in the upper lip. This tumor is the first case arising from the infraorbital nerve.

The epidemiological characteristics of NSM of the lip are the following: age range 24 to 71 years female predominance (2:1). Often they are a painless mass in the lip.[2-4] In our case there was painless mass too, so radiologic examination was not arranged.

Table 1. The characteristics of lip tumors including present case

Authors	Age/sex	Size (cm)
Nishioka et al.[2]	52/F	0.7x0.7
Mason et al.[3]	32/F	1x0.7x0.5
Terrier et al.[4]	35/M	5x3
Tiffee et al.[5]	71/F	0.9x0.6
Katsourakis et al.[6]	24/M	2x2x1.5
Present report	28/F	0.8x0.8

Generally, NSM is classified as classical, cellular and mixed types based on amount of myxoid component.[2] The differential diagnosis of NSM is extensive and includes other neural entities, such as schwannoma, neurofibroma and malignant fibrous histiocytoma especially myxoid malignant fibrous histiocytoma.[7]

Nerve sheath myxomas are managed by complete surgical excision, wide excision is recommended.[8,9] Because NSM show high (47%) local recurrences after surgery.[1] We excised the mass by intraoral approach. The patient has followed up for 16 months and there is no recurrence.

This report describes the clinicopathologic findings in case nerve sheath myxoma of the upper lip. It is the first case arising from the infraorbital nerve. Nerve sheath myxoma of the lip is a benign, slow growing mass. Wide excision appears to be curative. Although very rare, NSM should be considered in the differential diagnosis of lip mass and widely excised to avoid local recurrence.

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