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Case Report / Olgu Sunumu

Malignant peripheral nerve sheath tumor of lower lip

Alt dudağın malign periferik sinir kılıfı tümörü

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Malignant peripheral nerve sheath tumors (MPNSTs) are rare, highly aggressive tumors capable of arising de novo or from preexisting benign neurofibromas or schwannomas. The treatment of choice is surgery. Trunk and extremities are the commonest sites involved. Malignant peripheral nerve sheath tumors are rarely encountered in head and neck region. Here, we report a patient with MPNST of lower lip who had neither a family history nor stigmata of neurofibromatosis.

Key Words: Head and neck; lower lip; malignant peripheral nerve sheath tumor.

Malign periferik sinir kılıfı tümörleri (MPSKT) de novo ya da önceden mevcut olan nörofibromlardan veya schwannomlardan ortaya çıkabilen nadir ve yüksek seviyede agresif tümörlerdir. Tercih edilen tedavi cerrahidir. Gövde ve ekstremiteler en sık şekilde tutulan bölgelerdir. Malign periferik sinir kılıfı tümörlerine baş ve boyun bölgesinde nadiren rastlanır. Burada, nörofibromatozise ilişkin bir aile öyküsüne ya da belirtilere sahip olmayan ve alt dudağında MPSKT bulunan bir hasta bildirmekteyiz.

Anahtar Sözcükler: Baş ve boyun; alt dudak; malign periferik sinir kılıfı tümörü.

Malignant peripheral nerve sheath tumors (MPNSTs) are defined as any malignant tumor arising from or differentiating toward cells of the peripheral nerve sheath. They are very rare tumors, with an incidence of 1 per 100,000 population.[1] The World Health Organization (WHO) coined the term MPNST, replacing previous heterogeneous and often confusing terminology such as malignant schwannoma, malignant neurilemmoma and neurofibrosarcoma for tumors of neurogenic origin and similar biological behavior.[2]

Malignant peripheral nerve sheath tumors have a tendency to recur locally and spread hematogenously.[3] Alhough they are biologically aggressive, surgery is the mainstay of treatment.[3] Despite aggressive surgery and adjuvant therapy, the prognosis for patients with MPNST remains poor.[4,5]

Malignant peripheral nerve sheath tumors arise from major or minor peripheral nerve branches or sheaths of peripheral nerve fibers.[6] The trunk and extremities are the commonest sites involved.[3] Malignant peripheral nerve sheath tumors are rarely encountered in the head and neck region. A patient who had been diagnosed and treated for MPNST of the lower lip was presented in this study.

CASE REPORT

A 46-year-old man was admitted to our clinic in June 2007 for six months' progressive swelling

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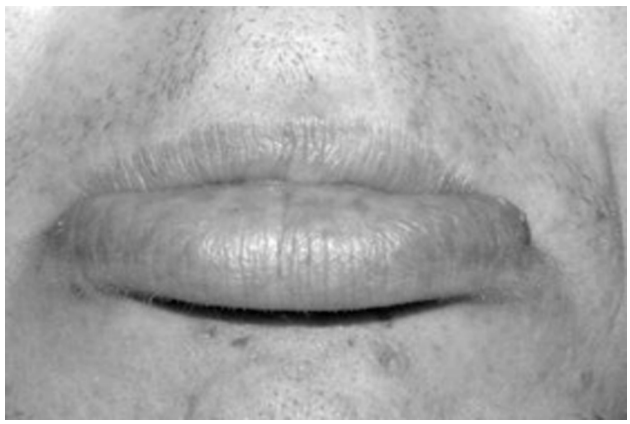

Figure 1. *Swelling of patient's lower lip.*

of his lower lip (figure 1). Otolaryngologic examination revealed a non-tender, mobile protruding mass in the lower lip measuring 3 cm in diameter, covered by normal mucosa. It was firm to elastic but not fixed to the surrounding tissue. There was no paresthesia of surrounding areas. The past medical history was unremarkable. Routine laboratory studies were normal. Resection of the tumor was performed under local anesthesia. The mass was encapsulated with regular borders. It was dissected from surrounding tissues without any difficulty.

Histopathologic examination disclosed a low-grade epithelioid-type malignant peripheral nerve sheath tumor (figure 2a-c). After histological diagnosis, the patient was assessed clinically for the presence of neurofibromatosis type 1 syndrome. After documenting the absence of cafe-au-lait spots, skin-fold freckles, skeletal dysplasia, Lisch nodules, optic glioma, cutaneous neurofibromas and family history, the tumor was evaluated as sporadic. Adjuvant radiotherapy was proposed but the patient refused. No complication occurred related to surgery and the patient was doing well for six months after surgery without any recurrence.

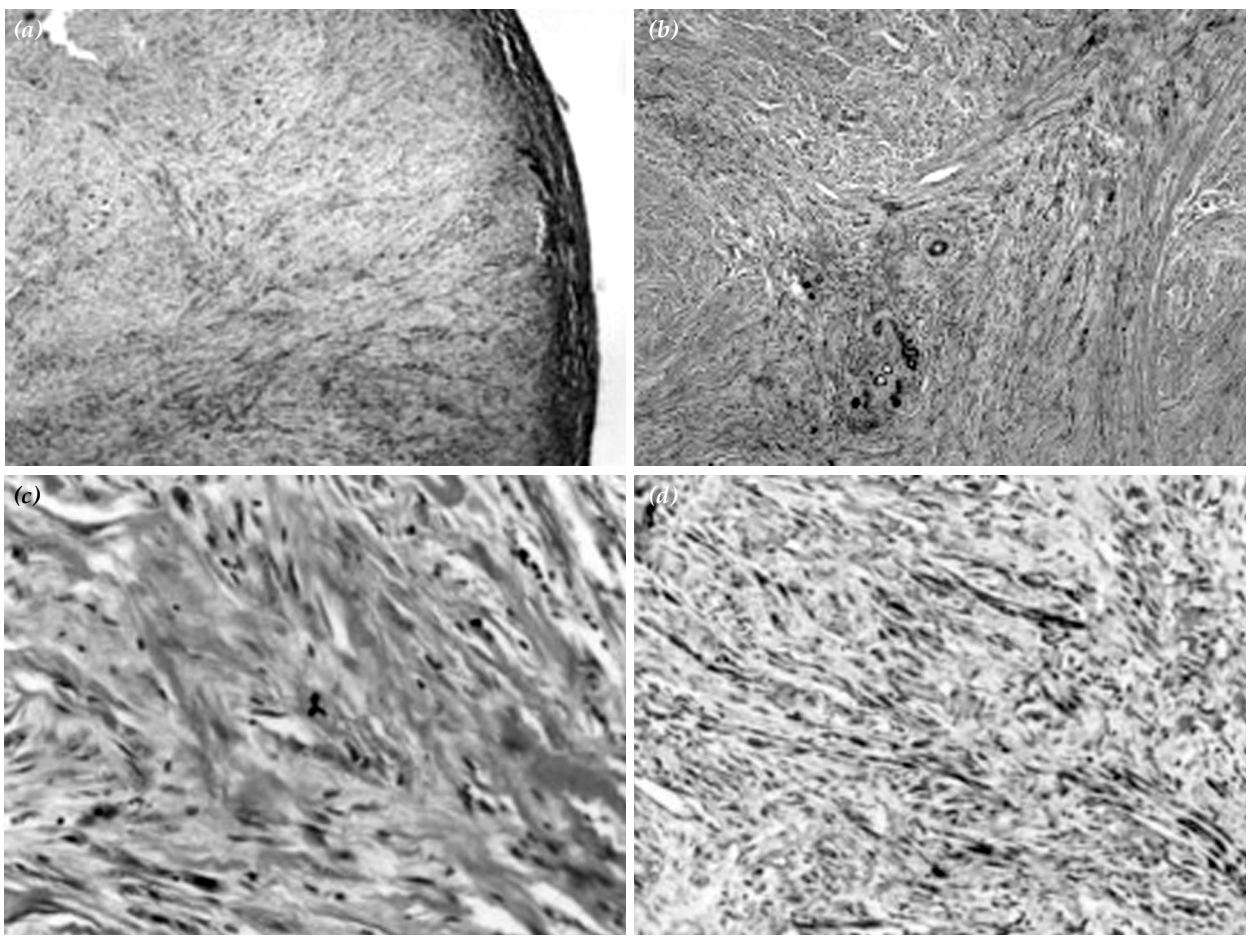

Figure 2. ***(a, b)*** *The tumor is relatively well circumscribed but invaded striated muscle fibers in some areas (H-E x 40).* ***(c)*** *An atypical mitosis in the tumor (H-E x 200).* ***(d)*** *The tumor expressed a diffuse and strong S-100 positivity (S-100 x 100).*

DISCUSSION

Malignant peripheral nerve sheath tumors are very rare, constituting between 5 to 10% of all soft tissue sarcomas.[1] Hence, they are often managed as a sub-category of soft tissue sarcomas. Malignant peripheral nerve sheath tumors usually arise in adults between 20 and 50 years of age, with a roughly equal distribution between men and women.[3] These tumors may arise spontaneously, although approximately 20% of MPNST have an association with neurofibromatosis type 1 (von Recklinghausen's disease), an autosomal-dominant disorder that involves the NF1 tumor suppressor gene, which is located on chromosome 17.[3,4]

They arise from major or minor peripheral nerve branches or sheaths of peripheral nerve fibers. However, it is not always possible to demonstrate the origin from a nerve, especially when it arises from a small peripheral branch.[7] A combination of gross and microscopic findings along with immunohistochemical studies is commonly used to diagnose a case of MPNST. Nonetheless, mitotic index, cellularity, nuclear atypia, and necrosis are the most reliable histologic criteria of malignancy.

According to the largest reported MPNST series comprising 205 patients, only 4% of MPNSTs are located in the head and neck region.[3] In the head and neck region, the most common localizations of the tumor are neck and lateral skull base.[8,9] Only a few cases of MPNSTs in the lower lip have been reported.[10,11]

Radical surgical resection is the treatment of choice in MPNST.[3,5] Adequate initial treatment gives the best chance of survival and complete tumor resection with negative margins should be the objective of surgery. Routine nodal dissection is not indicated.[4] However when a major nerve is identified, the cut end should be sent for frozen section to assess the tumor free margin of resection.[4] Adjuvant radiation therapy should be delivered to improve local control and may also be beneficial for survival. However, local recurrence is also a feature of this disease.[3] Despite aggressive surgery and adjuvant therapy, the prognosis for patients with MPNST remains poor.[3,5] The probability of achieving a cure is no greater than 40%.[3] Presentation with either primary or recurrent disease, tumor size and the site of origin are the most important prognosticators for cause-specific survival in patients with MPNST.[3]

The lower lip is a very unusual site of involvement for MPNSTs. Although very rare, MPNSTs should be excised totally as an initial treatment when encountered in the head and neck region.

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