OLGU SUNUMU CASE REPORT

DOI: 10.24179/kbbbbc.2021-84729

Neurofibroma in the Nasal Cavity: A Rare Clinical Entity

Nazal Kavite Yerleşimli Nörofibrom: Nadir Bir Klinik Durum

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This case report was presented as as a poster presentation in 11. National Turkish Rhinology Meeting, 16-19 April 2015, Antalya, Türkiye.

ABSTRACT Most peripheral nerve sheath tumors are benign. Neurofibroma, one of the peripheral nerve sheath tumors, is rarely seen in the nasal cavity. Definitive diagnosis of neurofibroma can be made by histopathological examination of the specimen. A 46-year-old female patient was admitted to Fatih Sultan Mehmet Health Practice and Research Center Ear Nose and Throat clinic with swelling complaints that have been slowly growing at the entrance to the left nasal cavity for a long time. In the examination, approximately 1x1 cm mass was detected under the skin in the left medial crus of the alar cartilage. The patient underwent mass excision from the left nasal passage under local anesthesia. The patient had no postoperative complications. Histopathological examination of the operation material was reported as a neurofibroma. The aim of this case report is discuss case in light of literature in the terms of symptoms, diagnosis and surgical methods.

Keywords: Neurofibroma; nasal cavity; peripheral nerve sheath tumor

ÖZET Periferik sinir kılıfı tümörlerinin çoğu benignder. Periferik sinir kılıfı tümörlerinden olan nörofibroma nazal kavitede nadiren görülmektedir. Nörofibromun kesin tanısı; spesimenin histopatolojik inceleme sonucuyla konulabilir. 46 yaşında kadın hasta, uzun senelerdir sol nazal kavite girişinde yavaş bir şekilde büyüme gösteren kitle şikayeti ile Fatih Sultan Mehmet Eğitim ve Araştırma Hastanesi Kulak Burun Boğaz polikliniğine başvurdu. Hastanın preoperatif muayenesinde solda alar kartilaj medial yüzü üzerinde cilt altında yaklaşık 1x1 cm boyutlarında kitle tespit edildi. Hastaya lokal anestezi altında sol nazal pasajdan kitle eksizyonu operasyonu yapıldı. Hastanın postoperatif komplikasyonu olmadı. Operasyon materyalinin histopatolojik inceleme sonucu nörofibrom olarak raporlandı. Ameliyat spesmenin histopatolojik sonucu; nörofibrom gelmiştir. Bu bildirinin amacı; olgunun semptom, tanı ve tercih edilecek cerrahi yöntemleri açısından literatür ışığında tartışılmasıdır.

Anahtar Kelimeler: Nörofibrom; nazal kavite; periferik sinir kılıfı tümörü

Peripheral nerve sheath tumors consist of cells of neural crest and mesenchymal origin.[1] Neurofibroma, one of the peripheral nerve sheath tumors, can be seen in the head and neck region, also on the flexor faces of the upper and lower extremities, yet; it is rarely seen in the nasal cavity and paranasal sinuses (PNSs).[2-4] The purpose of this case report, in which a case diagnosed with neurofibroma located in the nasal cavity is presented, is to discuss the case in the light of the literature in terms of symptoms, diagnosis and preferred surgical methods.

CASE REPORT

A 46-year-old female patient was admitted to Fatih Sultan Mehmet Health Practice and Research Center Ear Nose and Throat clinic with swelling that has been growing gradually for a long time at the entrance to the left nasal cavity. The examination showed a mass thought to be associated with medial crus, which closed the left nasal cavity (Figure 1a).

Nodular lesion, in 10x9 mm size with a neat contoured soft tissue, was detected in the patient's PNS

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Peer review under responsibility of Journal of Ear Nose Throat and Head Neck Surgery.

Received: 30 May 2021 ***Received in revised form:*** 26 Aug 2021 ***Accepted:*** 24 Nov 2021 ***Available online:*** 02 Dec 2021

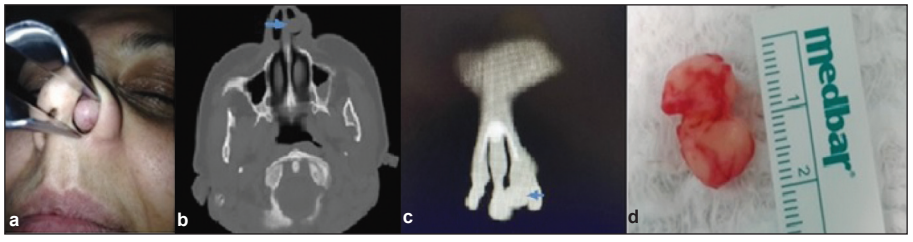

FIGURE 1: a) Preoperative view of the mass. Preoperative computed tomography imagings (**b:** Transverse image; **c:** Coronal image, blue arrow indicates the mass). **d)** Post-excision view of the mass.

computed tomography (PNS CT) film, extending from the inner wall of left alar cartilage to the nasal cavity. No destruction was observed in the bony and orbital structures in the PNS CT (Figure 1b; transverse image, Figure 1c; coronal image). The excision of the mass was planned under local anesthesia. In the left nasal passage, a mass with hard nature approximately in size of 1x2 cm that originating from the lower lateral cartilage was excised preserving the overlying mucosa. The lateral crus cartilage appeared to be invasive with the mass. This region was also included in the specimen. Cartilage defect occurred 2 mm lateral from the dome. The preserved mucosa was used for reconstruction of defective area. Gauze with furacin was placed on the inner and outer faces and transfection sutures were discarded. The operation was terminated by putting an extrafor tampon with furacin in the left nasal passage and operation material was sent to pathology for examination. The patient had no postoperative complications. Cartilage grafts were planned for the lateral cartilage after the healing of the mucosal tissue in the second session, but the patient did not want a second operation. The patient was informed about her situation and informed consent form was obtained from the patient for using in the case report.

Histopathological examination of the operation material was reported as a neurofibroma. The mass was found to be 20x10x10 mm in size on macroscopy in pathological examination (Figure 1d). A solid spindle cell tumor with a lobulated structure with irregular bundles was observed in microscopically examination (Figure 2). It was observed that the tumor consisted of cells with smooth and elongated oval nuclei and thin chromatin, forming irregular bundles on a collagenized and slightly myxoid background, the cytoplasm boundaries of which could not be identified (Figure 3a). Immunohistochemical staining showed diffuse positivity with tumor cells S-100 (Figure 3b). Some cells had CD34 positive staining (Figure 4a). No staining was seen with smooth muscle actin except vessels (Figure 4b).

DISCUSSION

Differential diagnosis of nasal masses is difficult. Since nasal masses can be of neoplastic, developmental and inflammatory origin, their differential diagnosis is quite wide. Benign tumors of the nasal cavity usually originate from the epithelium, but tumors originating from the peripheral nerve sheath are very rare.

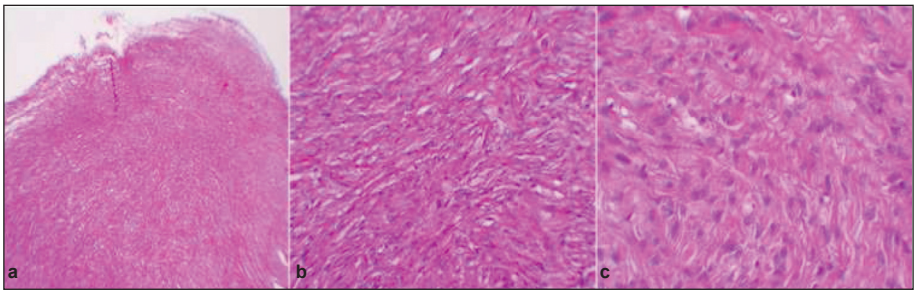

FIGURE 2: Solid spindle cell tumor that forms irregular bundles in a lobulated structure. (**a:** Hematoxilyn- Eosine (H-E) x100, **b:** H-E x200, **c:** H-E x400).

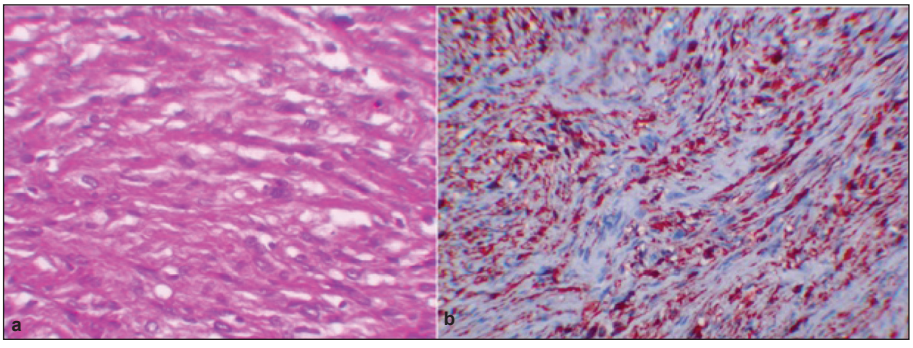

FIGURE 3: a/b) Cells with elongated oval nuclei and fine chromatin, whose cytopic borders cannot be detected/Common S-100 positivity. (**a:** H-E x400, **b:** S-100 x400).

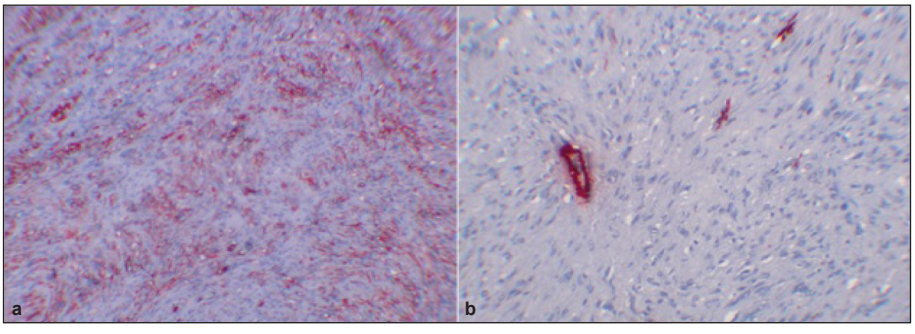

FIGURE 4: a/b) CD34 positivity/No staining was seen with smooth muscle actin other than vessels. (**a:** CD34 x200, SMA x200).

Peripheral nevre sheath tumours are classified into benign and malignant; and neurofibroma is one of the benigns. Neurofibromas are nerve sheath tumors that originate from the peripheral nervous system. They develop from non-myelin schwann cells. Neurofibromas originate from a combined proliferation of several elements of a peripheral nerve, including axons, Schwann cells, fibroblasts, perineurial cells,and endoneurium.[5] In the differential diagnosis of neurofibroma; inflammatory polyps, juvenile angiofibroma, inverted papilloma, meningioma, melanoma and olfactory neuroblastoma should be considered.[2-8] Although generally associated with type-1 neurofibromatosis, it may be isolated, and is then known as solitary neurofibroma. They are commonly seen in the head and neck region, extremities, mediastinum and retroperitoneal area. Nasal and paranasal location is extremely rare.[6,7] While it is seen 45% in the head and neck region, it is seen 4% in the paranasal sinus and nose.[4,7] Although neural origin could not be determined in the etiopathogenesis of nasal schwannoma; terminal somatosensory branches of the trigeminal nerve may play a role.[3-6] Cause of that, neurofibroma, which is thought to originate from peripheral nerves, is more common around the nasal vestibule than in other regions in the sinonasal tract. It is most common seen in the nasal cavity (64%), and followed by the maxillary sinus (17%).[5,9] Neurofibromas are slowly growing benign tumors. However; they can be grow that causes destruction due to local compression, and also could suggest a malignant process rarely.[2,10] Although it is seen in all age groups, its incidence is higher in the fourth and sixth decades. It affects men and women equally.[1-6]

Clinical findings of nasal schwannomas are nonspesific and similar to other nasal tumors. Clinical symptomatology is as in any process occupying the nasal and sinus cavities. Mucopurulent nasal discharge, nasal congestion, bleeding, inability to smell and pain are the most common symptoms, and these symptoms have been present for a long time.[3-8] Therefore; neurofibroma of the nasal cavity is frequently misdiagnosed.[5,11] Our case had a nasal mass and nasal obstruction on the left side.

Schwannom is macroscopically well-circumscribed, oval-round shaped, its cross-sectional surface has an elastic consistency, yellowish gray skin color and solid. Those located in the superficial soft tissue are usually 1-5 cm in diameter, while in the mediastinum and retroperitoneum that can reach up to 10 cm.[1,6]

The diagnosis is made by histological and immunohistochemical findings. Histological examination reveals that spindle cell groups with nuclei lined up in an elongated palisad pattern. S-100 protein positivity is an immunohistochemical indicator that the mass originates from Schwann cells.[2,3] Furthermore, the presence of CD34 (+) can be seen as an immunohistochemical finding in these tumors.[3] Carcinoma, sarcoma and lymphoma should be considered in the differential diagnosis. The definitive diagnosis is made by histological examination of the biopsy specimen.

Conservative approach is recommended for these tumors due to their low risk of recurrence, slow growth and non-invasiveness. Tumors of peripheral nerve origin are resistant to radiotherapy. The most appropriate treatment approach is surgical resection of the tumor.[1-6] Recurrence is rare after total removal of the mass. In our case, the mass was totally removed by surgical excision. There was no recurrence in the six-month follow-up after excision.

Although it is rarely seen in those who are admitted with nasal mass in otorhinolaryngology policlinics, it should think in patients who have a mass for a long time, neurofibroma should also be considered in the differential diagnosis. It can be diagnosed with the characteristic microscopic features in the histopathological examination to be made after it is thinking in the differential diagnosis and local excision is the curative treatment.

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Source of Finance

During this study, no financial or spiritual support was received neither from any pharmaceutical company that has a direct connection with the research subject, nor from a company that provides or produces medical instruments and materials which may negatively affect the evaluation process of this study.

Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: *Serhan Keskin, Tuğba Aslan Dündar, Arzu Tatlıpınar;* ***Design:*** *Serhan Keskin, Emrah Tekdemir, Arzu Tatlıpınar;* ***Control/Supervision:*** *Serhan Keskin, Tuğba Aslan Dündar, Arzu Tatlıpınar;* ***Data Collection and/or Processing:*** *Emrah Tekdemir, Adnan Somay;* ***Analysis and/or Interpretation:*** *Serhan Keskin, Arzu Tatlıpınar, Adnan Somay;* ***Literature Review:*** *Serhan Keskin, Tuğba Aslan Dündar, Emrah Tekdemir;* ***Writing the Article:*** *Serhan Keskin, Arzu Tatlıpınar, Tuğba Aslan Dündar;* ***Critical Review:*** *Arzu Tatlıpınar, Serhan Keskin.*

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