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What is your diagnosis?

Tanınız nedir?

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Solitary mass of the external nose

Case

A 72-year-old male patient was brought to our clinic due to a gradually enlarging lump on the right side of his ala nasi, which had been present for two years (Figure 1a). He had no significant medical history. An immobile mass of rubber consistency 2.5x2.5 cm in size with a polypoid appearance was found on the right ala nasi that expanded the right nasal wing and infiltrated the surrounding cheek tissue. No lymphadenopathy was found during the examination of the neck.

A punch biopsy was performed. Histopathological examination revealed that it was a mesenchymal neoplasm with myxoid features.

Under general anesthesia, the mass on the right nasal wing was completely resected, including the infiltrating cheek tissue with 0.5 mm safety margins. The defect was repaired by advancing the decollated cheek tissue towards the nose and suturing the remnant of the nasal wing to the alar crease. Although the right nostril was narrower than the opposite side, an aesthetically acceptable appearance was obtained. The patient underwent follow-up care with dressings and experienced a smooth recovery (Figure 1b). At 11-month follow-up, there were no signs of recurrence.

Diagnosis: Solitary myxoid neurofibroma originating from the external surface of the nose.

Microscopic findings and laboratory investigations: Histopathology revealed nodular formations in the dermis. It contained variable-sized collagen bundles, spindle cells, and mast cells interspersed in the myxoid edematous stroma in the dermis. Immunohistochemically, neural cells were positive for S100. The findings were compatible with myxoid neurofibroma (Figure 1c, d).

Since there was no family history and no findings related to neurofibromatosis type 1 or 2 in our case, the lesion on the nasal wing was considered a solitary neurofibroma.

Discussion

Neurofibromas are benign tumors originating from peripheral nerve cells. Histological examinations reveal three distinct categories of neurofibromas: Plexiform, diffuse, and solitary. Among these, plexiform neurofibromas are associated with neurofibromatosis type 1, also referred to as von Recklinghausen's disease. Growing multifocally along the affected nerve, they infiltrate soft tissue and show a more aggressive course. Diffuse neurofibromas present themselves as plaques and may be extremely large. Conversely, solitary neurofibromas present as glossy masses with clear boundaries and a rubber-like texture. The precise origin of solitary neurofibroma remains unclear; it is regarded more as a hyperplastic hamartomatous malformation than a true neoplastic condition[1-3].

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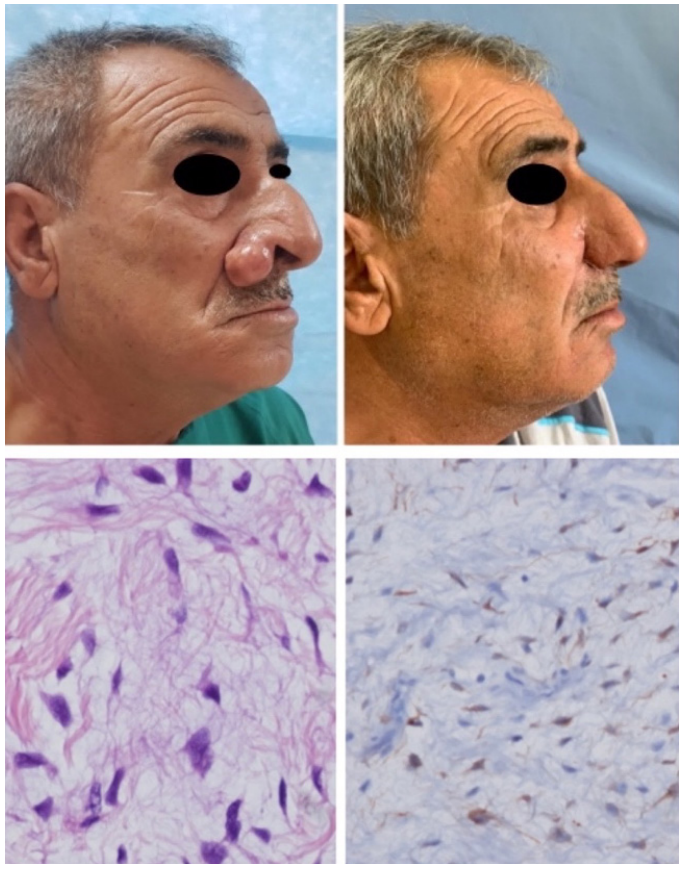

Figure 1. (a) Preoperative appearance of the patient (Lateral view). **(b)** Postoperative appearance of the patient (Lateral view). **(c)** Collagen bundles and spindle cells in the myxoid stroma (HE, x400). **(d)** S-100 positivity in spindle cells (HE, x200)

Solitary neurofibromas are usually located in the head and neck region and on the flexor surfaces of the extremities. The most common source of these tumors is the vestibular nerve in the head and neck region. However, solitary neurofibromas on the nose's outer part are very rare. They may arise from the ophthalmic and maxillary branches of the trigeminal nerve. Clinically, they appear as a slow-growing, smooth-surfaced mass that can reach a few centimeters in diameter. They can cause symptoms depending on the region where they are located[2,4,5].

Our case had no complaints other than the appearance of the nasal wing.

Neurofibromas can occur in all anatomical regions with myelin-sheathed nerves. They are well-differentiated, non-encapsulated tumors. They consist of Schwann cells, perineural cells, and fibroblasts. Nested thin spindle cell bundles, wavy dark-stained nuclei, abundant collagen bundles, and mast cells are located in the myxoid stroma. Specific diagnosis is made with S-100 positivity[6,7].

A differential diagnosis should involve malignant tumors, inflammatory or cystic lesions, lipomas, schwannomas, leiomyomas, granular cell tumors, neuromas, and adenomas[7].

Complete resection of the tumor is the best treatment option since it can recur if not completely removed. Subsequent repair of the defect is to be planned according to the location and size of the tumor.

In our case, a narrowing occurred in the right nostril compared to the left after the repair. It was planned to enlarge the nostril using a nostril retainer[7,8].

Neurofibromas require regular follow-up of the cases, as they can recur even after many years and rarely transform into malignancy[6,9].

In summary, by presenting this case, we aimed to raise awareness of this rare disease and reduce morbidity with early diagnosis and treatment supported by gathering clinical evidence.

Ethics

Informed Consent: It was obtained.

Authorship Contributions

Surgical and Medical Practices: S.S.E., A.K., Concept: S.S.E., A.K., Design: S.S.E., A.K., Data Collection or Processing: S.S.E., A.K., Analysis or Interpretation: S.S.E., A.K., Literature Search: S.S.E., A.K., Writing: S.S.E., A.K.

Conflict of Interest: No conflict of interest was declared by the authors.

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