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Case Report

A case of solitary neurofibroma in outer surface of nose

Shahin Abdollahi-Fakhim[1], Saman Rezaeian[2], Hojjat Hosseinpour-Feizi[3], Mohammad Naghavi-Behzad[4], Reza Piri*[5]

[1] *Associate Professor, Department of Ear, Nose and Throat (ENT), School of Medicine, Tabriz University of Medical Sciences, Tabriz, Iran*

[2] *Resident, Department of Ear, Nose and Throat (ENT), School of Medicine, Tabriz University of Medical Sciences, Tabriz, Iran*

[3] *Associate Professor, Department of Orthopedics Surgery, Shohada Educational Medical Hospital, Tabriz University of Medical Sciences, Tabriz, Iran*

[4] *Student of Medicine, Students' Research Committee, Tabriz University of Medical Sciences, Tabriz, Iran*

[5] *Student of Medicine, Medical Philosophy and History Research Center, Tabriz University of Medical Sciences, Tabriz, Iran*

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Abstract
Introduction: Solitary neurofibroma is a benign tumor originating from Schwann cells in peripheral nerves. In an investigation of 430000 pathologic samples, only 6 cases of neurofibroma had been reported in nose and paranasal sinuses.
Case Report: Present patient was a 15 year old girl with left-sided nasal and malar mass, whose computed tomography (CT) scan and magnetic resonance imaging (MRI) showed a non-circumscribed solid mass with total thickness affecting outer part of nose in left side as well as surface and deep soft tissue. The mentioned mass was excised by sub-labial incision and open rhinoplasty keeping infra-orbit nerve intact. Pathologic investigation of samples showed adipose fibroblast and skeletal muscle fibers with proliferation of spindle-shaped cells and staining was negative for glial fibrillary acidic protein (GFAP) and positive for S-100 which represented the diagnosis of neurofibroma.
Conclusion: Main differential diagnoses of benign tumors of paranasal sinuses and nasal cavity include neurofibroma, fibroma, leiomyoma, papilloma, and schwannoma. After resection of the tumor, patient should be monitored precisely since there is a possibility for malignant transformation in solitary neurofibromas.

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Introduction

Peripheral nerve sheath tumors include neurofibromas, schwannomas and neurogenic sarcomas. Neurofibromas and schwannomas are classified as benign tumors and both are believed to arise from Schwann cells.[1,2] Solitary neurofibroma is a benign, slow-growing, circumscribed, but not encapsulated tumor. This tumor essentially is composed of Schwann cells arising from a peripheral nerve and usually affects skin or subcutaneous tissue.[3,4] Neurofibroma is generally presented in background of Von Recklinghausen's disease, but it can also appear solitarily.[5] This tumor is usually found in the head and neck and the flexor surfaces of the extremities. Vestibular nerve is the most common origin of these tumors in the head and neck region.[6] However, neurofibromas arising from the nose and paranasal sinuses are rare.[7] The great majority of the peripheral nerve sheath tumors reported were schwannomas in the paranasal sinuses and nasal cavity. In a review study of 430000 pathological samples, six neurofibromas involving the paranasal sinuses or nasal cavity were detected.[8]

In this article we report a neurofibroma

*** Corresponding Author:** Reza Piri, Email: dr.reza.piri@gmail.com

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case in nose which was successfully excised surgically.

Case Report

Patient was a 15 year old girl with left-sided nasal and malar mass, which had developed during a 10 year period. Patient did not complain of face or nose pain, headache, epistaxis, nasal discharge, or adenoid. She mentioned a history of a blunt trauma to nose which had not been followed-up. Patient had no history of systemic diseases in the past. In physical examination, a subcutaneous mass was observed in left lateral side of the nose leading to elephantiasis of left side of the nose. The mass had spread towards the face medially and had also caused deformity in the face. There was alar cartilage, nostril and left nasolabial chin downward dislocation. While touching, there was a mass with approximate size of 7 × 5 cm, with non-circumscribed, soft and non-tender consistency on left side of the nose and subcutaneous tissue. Skin on mass seemed unfiltered and thick but lacked any wound and superficial lesion. Mass had no adhesion to the underlying tissue. In intranasal examination, there was no symptom of wound, mass or mucosal lesion. Eye examination was normal. There was no lymphadenopathy in the neck. Neurologic examination of the patient was normal. There was no pigmentation and mass on the skin of other parts of the body (Figure 1).

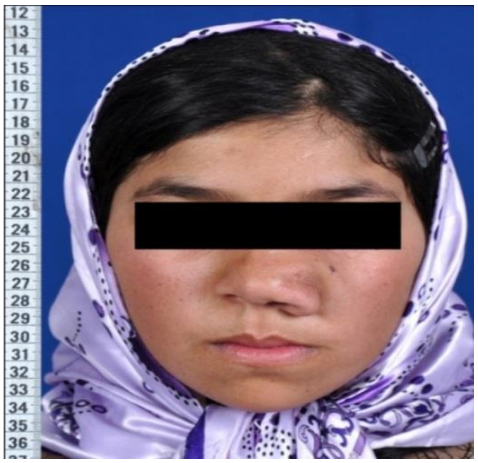

Figure 1. Photograph of the patient before surgery (notice the left external nasal mass)

Radiography of the chest of the patient was normal. Computed tomography (CT) scan and magnetic resonance imaging (MRI) showed a non-circumscribed solid mass with total thickness affecting outer part of the nose on left side. Surface and deep soft tissue affection of the left cheek continued to the surface of maxillary bone. Lower part of the mass continued to the midline up to upper lip philtrum but there were no bone destruction or erosion. The mass was hyperemic and had enhancement after injecting contrast media on MRI.

In operation room excision was done on the mass (with relatively soft consistency and fibro fatty appearance) as much as possible first by sub-labial cutting and by keeping infra-orbit nerve intact. Then in nasal part, open rhinoplasty cutting was done, flap was raised and by help of sub-labial approach, the mass was completely excised. Then suturing was done and drainage was placed under lip and inter alar base sutures. Additional skin was not removed.

Pathologic investigation of samples showed adipose fibroblast and skeletal muscle fibers with proliferation of spindle-shaped cells whose long nuclei were stream, twisted bundles, and arranged in plexiforms (Figure 2).

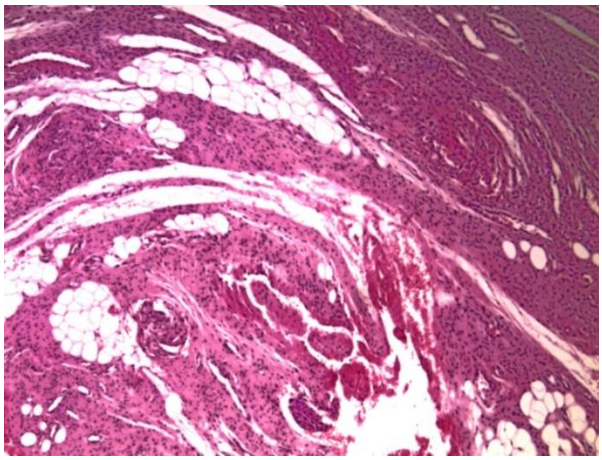

Figure 2. Pathologic view of the mass

There was growth of spindle-shaped cells and fibroblasts around nerves and these findings presented neurogenic tumors such as glioma and neurofibroma. In further

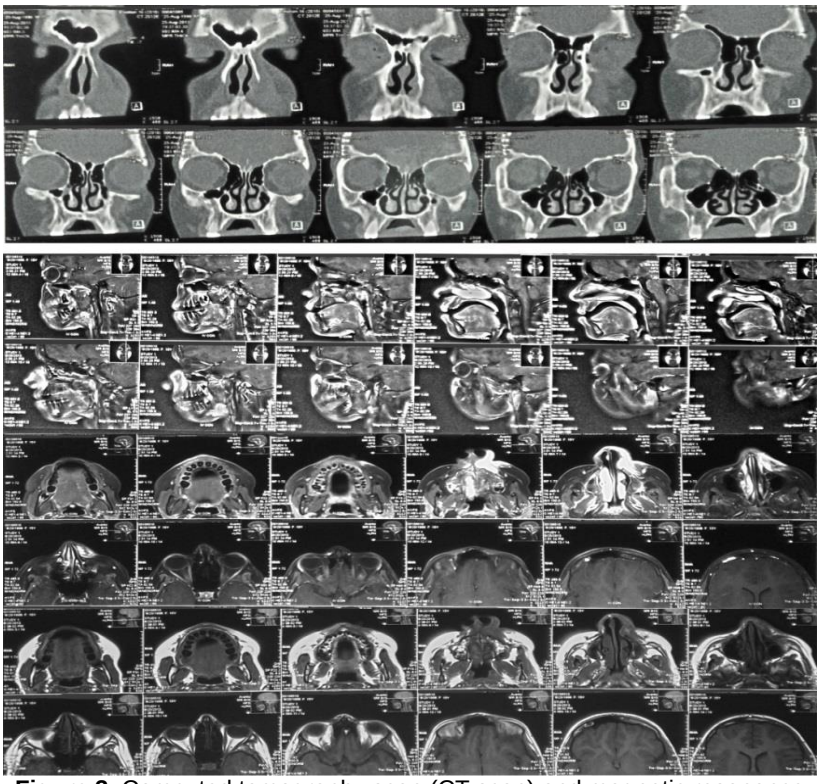

Figure 3. Computed tomography scan (CT scan) and magnetic resonance imaging (MRI) of maxillofacial region

investigations, staining was negative for glial fibrillary acidic protein (GFPA) and positive for S-100 which represented the diagnosis of neurofibroma (Figure 3).

After the first surgery due to skin excess in nasal and lateral malar, further surgeries were advised in several steps in order to reduce deformation. Patient did not receive other therapies. Patient did not have any specific problem in post-operative period. Patient was regularly monitored and during a year after surgery no complications of surgery or symptoms of tumor recurrence were observed.

Discussion

Solitary neurofibromas are slow-growing benign tumors, which are circumscribed, but not encapsulated. These tumors are essentially composed of Schwann cells arising from a peripheral nerve, which usually involve the subcutaneous tissue or skin.[2,4] Involvement of paranasal sinuses in those neurogenic tumors originating from head, neck and flexural surfaces of the extremities have been reported to be rare.[9] In paranasal sinuses and nose, the tumor arises from autonomic plexuses and from the 1st and 2nd division of the trigeminal nerve, but structures such as olfactory nerve which have no Schwann cells cannot harbor these tumors.[7,10] These tumors can become very large causing dislocation and compression of local structures (such as expansion and erosion of local bone).[11] Like any other benign lesions, basically, neurofibromas present symptoms and signs depending on the site of the tumor and the surrounding structures involved.[7]

The affected age group is usually from 25 to 55 years old. Most of the symptoms which patients present with are non-specific like facial pain (especially maxillary tumors), nasal obstruction and epistaxis (more in ethmoidal sinus and nasal fossa). Of conditions faced in these tumors are thrombi and hemorrhagic necrosis which may be the reason for misdiagnosing them as vascular tumors like angiofibromas or fibrotic polyps of nasal cavity. Thus, they are difficult to diagnose clinically in the absence of Von Recklinghausen's disease.[12]

Symptoms and signs mostly depend on the site of the tumor. The most common symptoms and signs are nasal obstruction, facial pain, epistaxis, proptosis, and swelling. The main diagnostic procedure for neurofibroma is biopsy, because most of these symptoms and signs are non-specific.[7] Despite their indolent growth, neurofibromas can occasionally become very large, resulting in local bone destruction and intracranial extension. This tumor may distort tissues by pressure or become symptomatic by obstruction of a sinus ostium.[1]

Considering histological aspects, neurofibroma is ill-defined and poorly circumscribed with non-encapsulated margin. This is characterized by hypocellular proliferation eosinophilic spindle cells with rather wavy, shaped or buckled nuclei set in a copious fibrillary or rather myxoid background.[13] Mostly mitosis is not seen. Usually within the tumor, small nerve fibers are readily identified. Occasionally the stroma of these lesions undergoes marked myxoid or hyaline change. Considering ultrastructural features, this is composed of a mixture of perineural fibroblasts and Schwann cells, which are S-100 positive.[7,14]

CT scanning and MRI are the main imaging methods used to reveal the size of the tumor, which is essential to plan further interventions such as surgical excision. Mostly MRI is thought to outweigh CT images since it can allow better distinction of the tumor from neighboring soft tissues, beside enhanced assessment of possible intraorbital and intracranial extension.[12,14]

The general management of neurofibromas mostly depends on the signs and symptoms which patient presents with. Small lesions not causing any issues may not require any invasive interventions, other than monitoring. Surgical exploration, biopsy, and excision are necessary if there is a suspicion of the tumor being malignant.[12]

Neurofibromas tend to infiltrate more extensively than schwannomas, and thus may require extensive surgical resection. However, functional and cosmetic considerations should be taken into account because neurofibromas, even if incompletely excised, may recur after many years. From this point of view, en bloc tumor resection via an external rhinoplasty approach seems an appropriate choice in terms of the aesthetic and oncological considerations. After en bloc resection, the tumor should be closely followed because the malignant transformation of solitary neurofibroma is also possible even though it is rare as compared with the sarcomatous changes that occur in 5.5% to 16% of neurofibromatosis patients.[15]

Conclusion

After surgery due to skin excess in nasal and lateral malar, further surgeries were advised in several steps in order to reduce deformation in this patient. Patient was regularly monitored and during one year follow up after surgery, no complications of surgery or symptoms of tumor recurrence were observed.

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Authors' Contribution

Shahin Abdollahi-Fakhim, Saman Rezaeian: conception and design, acquisition of data or analysis and interpretation of data; Hojjat Hossein Pourfeizi, Mohammad Naghavi-Behzad: drafting the article or revising it critically for important intellectual content; Reza Piri: conception and design, acquisition of data or analysis and interpretation of data, final approval of the published version.

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Conflict of Interest

Authors have no conflict of interest.

Ethic Approval

This study was approved by Tabriz University of Medical Sciences Ethics Committee.

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