

Rare Case of Solitary Neurofibroma Dorsum of Nose

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ABSTRACT

Introduction: Neurofibromas are usually solitary, although multiple tumours can be seen in patients affected with neurofibromatosis. These tumour can develop anywhere in nervous system including brain, spinal cord and nerves, but are relatively uncommon in sinonasal tract and paranasal sinuses, with only a few cases reported in literature. They generally arise in intimate association with peripheral nerve trunk, developing from Schwann cells, perineuritis and blended with fibroblast if cells. Benign tumours are usually solitary, although multiple tumors are seen more often in patients affected with neurofibromatosis. Neurofibromatosis is considered within differential diagnosis of other spindle cell tumors of sinonasal tract.

Case report: The article presents a rare case of neurofibroma on dorsum of nose crowding pattern in mixed dentition and discuss the appropriateness of this appliance against other conventional appliance.

Conclusion: Tumors of mesodermal origins, have high suspicion of malignancy & should always go for surgical excision.

Keywords: Neurofibroma, Fibroblast, Schwann Cells, Malignancy

INTRODUCTION

According to the 2017 World Health Organization Classification of Head and Neck Tumors, neurofibromas are benign peripheral nerve sheath tumors that consist of a mixture of Schwann cells, perineurial cells, fibroblasts, and axons. Neurofibromas resemble schwannomas and are often seen as solitary, elastic, hard, well defined masses with no capsule. [2]. Dermal neurofibromas arise from a single peripheral nerve, while plexiform neurofibromas are associated with multiple nerve bundles. Neurofibromas may appear as part of neurofibromatosis type I (NF-1) or von Recklinghausen disease, and they present most frequently with café-au-lait spots on the skin [3].

These tumours can develop anywhere in nervous system, including brain, spinal cord and nerves, but are relatively very uncommon in sinonasal tract and paranasal sinuses, with only a few reported cases in literature. They generally arise in intimate association with peripheral nerve trunk, developing from Schwann cells, perineurites, and blended with fibroblastic cells. Neurofibromatosis is frequently considered within differential diagnosis of other spindle cell tumours of sinonasal tract. We describe a rare case of fibroma in sinonasal tract and para nasal sinus reported in a patient.

CASE REPORT

A 51 year old male presented in the outpatient department with a swelling over the right dorsum of the nose, which

started 2 months back. It was gradual in onset and had a progressive nature. On palpating, it was a painless palpable mass. There was no reported history of itching or discharge from the nose. On being asked about any relevant past medical history, there haven't been any significant surgical/medical intervention. All the family members are healthy and apparently enjoying good health, hence, no relevant family history obtained.

The patient underwent general physical and local examination at the department itself.



Figure-1:



Figure-2:

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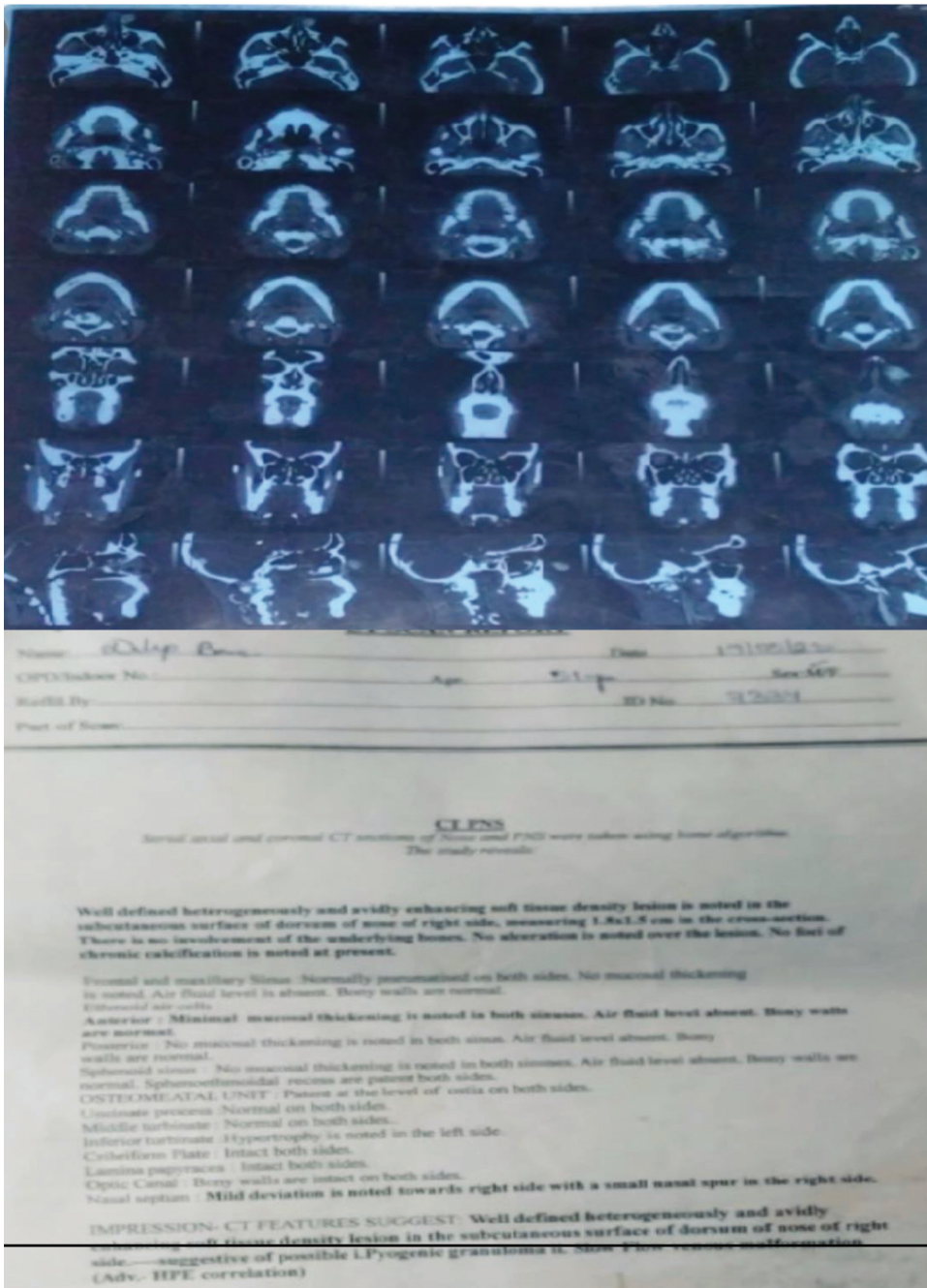


Figure-3:

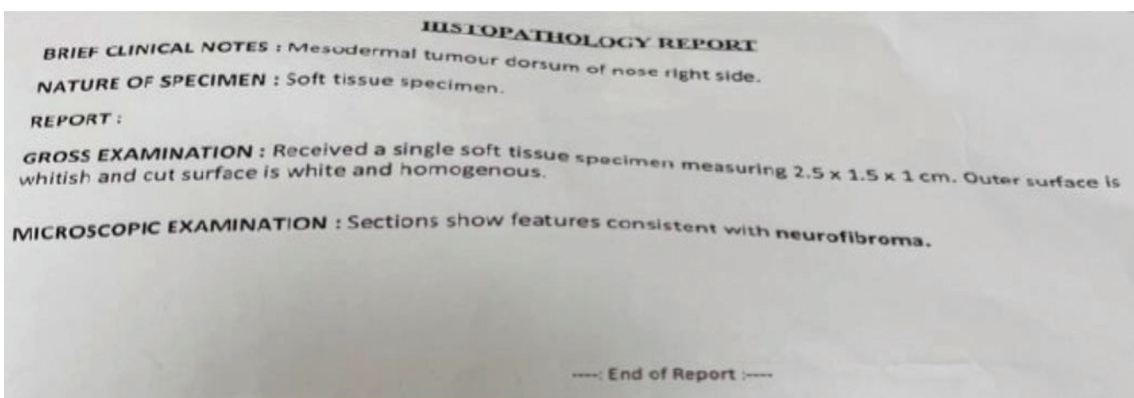


Figure-4:



Figure-5:

During general physical examination, the patient was well oriented to time, place and person and conscious. While on local examination there was a swelling observed over the right dorsum of nose. We observed a well-defined dome-shaped mass measuring approximately 2.5*2cm. The surface was observed and found to be irregular and non-tender on touch with a normal mucosal colour. The non-cystic swelling was found to be horizontally mobile and had a broad base. Also, the patient didn't experience any pain, tenderness or surface hypesthesia. CT PNS was suggestive of well defined heterogeneously and avidly enhancing soft tissue density lesion, suggestive of:

1) Neurofibroma 2) slow flow venous malformations.

FNAC from nasal mass showed haemorrhagic material. Completely excised mass sent for HPE evaluation, which was suggestive of neurofibroma.

Investigations

In local examination of ear, nose and throat we saw tip raising test was done on it to examine the bilateral nasal cavities, which turned out to be normal.

Anterior rhinoscopy was also, nasal mucosa was congested, on posterior rhinoscopy change and posterior turbinate's appear normal.

Provisional diagnosis of Fibroma was made

All the routine investigations were done, along with radiological and pathological investigations. All the blood investigations were apparently normal. CT scan of PNS shows, well defined heterogeneously and avidly enhancing soft tissue density lesion in the subcutaneous surface of dorsum of nose of right side, suggestive of possible (i) pyogenic granuloma (ii) slow flow venous malformation. Histopathological report showed, a single soft tissue specimen measuring 2.5*1.5*1cm, with outer surface showing whitish and cut surface is white and homogenous. Microscopic examination shows feature suggestive of neurofibroma

DISCUSSION

Neurofibroma is a benign peripheral nerve sheath tumor with classic identifiable features including presence of a neuronal component comprising transformed Schwann cells and a non-neoplastic fibrous component involving fibroblasts.

The essential features include interlacing bundles of elongated cells with wavy darkly stained nuclei, interspersed with mast cells and collagen bundles.

CD34 positivity is present in cells of unclear histogenesis. S100 positivity in neural cells. It occurs mostly during the 2nd or 3rd decades of life, with equal male to female predisposition.

90% of solitary NF are sporadic and 10% inherited. Types of NF are: Localized, diffuse and plexiform. Clinical features consist of sporadic, cutaneous and subcutaneous. Associated with NF, that is type 1 and type 2.

DIFFERENTIAL DIAGNOSIS

Schwannoma

Ganglioneuroma

Myxoid liposarcoma

Nerve sheath myxoma

Perineuroma

CONCLUSION

Tumors of mesodermal origins, have high suspicion of malignancy & should always go for surgical excision.

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