Neurofibroma of Middle Meatus: A case report and Review of Literature

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This is a case report of a 46 years old female diagnosed to be neurofibroma arising from the left middle meatus. She underwent endoscopic sinus surgery and there is no recurrence till 3 months. A short review of literature is also presented.

Keywords:
Neurofibroma, middle meatus, endoscopic sinus surgery.

INTRODUCTION:

Peripheral nerve sheath tumours are divided into neurofibroma, schwannoma and neurogenic sarcoma. Neurofibroma and Schwannoma are classified as benign and both are believed to arise from a common origin - the Schwann cell.[1] Though these neurogenic tumors are found in the head, neck and flexoral surfaces of the upper and lower limbs, their affection of nose and paranasal sinuses in the reported literatures have been far and few.[2] In this paper, neurofibroma arising from left middle meatus is described along with a review of literature.

CASE REPORT:

A 46 years old female from Kathmandu presented to us with left sided nasal obstruction for 4 months. It was insidious, gradually progressive, partial, continuous aggravated by upper respiratory tract infection. There was no history of excessive sneezing, nasal discharge, nasal bleeding or headache. There was no systemic history in the past.

On examination, she was of average built and well nourished. The osteocartilaginous framework of the nose appeared normal with decreased patency on the left side. Anterior rhinoscopy revealed a pinkish, polypoidal mass occupying left middle meatus. It was soft, non shrinkable, did not bleed on touch and probe could be passed superiorly, medially, inferiorly but not laterally. The contralateral nostril and posterior rhinoscopy were normal. There were no enlarged lymph nodes in the neck, and ear and throat examination were normal. Her systemic examination, preoperative investigations and chest radiograph was normal.

Computed tomography (CT) scan was suggestive of a soft tissue density mass obliterating the left nasal cavity (Fig-1). There was no bony erosion or antral soft tissue extension. She underwent endoscopic sinus surgery under general anaesthesia. Per operatively a pinkish, pedunculated, leafy mass was seen arising from the left middle meatus. With a straight forceps the mass was avulsed in toto. On cross section it was firm in consistency. Nasal cavity was packed with gel foam following surgery and the patient was sent home on second postoperative day.

Postoperatively, the histopathology report of the operated specimen showed nasal tissue with submucosal nodules composed of spindle cells with wavy nuclei and scanty cytoplasm separated by wavy collagen fibres and myxoid stroma. Mast cells were also seen. So, histopathological examination was considered to be neurofibroma (Fig-2& Fig-3). The patient has turned up twice for follow up in the OPD- once at one week following discharge and the other three months later and no recurrence has been seen.

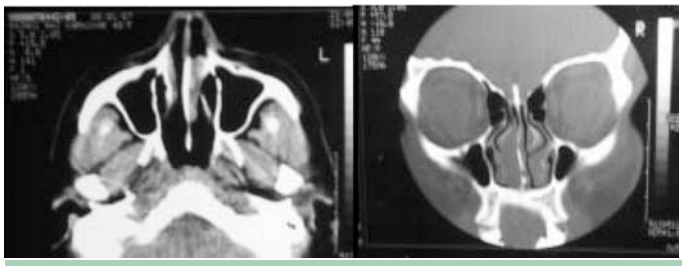

Fig:1. CT Scan axial and coronal cut showing homogeneous opacity in the left nasal cavity.

DISCUSSION:

Neurofibromas are benign, heterogeneous peripheral nerve sheath tumors that arise from the connective tissue of peripheral nerve sheaths, especially the endoneurium.[3] In the area of the nose and paranasal sinuses, neurofibroma arises from the first and second division of the trigeminal nerve and from autonomic plexuses.[1,2] The olfactory nerve can be excluded as a possible origin for these tumors, since the olfactory nerve contains no Schwann cells.[2]

They may occur as an isolated, sporadic lesions but are much more common in association with neurofibromatosis type[1.4] Solitary neurofi bromas of the maxillary sinus are exceedingly rare tumors.[4] Therefore, cases suspicious for neurosi broma or neurofibromatosis, i.e., with cafe-au-lait spots and nodules indicative of cutaneous neurofibroma, should be thoroughly examined to exclude the possibility of neurofi bromatosis.[5] In our case, the patient showed no skin pigmentation or nodule and was believed to have a solitary neurofibroma.

Depending on their location and size, neurosi bromas of the nose and paranasal sinuses may present with a variety of signs and non-specisi csymptoms, including nasal obstruction, epistaxis, rhinorrhoea, epiphora, anosmia, facial swelling, headache and serous otitis media.[2,4,6,7] Despite their indolent growth rate, neurofibromas can occasionally become very large, resulting in local bony destruction and intracranial extension. The tumours may distort tissues by pressure

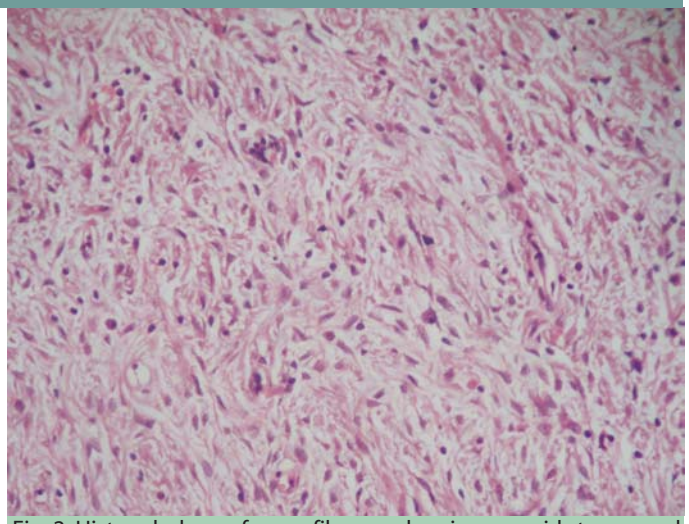

Fig: 2. Histopahology of neurofibroma showing myxoid stroma and spindle cells

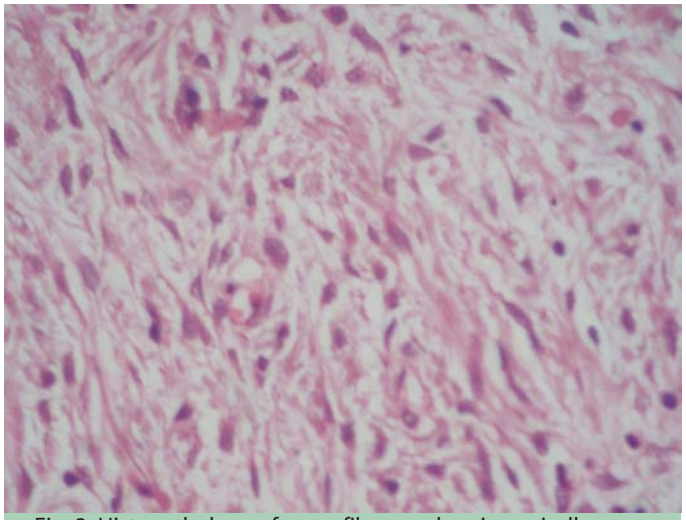

Fig: 3. Histopathology of neurofibroma showing spindle cells with wavy nuclei

or become symptomatic by obstruction of a sinus ostium.[8,9] Neurofibromas involving the sinonasal tract are predominantly solitary lesions. Their clinical presentation and imaging characteristics are not easily distinguishable from those of other sinonasal tumours. Naso-endoscopy may often add further information by identifying the origin of the tumour. CT scan is particularly important in the initial assessment in order to evaluate the origin, localisation and extension of the lesion.

Microscopically, neurofibromas are composed of a cellular proliferation of randomly arranged, spindle-shaped cells with fusiform or wavy, comma-shaped nuclei distributed on a background of a sibro-myxoid matrix, rich in mucopolysaccharides.[10,11] Immunohistochemically, neurofibromas show immunoreactivity for S-100 protein, NSE, and vimentin.[4] However in our case, immunohistochemistry could not be done due to lack of readily available facilities, financial constraint, lack of patient compliance and unwillingness on the part of the patient for the same.

Neurofibromas have to be differentiated from schwannomas and malignant peripheral nerve sheath tumors (MPNST). Schwannomas have a typical palisade pattern of the nuclei and tumor cell density is higher compared within neurofibromas, which usually show a mucoid extracellular matrix with only scattered tumor cells.[11] MPNSTs are characterized by hypercellularity and pleomorphic tumorcells and nuclei, features not presented by this tumor. Other spindle cell lesions such as fibrosarcoma, synovial sarcoma, and brous histiocytoma are uniformly S-100 protein-negative.[11]

Solitary neurofibroma should be closely followed up following surgery because the malignant transformation of solitary neurofibroma is also possible. Recurrence is rare, although neurofibroma recurs more often than schwannoma. But malignant transformation is reported to be at the rate of 10 per cent.[12] For this reason the patient has been kept on close follow up.

CONCLUSION:

Neurofibroma of the middle meatus is a rare disease. To the best of our knowledge, this case report is the first of its kind reported from Nepal.

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