The Journal of Laryngology & Otology, E44, 1 of 4.

doi:10.1017/S0022215106004440
Printed in the United Kingdom

Online Only Clinical Record

A peripheral nerve sheath tumour as a cause of nasal obstruction

A Manganaris, C Tsompanidou*, T Manganaris

Abstract

Neurogenic tumours form a very small percentage of all neoplastic lesions of the head and neck region. However, the head and neck region is by far the most common location for benign peripheral nerve tumours. Several cases involving the nose have been sporadically documented throughout the medical literature.

We present a rare case of a solitary neurofibroma arising from the lateral nasal wall of a 68-year-old woman. En bloc surgical resection of the mass was achieved by the lateral rhinotomy approach.

The clinical significance of this case report is due to its rare site. In recent medical literature, there has been only one report concerning a solitary neurofibroma arising from the inferior turbinate. This case also highlights the importance of considering this clinical entity in the differential diagnosis when encountering a unilateral soft tissue mass in the nasal cavity.

Key words: Neurofibroma, Nose; Lateral Rhinotomy

Introduction

Neurofibromas are benign, slow-growing, non-encapsulated tumours of neuroectodermal origin. They develop from the neural sheath of peripheral and cranial nerves as well as the nerve roots. Neurofibromas can occur in any part of the body, either as an isolated lesion or as part of the generalised syndrome of neurofibromatosis (von Recklinghausen disease). However, a solitary, localised neurofibroma will not usually be associated with neurofibromatosis. Common sites in the head and neck region include the oral cavity and tongue, the larynx, the face and orbit, the pharynx and oesophagus, and the nose and paranasal sinuses.[1–11] Neurofibromas arising in the sinonasal tract account for only a small proportion of the reported cases and are therefore considered extremely rare.

Depending on their location and size, neurofibromas of the nose and paranasal sinuses may present with a variety of signs and non-specific symptoms, including nasal obstruction, epistaxis, rhinorrhoea, epiphora, anosmia, facial swelling, headache and serous otitis media.[2–5] 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 or become symptomatic by obstruction of a sinus ostium.[12,13] Neurofibromas involving the face can even manifest clinically with facial or periorbital pain, proptosis or transient diplopia. Rapid clinical enlargement of neurofibromas is relatively uncommon but is nevertheless suggestive of possible malignant transformation.

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. Computed tomography (CT) scanning is particularly important in the initial assessment in order to evaluate the origin, localisation and extension of the lesion. Computed tomography of the paranasal sinuses is superior to magnetic resonance imaging (MRI) in demonstrating possible bony involvement. However, MRI is very useful in differentiating the tumour from secondary sinus disease.

The primary treatment modality for the management of neurofibromas of the nose is surgical excision.[14] The approach to the tumour is dictated primarily by its site and extension, as well as by the available surgical expertise and personal preference.

Case report

A 68-year-old woman presented suffering from progressive nasal obstruction for two years. No history of rhinorrhoea, epistaxis, anosmia, diplopia or epiphora, headache, or localised pain was given.

After admission, anterior rhinoscopy revealed a greyish, firm, smooth-surfaced and partially mobile mass occupying the right nasal cavity and pushing the septum to the opposite side. The left nasal cavity and the nasopharynx were free of tumour. There was no evidence of any associated lymphadenopathy, and the remainder of the head and neck examination was unremarkable.

Computed tomography revealed a well defined soft tissue mass in the right nasal cavity (Figure 1). The tumour had caused atrophy of the left middle and inferior turbinates. Obstruction of the sinus ostium had caused mucosal thickening in both the maxillary and ethmoidal sinuses. There was no evidence of bone destruction of the

From the Departments of ENT and *Histopathology, 'Theagenio' Anticancer Hospital, Thessaloniki, Greece.

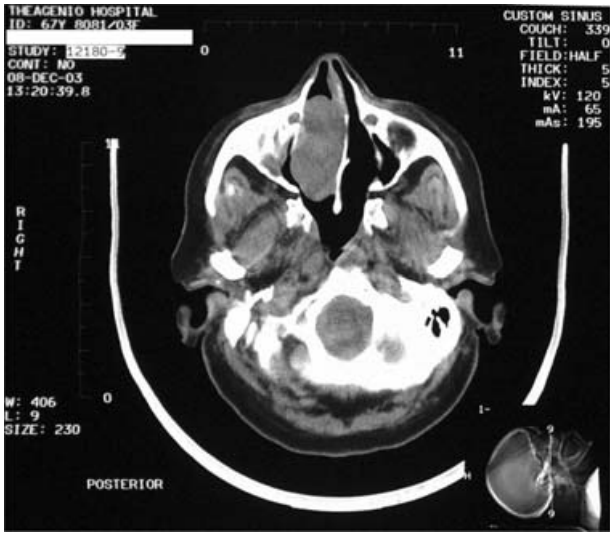

FIG. 1

Computed tomography scan showing a well defined soft tissue mass occupying the right nasal cavity and pushing the septum to the opposite side, and also mucosal thickening in the maxillary sinus.

medial wall of the maxillary sinus, the medial orbital wall or the anterior skull base. Both the history and the clinical and radiological findings were suggestive of a benign tumour, due to the benign course and lack of aggressive appearance on imaging.

The patient underwent en bloc tumour resection under general anaesthesia, through a right lateral rhinotomy incision (Figure 2). The tumour measured 4.5 × 3.5 × 0.8 cm and was found to originate from the medial surface of the right inferior turbinate.

Histological examination revealed that the tumour was well circumscribed, with partially irregular margins but definitely non-encapsulated. The tumour consisted of spindle-shaped cells that had elongated nuclei, with a wavy, serpentine configuration. The stroma contained a rich network of collagen fibres (Figure 3). Thin-walled blood vessels, numerous mast cells and mucinous changes were also present. No mitotic figures or necrosis were observed.

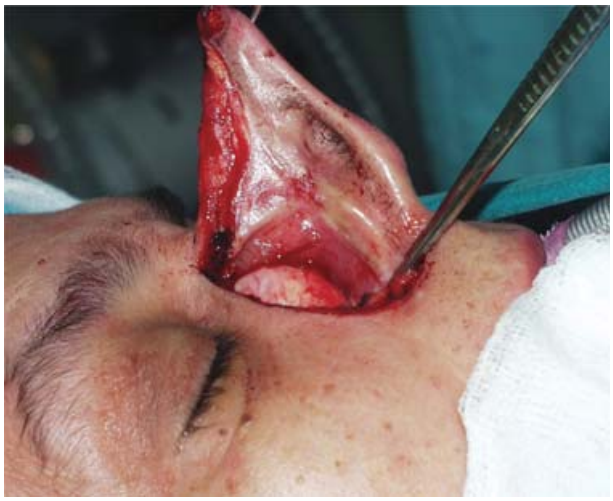

FIG. 2

Right lateral rhinotomy incision.

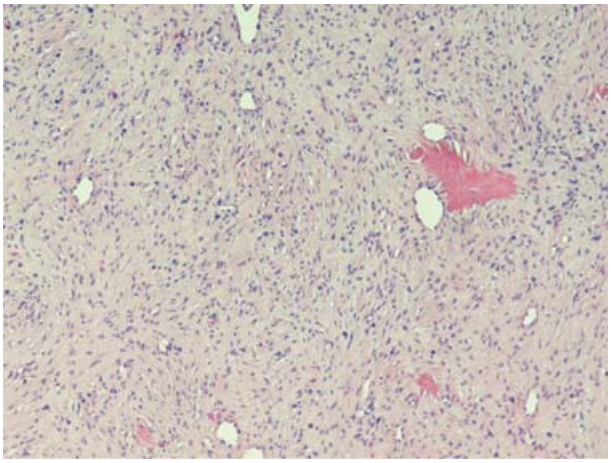

FIG. 3

Solitary neurofibroma showing serpentine configuration of the spindle-shaped cells, collagen fibres and blood vessels (H&E; ×100).

Immunohistochemical studies of the tumour cells revealed positivity to vimentin, N.S.E. (Neuron Specific Enolase) and S-100 protein (Figures 4 and 5). Smooth muscle actin, desmin, CD-34 and keratin E1/E3 were all negative.

At post-operative review, there were no stigmata of multiple neurofibromatosis. The final diagnosis was solitary neurofibroma arising from the right inferior turbinate. Follow up after 18 months showed no recurrence in the nose or any other complaints. The cosmetic result from the skin incision was excellent.

Discussion

In theory, any of the somatic or autonomic nerves supplying the nose and paranasal sinuses may give rise to a neurofibroma. Solitary neurofibromas may be well circumscribed lesions, or they may be diffuse with no apparent margins. The lesions can be papular, nodular or pedunculated and are usually greyish-white in colour.[13]

Solitary neurofibromas can present as either concentric or eccentric growths on the nerve of origin. They usually grow within the nerve sheath, encompassing the nerve.

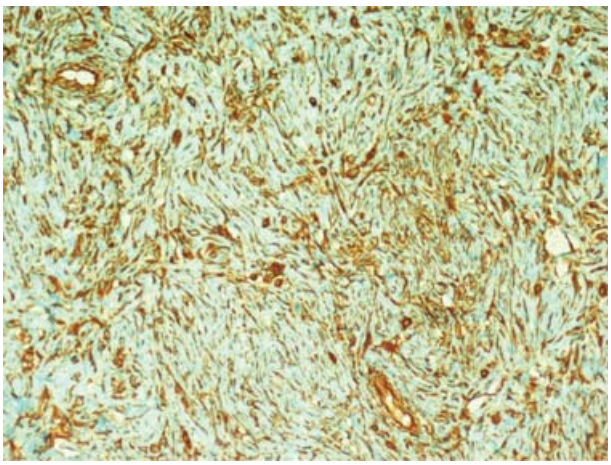

FIG. 4

Solitary neurofibroma showing positivity to vimentin (×200).

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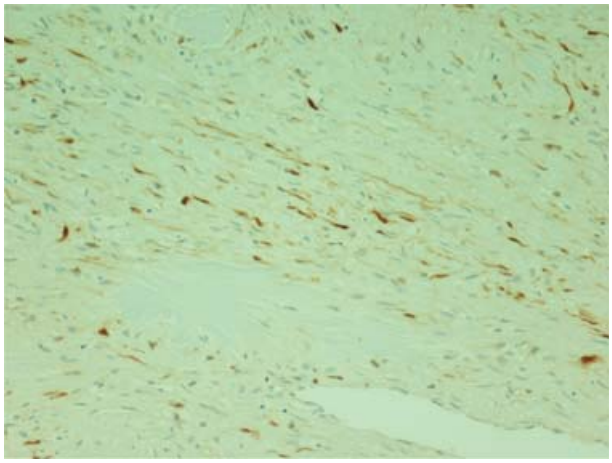

FIG. 5
Solitary neurofibroma showing S-100 protein positive immunohistochemical staining (×100).

Tumour proliferation may occur outside the perineurium, resulting in poor demarcation from the surrounding fibrovascular tissues, or it may be limited to within the perineurium, resulting in pseudo-encapsulation of the neural mass. Surgical identification of the particular nerve of origin is considered practically impossible due to their small size. Because of their growth characteristics, it is frequently impossible to completely excise neurofibromas without sacrificing the nerve of origin.[9,12,13]

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 fibro-myxoid matrix, rich in mucopolysaccharides.[11,15] Few, if any, Verocay bodies (so characteristic of the neurilemoma) are present. Tumour cells are not uniformly positive to S-100 protein, in contrast with neurilemomas (schwannomas), in which 100 per cent of the tumour cells are positive, signifying that they originate from neural crest derived tissue. The presence of axons within the neurofibroma can also help to distinguish it from a schwannoma, because axons are neurofilament positive.

Five different variants of solitary neurofibromas have been described:[16]

1 Myxoid neurofibroma: abundant mucin present in the matrix; S-100 immunohistochemical study is necessary to distinguish from myxoma.
2 Collagenous neurofibroma: thick collagen bundles present in matrix.
3 Epithelioid neurofibroma: tumour cells are rounded with eosinophilic cytoplasm.
4 Granular neurofibroma: cells contain granular, periodic acid-Schiff positive, diastase resistant cytoplasm.
5 Pigmented neurofibroma: scattered tumour cells contain melanin pigment; tumour cells are positive to S-100, Monoclonar Mouse Antihuman Melanosom (HMB45) and melan A.

Nuclear pleomorphism and mitotic activity is quite unusual in neurofibromas. However, degenerative nuclear pleomorphism may sometimes be present in atypical neurofibromas, thereby complicating their differentiation from malignant peripheral nerve sheath tumours. The presence of mitotic activity in neurofibromas is considered indicative of malignancy.[16]

A neurofibroma may often be diagnostically confused with a fibroma, neuro-fibrosarcoma, myxoma, haemangioma, lymphangioma, solitary fibrous tumour, dermoid or epidermoid cyst, or benign schwannoma.[11,12,16] In our case, the differential diagnosis included fibroma, neurofibroma and benign schwannoma.

Conclusion

Neurofibroma is considered a relatively common, benign peripheral nerve tumour. However, a thorough search of the English literature revealed only four well documented cases of solitary neurofibroma arising in the sinonasal tract, confirming this as a rather rare clinical entity.[2–5]

Immunohistochemistry and electron microscopy are used to confirm the initial diagnosis made by histological findings. S-100 protein, antiglial fibrillary acidic protein, vimentin and cytokeratin are routinely employed to exclude malignant schwannoma and to differentiate neurofibromas from other tumours.

The management of neurofibromas is based upon their symptoms. Excision of the mass is warranted in cases in which there is pain, cosmetic problems, progressive neurological deterioration, compression of adjacent tissues or loss of function, as well as suspicion of malignant degeneration.

The present consensus on the management of solitary neurofibromas not associated with a generalised syndrome dictates complete surgical excision. This is considered curative and offers an excellent prognosis. The malignant transformation potential of this tumour, when not associated with a syndrome, is considered minimal. Solitary neurofibromas of the sinonasal tract may also be excised via endoscopic techniques, always taking into consideration the possibilities of malignant transformation and local recurrence of incompletely excised lesions.

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https://doi.org/10.1017/S0022215106004440 Published online by Cambridge University Press

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Address for correspondence:
Dr Argyrios Manganaris,
17 Mitropolitou Iossif Str,
54622 Thessaloniki,
Greece.

E-mail: amanganaris@hotmail.com

Dr A Manganaris takes responsibility for the integrity of the content of the paper.
Competing interests: None declared