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CASE REPORT

Nasal Neurilemmoma in a Patient with Neurofibromatosis 2

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ABSTRACT

Neurilemmoma (schwannoma) is a benign peripheral nerve sheath tumour that rarely occurs in the nasal cavity. Lesions that present in the paranasal sinuses and nasal cavity account for approximately 4% of head and neck neurilemmomas. In patients with a known history of neurofibromatosis, computed tomography and magnetic resonance imaging can give a clue to this diagnosis, which requires confirmation by histology. This report is of a patient with nasal neurilemmoma and neurofibromatosis 2.

Key Words: *Neurilemmoma; Neurofibromatoses; Neurofibromatosis 2; Paranasal sinus neoplasms*

INTRODUCTION

Neurilemmomas are benign peripheral nerve sheath tumours that occur throughout the body. They typically present in the third to sixth decades of life. The estimated incidence is 1 per 3000 population and the condition is 2 to 4 times more common in women than in men.[1]

Neurofibromatosis 2 (NF2) is a rare autosomal dominant disorder, with an incidence of 1 per 37,000 population.[2] The hallmark of the condition is the presence of bilateral vestibular schwannomas. Patients with NF2 also develop other cranial, spinal, and peripheral schwannomas, cranial and spinal meningiomas, and cataracts.[3]

Solitary neurilemmoma in the nasal cavity is rare. Only 63 patients with this condition have been reported in the English language literature since January 1943.[4-24] This report is of a patient with neurilemmoma of the nasal cavity and NF2.

CASE REPORT

A 44-year-old woman with a history of NF2 presented in 2007 with right epistaxis. Examination by an ear, nose, and throat surgeon showed a polyp-like mass in the middle meatus of the right nasal cavity.

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Magnetic resonance imaging (MRI) was performed. T1-weighted post-contrast images showed enhancing extra-axial masses over the cerebral convexity and enhancing masses in the cerebello-pontine angles, with extension into the internal acoustic meati, consistent with multiple meningiomas and bilateral acoustic schwannomas (Figure 1).

T1-weighted images showed a hypointense right nasal mass (Figure 2a). T2-weighted images showed a slightly heterogeneous hyperintense mass in the right nasal cavity (Figure 2b). T1-weighted post-contrast images showed a heterogeneously enhancing mass (Figure 2c).

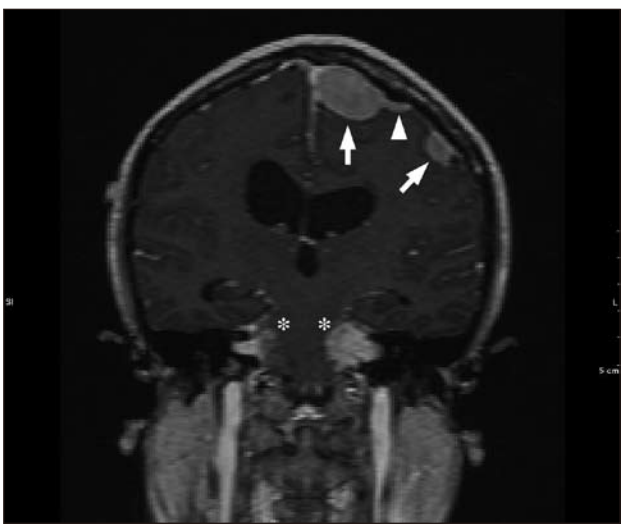

Figure 1. Typical magnetic resonance imaging features of neurofibromatosis 2. T1-weighted post-contrast coronal image showing multiple meningiomas (white arrows) with 'dural tail sign' (white arrowhead) and bilateral acoustic schwannomas (asterisks).

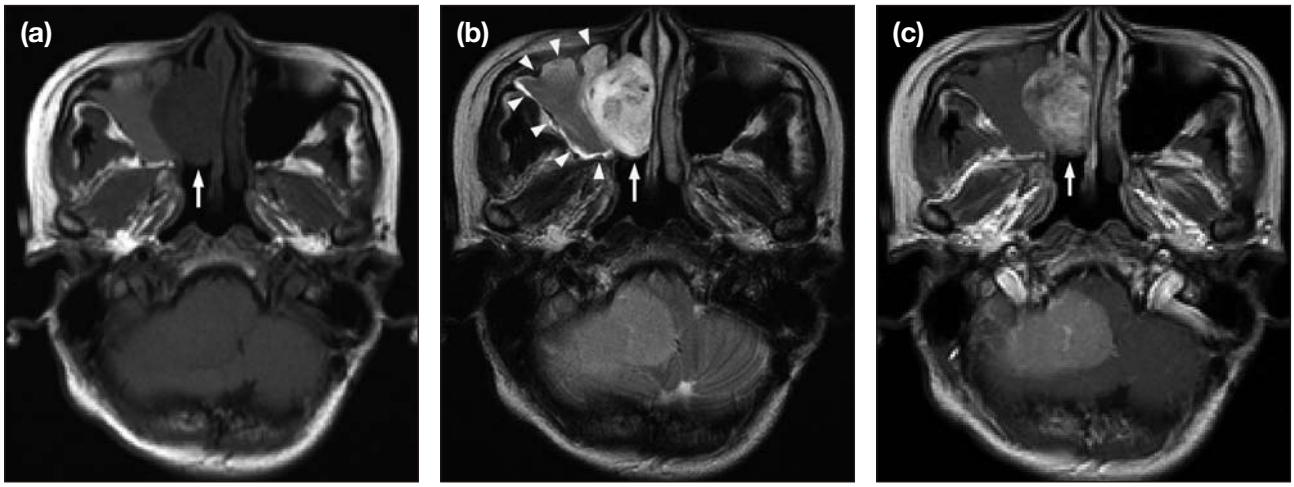

Figure 2. Magnetic resonance imaging features of the right nasal mass. (a) T1-weighted image showing a hypointense right nasal mass (arrow); (b) T2-weighted image showing a slightly heterogeneous hyperintense mass in the right nasal cavity (arrow) and an opacified right maxillary sinus (arrowheads); and (c) T1-weighted post-contrast image showing the corresponding right nasal mass with heterogeneous contrast enhancement (arrow).

Computed tomography (CT) without intravenous contrast of the paranasal sinuses (in the bone window) showed a corresponding well-defined mass with smooth expansion of the adjacent bony sinus wall and nasal septum (Figure 3). Radiological features were suggestive of NF2 with a non-aggressive right nasal tumour. Neurilemmoma was the primary diagnostic consideration given the patient's history of NF2.

Functional endoscopic sinus surgery was performed to resect the right nasal tumour. Pathology showed a mass with a polypoid outline with spindle cell proliferation beneath the respiratory epithelium. The tumour cells showed wavy outlines. There were dense and loose areas, known as Antoni type A and B areas, characteristic of neurilemmoma (Figure 4a). The dense area showed palisading of the nuclei, also characteristic of neurilemmoma (Figure 4b).

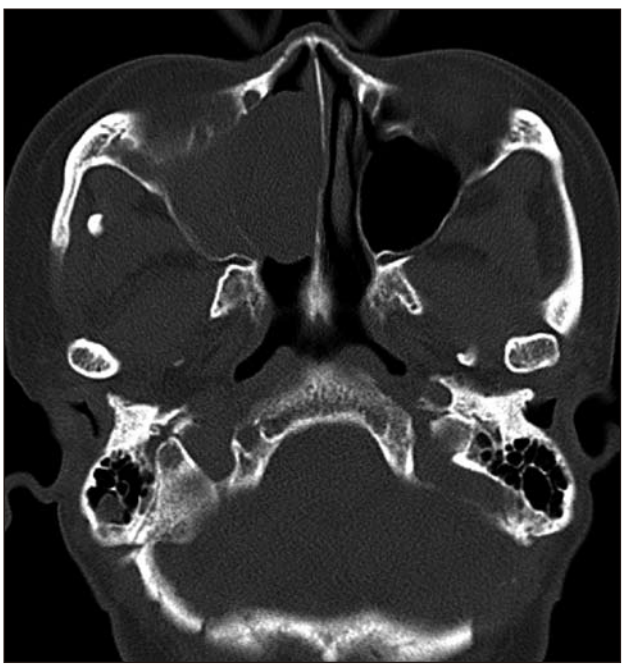

Figure 3. Computed tomography image without intravenous contrast of the paranasal sinus (in the bone window) showing a corresponding well-defined mass with smooth expansion of the adjacent bony sinus wall and nasal septum.

DISCUSSION

Neurilemmomas, or schwannomas, are benign neurogenic tumours that arise from the nerve sheaths of the peripheral nerves. Neurilemmomas may occur anywhere in the body but have a predilection for the head, neck, and flexor surfaces of the upper and lower extremities. Deeply situated tumours predominate in the retroperitoneum and posterior mediastinum.[25] Patients with NF2 often develop cranial, spinal, and peripheral schwannomas.

Solitary nasal neurilemmoma is rare. The symptoms are non-specific and are the result of the mass effect or tumour necrosis. Patients may present with nasal obstruction, rhinorrhoea, or recurrent epistaxis, as for this patient. Facial swelling and pain are associated with paranasal sinus involvement.[11] These lesions rarely undergo malignant transformation.[26]

Microscopically, neurilemmomas can exhibit 2 architectural patterns, Antoni type A and Antoni type B, in different proportions. Antoni type A tissue is composed of an organised compact cellular stroma with elongated spindle cells. Parallel rows of palisading nuclei can be

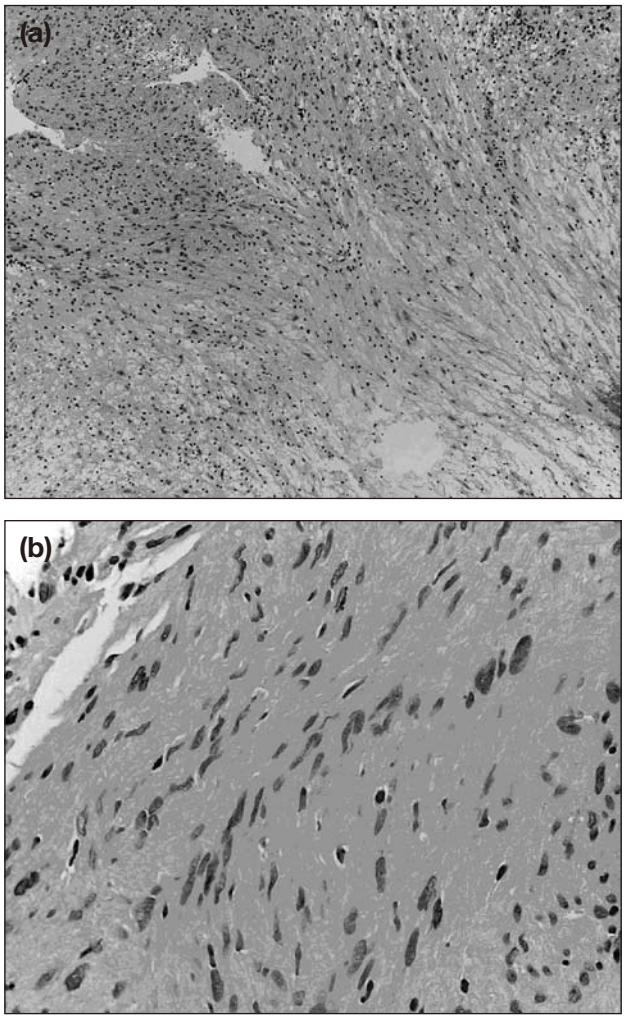

Figure 4. Photomicrographs showing histological features characteristic of neurilemmoma. (a) Dense and loose areas characteristic of neurilemmoma; and (b) the dense area showing palisading of the nuclei (haematoxylin and eosin stain; original magnification, x 60).

seen in this highly differentiated tissue. Antoni type B tissue is composed of disorganised loose myxoid stroma with few spindle cells.

Neurilemmomas are slow-growing lesions with a tendency to cause bony expansion rather than the bony destruction seen with malignant lesions.[27] CT is excellent for demonstrating the fine bony details.

MRI findings for neurilemmoma have been described as masses with low-signal intensity on T1-weighted images and inhomogeneous high-signal intensity on T2-weighted images due to alternating Antoni type A and B areas and secondary degenerative changes. Contrast-enhanced T1-weighted MRI can clearly depict cystic necrotic areas and well-enhanced peripheral and intervening solid areas of the mass. These patterns of signal intensity are not specific to neural tumours, nor do they allow differentiation between benign and malignant nerve sheath tumours.

Although only 63 patients with solitary neurilemmoma in the nasal cavity have been reported in the English language literature since January 1943,[4-24] the nasal septum appears to be the most common location for this lesion. The main differential diagnoses of nasal masses with similar imaging findings as neurilemmoma include nasal polyp, malignant nerve sheath tumours, fibromyxoma, and sarcoma. Nasal polyps represent hyperplasia of the mucosa in response to chronic inflammation, usually from chronic sinusitis. Nasal polyps usually have high signal intensity on T2-weighted images, which helps to distinguish them from tumours. Fibromyxoma is a rare slow-growing tumour originating from fibroblasts. This tumour can manifest as a well-marginated mass but may also show local infiltration and local invasion.[28] In patients with a known history of NF, a nasal mass with such imaging findings provides a clue for this diagnosis, which requires confirmation by histology.

The preferred treatment for neurilemmoma is complete surgical excision as neurilemmomas are generally radioresistant.[29] Functional and cosmetic considerations are important for the surgical planning because of the non-invasive and slow-growing nature of this tumour.

Nasal neurilemmoma is a rare benign peripheral nerve sheath tumour. This report describes a patient with NF2 showing concordant radiological-pathological correlation.

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