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Solitary neurofibroma of the nasal cavity: resection with endoscopic surgery

Mitsuaki Hirao*, Takashi Gushiken*, Hideki Imokawa†, Satoshi Kawai‡, Hayashi Inaba‡, Mamoru Tsukuda‡

Abstract

We present a case of neurofibroma of the nasal cavity treated by endoscopic surgery. A 71-year-old female had complained of left-sided nasal obstruction for the past four years. Anterior rhinoscopy, computed tomography (CT) and magnetic resonance imaging (MRI) revealed a tumour involving the left nasal cavity. Histological and immunohistochemical examination showed the tumour to be a neurofibroma. The tumour was resected with endoscopic surgery. Neurofibroma arising in the area of the nose and paranasal sinuses is rare. We discuss the clinical and pathological characters of neurofibroma arising in the nasal cavity.

Key words: Nasal Cavity; Neurofibroma; Immunohistochemistry; Endoscopsy; Surgical Procedures, Operative

Introduction

Peripheral nerve sheath tumours are divided into schwannoma, neurofibroma, and neurogenic sarcoma. They rarely arise in the area of the nose and paranasal sinuses,[1] especially neurofibroma. We present a case of neurofibroma of the nasal cavity treated by endoscopic surgery.

Case report

A 71-year-old female had complained of left-sided nasal obstruction for the past four years. She was taking oestriol, since she had a past history of cancer of the uterus and had undergone an operation. But there was no other relevant past history, no areas of skin pigmentation abnormalities, and no family history of neural tumours.

Anterior rhinoscopy revealed a whitish, firm, and haemorrhagic polypoid mass (Figure 1). Water's and Caldwell's view showed a diffuse shadow involving the left maxillary sinus, ethmoidal sinus, and frontal sinus. Contrast-enhanced computed tomography (CT) showed an irregularly enhanced high density region filling the left nasal cavity and displacing the nasal septum to the right. The lateral wall of the left nasal cavity was pushed laterally. In addition, it showed uniformly high-density regions occupying the left maxillary sinus, ethmoid sinus, and frontal sinus without apparent bone destruction (Figure 2). On magnetic resonance imaging (MRI), the left nasal cavity and the left paranasal sinuses showed low intensity in T1-weighted and high intensity in T2-weighted, but the enhancement was more irregular and the intensity was lower in the nasal cavity than in the paransal sinuses. From these findings we suspected a tumour arising from the left nasal cavity (Figure 3).

A biopsy was taken for pathological study. Histological and immunohistochemical examinations showed the tumour to be a neurofibroma. A total resection of the tumour was performed with endoscopic surgery. The tumour was whitish-gray, solid, and so large as to be attached to the lateral side of the nasal cavity, the inferior nasal turbinate, and the choana, but no adhesion was seen. The stem of the tumour was at the nasal septum and it was cauterized and ablated from the nasal septum by using a Nd-YAG laser and a monopolar cautery. *En bloc* resection of the tumour was attempted but as the tumour was strangulated at the choana, it was removed piecemeal. After the extraction, the left ethmoid bulla and the left maxillary sinus were visualized via the middle nasal meatus and the left sphenoid sinus and the left frontal sinus were visualized via the superior nasal meatus. Mucous fluid was found in each paranasal sinus. From these findings, we

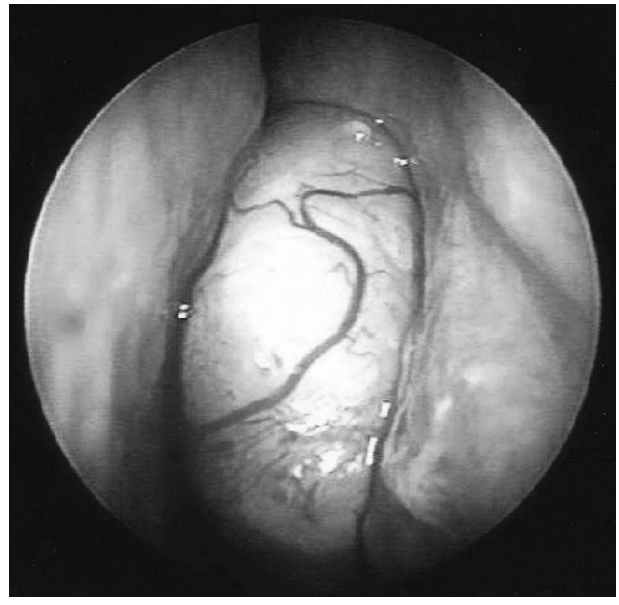

Fig. 1

Anterior rhinoscopy revealed a whitish, firm, haemorrhagic polypoid mass.

From the Ohfuna Central Hospital, Imokawa Otorhinolaryngology Clinic† and the Department of Otorhinolaryngology‡, Yokohama City University School of Medicine, Japan.
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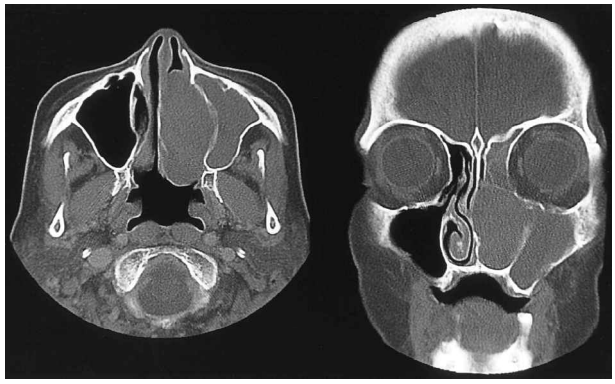

FIG. 2

Contrast-enhanced CT revealed lesions in the left nasal cavity and paranasal sinuses. The lesion of the nasal cavity showed an expansive growth without bone destruction.

drew the conclusion that the tumour was localized to the left nasal cavity and that it had caused secondary paranasal sinusitis. We were unable to demonstrate the nerve of origin which gave rise to the tumour.

On macroscopic examination of the resected specimen, the tumour had a quite thin fibrous capsule and the cut surface was yellowish-gray. Microscopic examination revealed interlacing bundles of spindle cells. These cells had hyperchromatic serpentine nuclei. A few of them had large and slightly atypical nuclei, but no nuclear mitoses (Figure 4). Immunohistochemical study revealed that the cells of the tumour were immunoreactive for S100 protein, neuron specific enolase (NSE), and vimentin, but not for desmin, or smooth muscle actin (Table I). From these clinical and histopathological findings the tumour was diagnosed as a neurofibroma.

The operation has lead to a relief of symptoms, secondary paranasal sinusitis has been improved. Now after the follow-up period of 18 months, neither post-operative complications nor signs of recurrence have been observed.

Discussion

Peripheral nerve sheath tumours are classified into benign and malignant. The former include schwannoma and neurofibroma, and the latter comprises neurogenic sarcoma. Furthermore, neurofibroma is divided into solitary tumours and multiple tumours which include neurofibromatosis I and II. Schwann cells and perineural cells derived from neuroectoderm are thought to be the origin of all these neoplasms.[2,3] In the area of the nose and paranasal

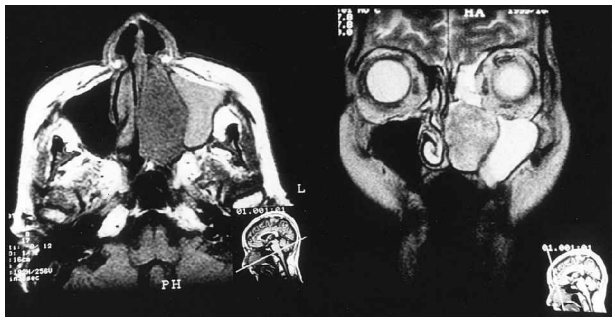

FIG. 3

MRI image. The left side is axial T1-weighted image and the right side is coronal T2-weighted image. The intensity is different between the nasal cavity and the paranasal sinuses.

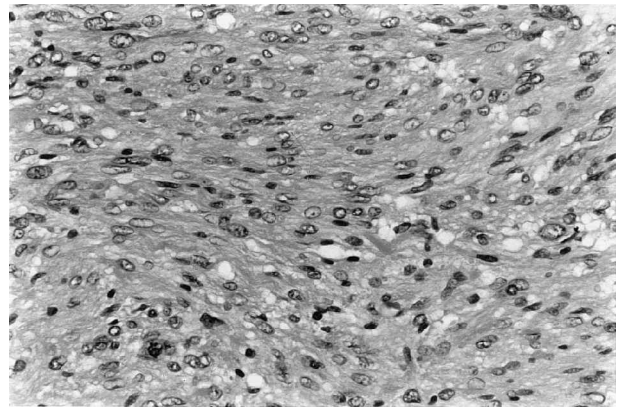

FIG. 4

Interlacing bundles of spindle cells without nuclear mitosis (H&E; ×100)

sinuses, neurofibroma arises from the first and second division of the trigeminal nerve and from autonomic plexuses.[2,4] These tumours are extremely uncommon in this area.

Neurofibroma in the area of the nose and paranasal sinuses, as other neoplasms in this area, leads to non-specific symptoms including nasal obstruction, epistaxis, cheek swelling and pain, and proptosis. Thus the histological examination is essential to the diagnosis.

Neurofibromas may be difficult to differentiate from other non-epithelial tumours especially in small biopsy or curretage specimens because the histological appearances tend to be non-specific.[5] Adequate biopsy or specimens sufficient for macroscopic and microscopic examination, and immunohistochemical studies are essential for exact diagnosis. Macroscopically neurofibromas have no capsule, and their cut surfaces are regular without secondary degenerative changes. Histologically, neurofibromas consist of interlacing bundles of elongated cells with wavy and dark-stained nuclei, wire-like strands of collagen, and myxoid stroma dotted with occasional mast cells and lymphocytes. Immunohistochemically, neurofibromas show immunoreactivity for S-100 protein, NSE, and vimentin. All these characteristics lead to the exact diagnosis of neurofibroma.

The treatment of neurofibroma is complete resection of the tumour because neurofibromas may infiltrate extensively. Other treatments are not effective. In more extensive cases, the extensive operation should be performed. Recurrence is rare, although neurofibroma recurs more after than schwannoma. But malignant transformation is reported to be at the rate of 10 per cent.[6] In our case, as CT and MRI revealed the tumour localized in the left nasal cavity, we attempted tumour resection under endoscopy. Fortunately, the tumour was not adherent and we could visualize the origin of the tumour at the nasal septum, and this allowed macroscopic complete resection under endoscopy. Our only worry was the possibiity of residual tumour at the nasal septum. Because of this we cauterized and ablated the stalk of the tumour down to the cartilage of the nasal septum. We were therefore unable to examine the histology of the deep margins of resection.

TABLE I

THE RESULT OF IMMUNOHISTOCHEMICAL EXAMINATION

Positive reaction for:	S-100 protein, NSE, vimentin
Negative reaction for:	desmin, smooth muscle actin

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In the literature, however, there are few reports of endoscopic surgery for total resection of neurofibroma in the nasal cavity. Yong reported a case in which a solitary neurofibroma arising from the nasal septum was resected under endoscopy and no signs of recurrence were observed after 12 months' follow-up.[7] Pablo reported a case in which a solitary neurofibroma arising from the inferior turbinate was resected with the inferior turbinate under endoscopy and no signs of recurrence were observed after two years follow-up.[4] Both of them identified the origin of the tumour under endoscopy but more details of the operation were not reported.

In conclusion, transnasal endoscopic resection is useful if the neurofibroma is solitary, small, and localized in the nasal cavity and the origin can be identified.

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Address for correspondence:
Mitsuaki Hirao,
Ohfuna Central Hospital,
6-2-24 Ohfuna,
Kamakura-shi,
Kanagawa Prefecture,
Japan.

Dr M. Hirao takes responsibility for the integrity of the content of the paper.
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