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Malignant schwannoma arising in a paranasal sinus

IKUO NAGAYAMA, M.D., TOSHIRO NISHIMURA, M.D., MITSURU FURUKAWA, M.D. (Kanazawa, Japan)

Abstract
A case of malignant schwannoma arising in a paranasal sinus is reported. In this case, histological hallmarks were lost because of the poorly differentiated nature of the tumour. Immunohistological techniques were applied to the diagnosis, confirming the neural origin of this tumour. Malignant schwannoma is a relatively rare disease in the nasal cavity and paranasal sinuses. For the treatment of this tumour, wide resection is recommended. In this patient, radical resection of the maxilla with orbital and ethmoid exenteration was performed. The patient recovered uneventfully with no evidence of disease three years after surgery.

Key words: Paranasal sinus neoplasms, malignant schwannoma; Immunohistochemistry.

Introduction

The definitive diagnosis of malignant schwannoma is based on the coexistence of a tumour with a nerve fibre or ultrastructural features showing elaborate, branched cytoplasmic processes covered by basal laminae. When these typical features cannot be confirmed because of the undifferentiated and highly pleomorphic nature of the tumour, immunohistological examination is a useful modality. Immunohistochemistry with S-100, antiglial fibrillary acidic protein (GFAP), vimetin and cytokeratin are employed to confirm malignant schwannoma, and differentiate it from other tumours. Malignant schwannoma arising in a paranasal sinus is a quite rare clinical entity. A search of the literature revealed only 11 previously reported cases. The treatment of this aggressive disease is wide resection. Local excision will result in a poor prognosis.

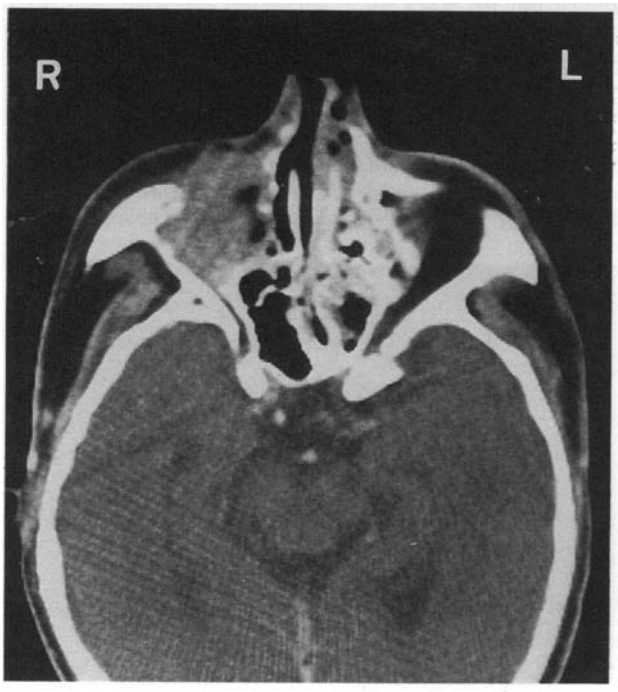

FIG. 1
Axial CT scan showing the lesion arising from the maxillary sinus and extending into the ethmoid sinus with destruction of the orbital wall.

Case report

A 60-year-old man was referred to our hospital because of right facial swelling and numbness. He first noted the swelling two months earlier and was concerned because it had been steadily enlarging, causing local facial paresthesia. He denied any history of epistaxis, mucopurulent rhinorrhea or hyposmia. He had been healthy for most of his life, and the family history was negative for neurofibromatosis.

On physical examination diffuse swelling of the right anterior antral wall and upward deviation of the eyeball were marked. No significant findings were observed in the nasal cavities, and there was no involvement of the lymph nodes. No skin lesions or café au lait spots were identified.

Computed tomography revealed the main tumour to arise in the antrum and extend into the ethmoid sinus (Fig. 1). Bone erosion and partial destruction of the orbital floor were demonstrated. The patient underwent excisional biopsy through a right

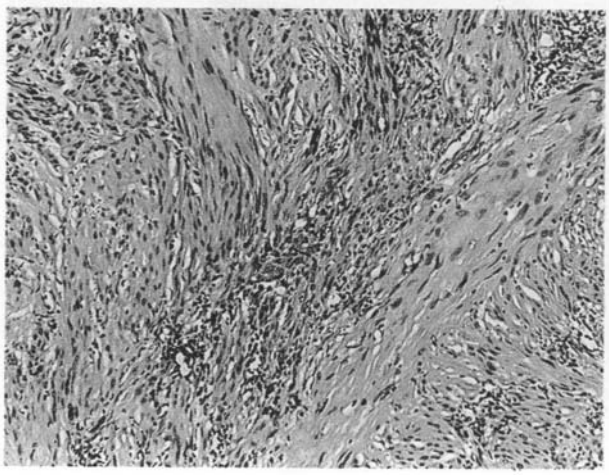

FIG. 2
Myxomatous stroma and perineural extension including fascicles and nodule of spindle-shaped tumour cells. H&E ×100.

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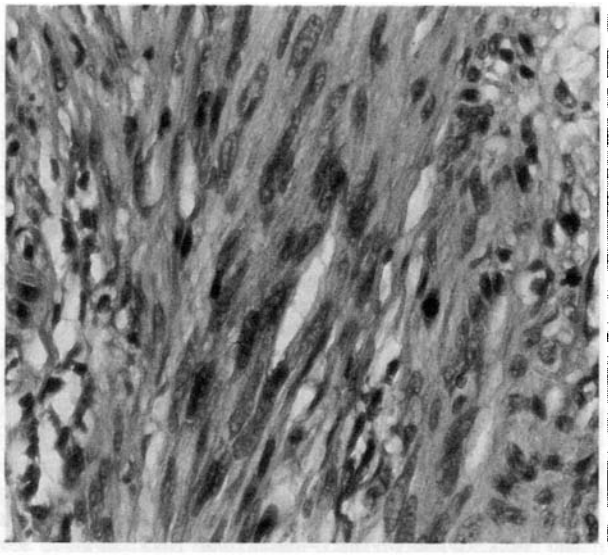

FIG. 3
Nuclei showing various degrees of pleomorphism and irregularity in shape ×400.

exploratory antrostomy. Haematoxylin and eosin staining revealed fascicles and nodules of spindle-shaped tumour cells with indistinct cytoplasmic borders. A myxomatous stroma and perineural extension were also observed (Fig. 2). Nuclei exhibited various degrees of pleomorphism and shape irregularity, unlike fibrosarcoma (Fig. 3). Electron microscopic and immunohistochemical studies were performed to elucidate the true origin of the tumour.

Immunohistopathological examination

Immunohistochemical specimens for S-100 protein, vimetin, cytokeratins and GFAP were prepared according to Hsu. Under microscopic observation, the following results were obtained. Immunohistopathological examination using the avidin-biotin-peroxidase complex method was positive for S-100 protein and vimetin (Fig. 4). In this slide, not only tumour cells but also nerve cells are positive for S-100 protein. Cytokeratin, Nos. 8, 10, 13, 16, 18 and 19 were negative. Desmin and GFAP were also negative. Desmin-positive cells represent muscle cells, but there were no positive cells in the tumour. These results were suggestive of malignant schwannoma.

An ultrastructural study, performed with previously formalin-fixed material, did not clearly demonstrate the cytoplasmic features because of the inadequacy of the specimens and the poorly differentiated nature of the tumour.

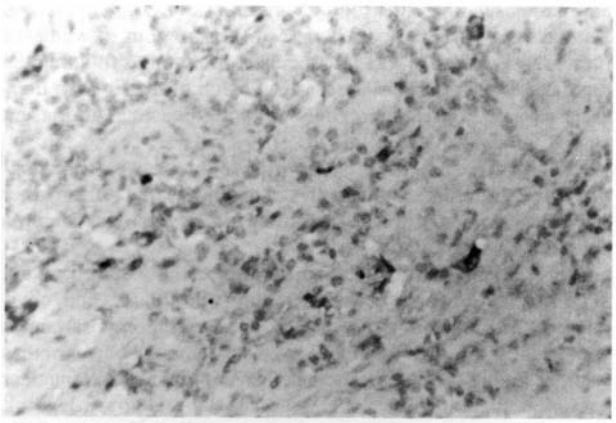

FIG. 4
S-100 protein in the tumour cells.

Clinical course and treatment

Because of its high malignancy, one cycle of neo-adjuvant chemotherapy using vincristin, adriamycin and cyclophosphamide (Goldman *et al.*, 1977) and 70 Gy of irradiation with superficial arterial infusion of 5FU were administered. The effect of combined therapy was poor. Subsequently, a right total maxillectomy with orbital exenteration was planned, and wide *en bloc* resection was successfully conducted. The tumour was found to extend into the right maxillary sinus and orbit and to grossly involve the hard palate. A major nerve trunk from which the tumour might have arisen was not detected. The post-operative course was uneventful. At present, the patient is alive with no evidence of disease three years after operation.

Discussion

Malignant schwannoma is uncommon, accounting for no more than 5 to 10 per cent of all sarcomas (Russel *et al.*, 1977; Hoffman *et al.*, 1988). Less than 10 per cent of all cases of malignant schwannoma are found in the head and neck. In a series from the Massachusetts General Hospital, 607 patients with neurilemmomas or neurofibromas were seen from 1962 to 1979 (Trojanowski *et al.*, 1980). During the same period there were only 24 patients with malignant schwannoma. In the nasal cavity

TABLE I
TUMOUR ARISING IN NASAL CAVITY OR PARANASAL SINUS

Year	Author	No.	Head and neck	Nasal cavity paranasal sinus	Treatment	Outcome
1951	Vieta	31	8	1	—	—
1970	White	15	2	1	Local excision	DOD 2 months
1977	Goepfert		7	2	Resection	NED 2.5 yr
					Radiation	
					Chemotherapy	
					Resection	NED 3 yr
					Radiation	
1985	Gullane		3	1	Subtotal maxillectomy	DOD 2 yr
1988	Hoffman		9	1	Craniofacial surgery	NED 11 yr
1989	Marvel		1	1	Craniofacial surgery	—
1991	Bailet		16	4	Radiation	Dead 3yr, NED
					Local excision	DOD 15.5 yr
					Local excision	DOD 5 months
					—	NED 18 yr
1992	Author		1	1	Extended	NED 3 yr
					Total maxillectomy	

DOD = Die of disease; NED = No evidence of disease.

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and paranasal sinus this tumour is quite rare. Das Guputa and Brasfield (1970) reported 232 cases of this tumour of which 18 tumours were in the head and neck with no tumour found in the nasal cavity or paranasal sinuses. A search of the English literature revealed only 11 previously reported cases. The present case is the twelfth case (Table I).

Usually, the malignant nature of this tumour is easily recognized and the major challenge lies in distinguishing it from other sarcomas such as fibrosarcoma, monophasic synovial sarcoma and leiomyosarcoma.

Fibrosarcomas have a more uniform fasciclar pattern, contain symmetrical fusiform cells resembling fibroblasts, and obviously lack features of neural differentiation. Leiomyosarcomas can usually be differentiated from malignant schwannomas without undue difficulty. Their cells have distinct juxtanuclear vacuoles. In ambiguous situations special stains are helpful, since the cytoplasm of leiomyosarcoma cells is fuchsinophilic with longitudinal striations (Masson trichrome) and contains at least moderate amounts of glycogen (Periodic acid-Schiff). On the other hand, the cytoplasm of malignant schwannoma is usually less fuchsinophilic (with no longitudinal striations) and contains little or no glycogen. Immunoreactive cytokeratin can be detected within a significant number of monophasic synovial sarcomas, while S-100 protein is rarely identified.

On the other hand, immunoreactive S-100 protein is present in 50 per cent to 90 per cent of malignant schwannomas, while cytokeratin and desmin are rarely identified (Nakajima *et al.*, 1982; Stefansson *et al.*, 1982). Although the resected specimen did not meet the requirement of arising in and being largely confined to a nerve, S-100 protein identification suggested it to be of nerve origin.

Malignant schwannoma occurs in isolation or secondary to von Recklinghausen's disease (Ghosh *et al.*, 1973; Sordillo *et al.*, 1981; Hoffman *et al.*, 1988). Von Recklinghausen's disease is present in 30 per cent to 40 per cent of patients with this tumour (Ghosh *et al.*, 1973; Sordillo *et al.*, 1981) while according to Sordillo *et al.* 8.5 per cent of patients have a history of radiation therapy to the affected region. All age groups are affected with the peak incidence found in the fifth decade. Ghosh *et al.* and White (1971) have both described a lower survival rate in patients associated with von Recklinghausen's disease.

The best curative therapy is wide *en bloc* excision (Das Guputa and Brasfield, 1970; White, 1971). From the Table it is clear that local excision results in a poor prognosis, so wide resection procedure including craniofacial surgery should be chosen.

Pulmonary metastases develop during life in 40 per cent of patients (Sordillo *et al.*, 1981) but are found in as many as 90 per cent at autopsy (Das Guputa and Brasfield, 1970).

Metastases to the brain and lymph nodes are notably rare, making prophylactic treatment of the lymph nodes unnecessary. The roles of radiotherapy and chemotherapy are still controversial. Malignant schwannoma has traditionally been considered radio-resistant (Hutcherson *et al.*, 1979; Marvel *et al.*, 1990), but some authors have described the usefulness of postoperative radiation combined with chemotherapy (Goepfert *et al.*, 1977; Hoffman *et al.*, 1988).

In the treatment of this often fatal disease, wide resection is the treatment of choice, but combined therapy may also have a role.

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Address for correspondence:
Ikuo Nagayama , M. D.,
Department of Otolaryngology,
School of Medicine,
Kanazawa University,
13-1 Takaramachi,
Kanazawashi, 920,
Ishikawaken, Japan.