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Myxofibrosarcoma of the sphenoid sinus


Abstract
Myxofibrosarcoma was originally described as the myxoid variant of malignant fibrous histiocytoma (MFH). It is one of the commonest sarcomas of the extremities and retroperitoneum in late adult life. The head and neck region, however, is an uncommon site with three to 10 per cent involvement. Primary tumour sites include the larynx, paranasal sinuses, oral cavity, salivary glands, face and scalp. Thirty per cent of cases were in the sinonasal tract, most commonly in the maxillary and ethmoid sinuses, whereas the sphenoid sinus is rarely involved. There are no specific clinical features of this particular tumour type, although its mass effect may occasionally be noted. Magnetic resonance imaging (MRI) and computerized tomographic (CT) scan may show malignant features such as local tissue invasion. Histopathological examination is, however, the gold standard in making a definitive diagnosis. Complete tumour resection with adequate resection margins remains the mainstay of treatment, but the roles of radiotherapy and chemotherapy are debatable.

Introduction
Myxofibrosarcoma was originally described as the myxoid variant of malignant fibrous histiocytoma (MFH). It is one of the commonest sarcomas of the extremities and retroperitoneum in late adult life. The head and neck region, however, is an uncommon site with three to 10 per cent involvement. Primary tumour sites include the larynx, paranasal sinuses, oral cavity, salivary glands, face and scalp. Thirty per cent of cases were in the sinonasal tract, most commonly in the maxillary and ethmoid sinuses, whereas the sphenoid sinus is rarely involved. There are no specific clinical features of this particular tumour type, although its mass effect may occasionally be noted. Magnetic resonance imaging (MRI) and computerized tomographic (CT) scan may show malignant features such as local tissue invasion. Histopathological examination is, however, the gold standard in making a definitive diagnosis. Complete tumour resection with adequate resection margins remains the mainstay of treatment, but the roles of radiotherapy and chemotherapy are debatable.

Case report
A 55-year-old Chinese gentleman was referred to our division for management of a left nasal mass in March 2000. He complained of left-sided blood-stained rhinorrhea for six months. On examination, a polypoid mass was found filling the left nasal cavity. No neck nodes were palpable. CT scan and MRI of the paranasal sinuses showed a soft tissue tumour in the sphenoid sinus extending through areas of cortical erosion to involve the nasopharynx and the base of the left pterygoid process and pterygoid fossa. Further biopsies showed superficial fragments of a bland, hypocellular fibromyxoid stromal lesion. Although no firm pre-operative histological diagnosis was preferred, several differential diagnoses were considered. These included benign entities such as myxoma, superficial angiomyxoma and myxoid neurofibroma (excluded as S100 stain was negative) as well as low-grade malignancies such as myxofibrosarcoma and low grade fibromyxoid sarcoma amongst other myxoid lesions. The lesion was excised via a mid-face degloving approach, in view of uncertain histology and radiologically aggressive features (Figure 3). The nasal septum was detached inferiorly to enhance exposure. The medial wall of the left maxillary sinus, left inferior turbinate and part of the...
posterior nasal septum were resected. The sphenoid sinus
was opened up and complete tumour resection was
achieved. The patient recovered uneventfully after the
operation.

Histological examination of the resected specimen
showed the typical appearances of an intermediate grade
myxofibrosarcoma (myxoid malignant fibrous histiocyt-
toma). The lesion was characterized by tumour nodules
separated by fibrous septa with prominent myxoid stroma
(Figures 4 and 5). Elongated, thin walled curvilinear
vessels with perivascular tumour cell condensation were
seen. The tumour cells stained positively for vimentin but
were consistently negative for other immuno histochemical
markers (including CD34, CD68, S100 and CAM5.2).

The patient was well and remained disease-free at eight
months follow-up. Repeated computerized tomographic
imaging and endoscopic examination showed no tumour
recurrence.

Discussion
MFH was first reported as a unique disease entity by
O’Brien and Stout, 1964. The male preponderance was found
with a ratio M:F = 3:1. The middle-aged to older adults
were more commonly involved. Neck node enlargement
uncommon. The patient recovered uneventfully after the
operation.

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markers (including CD34, CD68, S100 and CAM5.2).

MFH consists of several histological subtypes: storiform-
pleomorphic, myxoid, giant-cell, angiomatoid and inflam-
matory. Both angiomatoid and myxoid types carry better
prognosis and the giant cell type has the worst. Myxofibrosarcoma was described as the myxoid variant of MFH and is one of the commonest sarcomas of the extremities and retroperitoneum in late adult life. The
head and neck is uncommon for both myxofibrosarcoma and other variants of MFH.

The differential diagnosis of MFH consists of a variety of
fibrous tumours and inflammatory conditions. Neuri-
llelioma, fibroma, haemangiopericytoma and gran-
uloma may be confused with MFH. The clinical
presentation is usually not helpful in establishing diagnosis. MFH in the sinonasal tract usually exhibits features similar to other soft tissue neoplasm, namely epistaxis, nasal obstruction, rhinorrhea and non-specific nasal discomfort. There is no distinguishing physical sign to MFH. MRI and CT scan show the extent of tumour involvement and malignant features such as adjacent tissues invasion, but are not in themselves diagnostic. Histological examination is the gold standard for diagnosis.

We report the third case of MFH and possibly the first myxofibrosarcoma in the sphenoid sinuses in the English medical literature (Table 1). Considerable difficulty in making a definitive diagnosis prior to surgery was encountered, primarily attributed to its scarcity and non-specific clinical presentation. Complete tumour resection with adequate resection margin is essential. Local recurrence is frequent with positive resection margins. The midface degloving approach allows good exposure for surgery. The cosmetic result was good and the patient complained of transient infraorbital paresthesia only. Mild nasal crusting in the first few months could be cleaned with simple suction clearance.

Adjuvant radiotherapy is reserved for recurrent, unresectable or large lesions with a high chance of microscopic resection margin involvement; its efficacy is controversial. A post-operative watchful waiting policy was employed in this patient, because of the proximity to the optic nerves and the pituitary gland. Cyclophosphamide and vincristine have been used in metastatic myxofibrosarcoma, but the results are disappointing.

In summary, myxofibrosarcoma is rare and aggressive in the head and neck region. It has a high propensity for local recurrence and distant metastasis. Close post-operative observation is essential. Because of its non-specific clinical presentation and bland histological appearances, definitive pre-operative diagnosis may be problematic. The current treatment is surgery for resectable lesions and radiotherapy for recurrent, unresectable lesions or tumours with positive resection margins. The value of chemotherapy in myxofibrosarcoma is still debatable.

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Dr A. P. W. Yuen takes responsibility for the integrity of the content of the paper.
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