Rhinology, 39, 115-117, 2001

Leiomyosarcoma of the ethmoidal cells*

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SUMMARY

Leiomyosarcomas of the paranasal sinuses are rare malignant tumors. A case of a 68-year-old female with leiomyosarcoma of the ethmoidal cells is presented. Since half a year she had a stuffed nose on both sides. Preoperatively, several attacks of epistaxis on the right side occurred. CT scans showed a tumor of the ethmoidal cells on the right side. The tumor was completely removed via a functional endoscopic endonasal approach and right sphenoethmoidectomy and maxillary sinus surgery. The operation was followed by a radiotherapy with 72 Gy. Up to 29 months after the operation local recurrence could not be observed. Endonasal tumor resection followed by radiotherapy in a case of leiomyosarcoma without invasion of orbit and skull base can allow tumor control.

Key words: functional endoscopic sinus surgery (FESS), leiomyosarcoma, paranasal sinuses, radiotherapy

INTRODUCTION

Leiomyosarcomas are aggressive malignant tumors originating from smooth muscles. They rarely appear in the head and neck region. The relatively most frequent sites of occurrence in the head region are the maxillary sinus and the nasal cavity (Mindell et al., 1975). Due to the aggressiveness of this tumor, radical mutilating surgery has often been performed in spite of high local recurrence and poor prognosis (Som and Brandwein, 1990). In this report, a rare case of leiomyosarcoma of the ethmoidal cells and its treatment is presented.

CASE REPORT

A 68-year-old woman was admitted to the Department of Otorhinolaryngology, University of Ulm in January 1998 because of stuffed nose and recurrent epistaxis on the right side. The patient had first noticed frontal pain in the summer of 1997. There was no swelling of the maxillary or orbital region on the right side and no facial asymmetry. Endoscopic examination of the right nasal cavity revealed a protrusion of the ethmoidal bulla with an obstruction of the right nasal cavity. On a computed tomography (CT) scan a complete obstruction of the right nose, a total occupation of the right ethmoidal cells and a displacement of the lateral nasal wall and the nasal septum could be seen (Figure 1). Bone destruction of the orbit or frontal skull base could not be seen. The tumor extended to the floor of the nasal cavity without destructing the bony palate (Figure 2). Magnetic resonance imaging of the paranasal sinuses was not

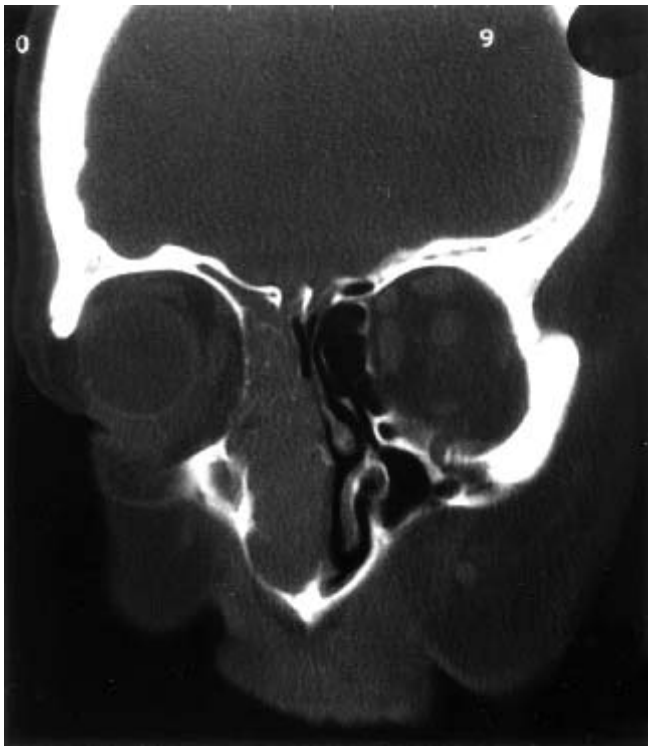


Figure 1. The coronar CT scan shows a complete obliteration of the right ethmoid cell system without destruction of the skull base bone or the medial orbital wall (arrow). The right middle turbinate is shifted towards the nasal septum. The entire right nasal cavity is obstructed and the nasal septum is bended to the left side.

* Received for publication July 11, 2000; accepted November 27, 2000

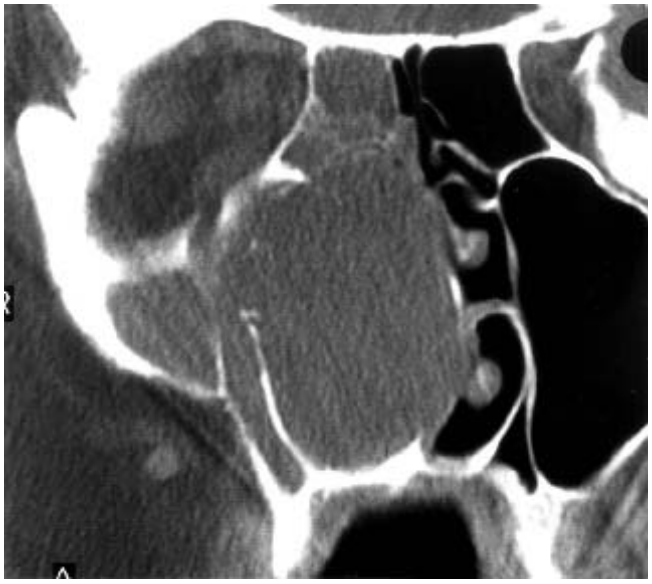


Figure 2. The right maxillary sinus is compressed by lateralization of the lateral nasal wall and tumor extension in the ostiomeatal complex (arrow).

performed. There were no signs of lymphonodal metastasis in physical examination and ultrasound- and CT imaging of the neck region. There were no other tumor localizations in positron emission tomography (PET), ultrasound of the liver, chest X-ray, and scintigraphy of the sceletal system.

A biopsy under local anaesthesia was not diagnostic. An endonasal endoscopic approach for right radical sphenoethmoidectomy, including subtotal resection of the middle turbinate and maxillary sinus surgery, was performed subsequently. The encapsulated tumor which seemed to originate from the anterior ethmoidal cells and the lateral part of the head of the middle turbinate could be totally removed by peacemeal resection. The postoperative course was uneventful.

Histopathological examination of the tumor (ethmoid, ostiomeatal complex) showed a mesenchymal tumor tissue with spindle cells, partially covered by hyperplastic respiratory epithelium (Figure 3). Nuclear polymorphism, several mitoses and focal necrosis could be seen. Immunohistochemical expression of vimentin was present, the expression of desmin and actin was strong. The proliferative marker MIB-1 (i.e., Ki67; Jensen et al., 1998) was positive in 10-20% of the tumor cell nuclei in the sections. The final histopathological diagnosis was a leiomyosarcoma of medium differentiation (G2).

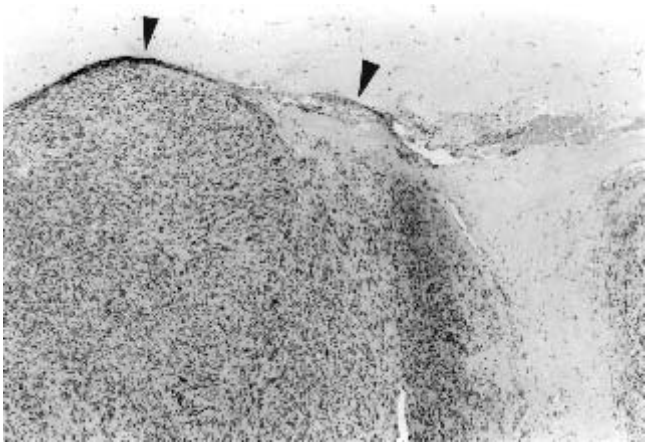
The surgical procedure was followed by a postoperative radiotherapy with 72 Gy over 8 weeks without additional chemotherapy. The patient presented no signs of local recurrence or distant metastases during the 29-month follow-up period, including endoscopical and CT examination (Figure 4).

DISCUSSION

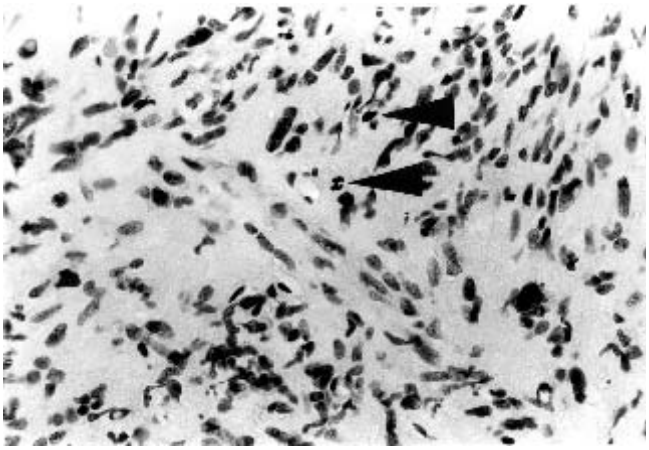
Leiomyosarcomas are rare tumors in the head and neck region. The tumor cells are originating from smooth muscle tissues. Since 1958 when Dobben first described the entity of sinonasal tract leiomyosarcomas (Dobben, 1958), several sporadic cases of occurrence of leiomyosarcomas in the paranasal sinuses were reported. Most of the leiomyosarcomas were found in the nasal cavity involving one or more of the paranasal sinuses or the maxillary sinus only (Ikeda et al., 1997; Josephson et al., 1985; Kuruvilla et al., 1990; Lippert et al., 1996; Tanaka et al., 1998). Localization of leiomyosarcomas in the ethmoid sinus exclusively is even more rare and was only recently described (Reich et al., 1995).

In case of a leiomyosarcoma of the nasal cavity or paranasal sinuses radical surgery was mostly performed via rhinotomy or a transoral or transfacial approach. Radiotherapy or chemotherapy seem not to affect the progression of tumor growth (Dropkin et al., 1976).

The prognosis of this disease is poor. Kuruvilla reported about nine patients with leiomyosarcomas of the sinonasal tract and compared the results of his study with 21 previously reported cases (Kuruvilla et al., 1990). In 10 of the 30 patients who were alive after a follow-up time of 1 to 3 years the main tumor site was found in the nasal cavity. Better outcome after surgical



3A



3B

Figure 3. Light micrographs of the leiomyosarcoma of the ethmoidal cells. A (overview magnification): Extensive mesenchymal tumor tissue infiltrates the soft tissue of the ethmoidal cavity and has reached the covering mucosa (arrows), which is partially eroded. B (detail): The tumor cells have a spindle shape. They are arranged in larger bundles and, although neoplastic, still basically resemble smooth muscle cells. The nuclei are essentially cigar-shaped, but rather pleomorphic. Mitoses are indicated by arrows.

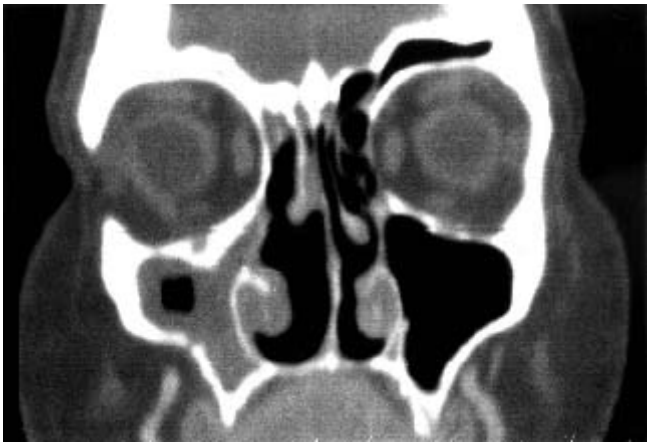


Figure 4. The coronar CT 29 months after surgery only showed mucosal thickening in the right maxillary sinus. Tumor growth was not found in histological examination after endoscopically guided biopsy of the mucosa.

treatment in these 10 cases was suggested to be related to earlier stage of tumor disease or better surgical treatment due to tumor size and localization. Recent reports about postoperative neutron irradiation (Schmäl and Laubert, 1995) and preoperative telecobalt irradiation (Lippert et al., 1996) in the treatment of a leimoyosarcoma of the nasal cavity underline the importance of surgical treatment but proposed extended irradiation in spite of less radiosensitivity of this tumor. In a case of an ethmoid sinus leiomyosarcoma, radical surgery and postoperative external beam radiation has been performed after cyclophosphamide treatment (Reich et al., 1995). Thirteen months after radiotherapy no recurrence of the disease has developed.

Our patient showed unspecific symptoms like epistaxis and nasal obstruction; these are the most common initial symptoms of this tumor. A large tumor mass in the right ethmoid cell system could be seen in CT scans. No tumor growth could be found in the nasal cavity preoperatively or intraoperatively. The ostium of the maxillary sinus was blocked by the tumor mass but tumor growth in the maxillary sinus with bone destruction could not be seen. After an endoscopic approach and sphenoethmoidectomy and maxillary sinus surgery on the right side radiotherapy with 72 Gy followed. All surgical and radiotherapeutical procedures were well tolerated. After 29 months there is no evidence of tumor recurrence or metastatic spreading.

A limited extended surgical approach for tumor resection in combination with irradiation could be performed to achieve local tumor control in a 2-year follow-up. Endoscopic control demonstrated no evidence for residual tumor or bone destruction of the skull base or the medial orbital wall.

We conclude that in our patient relatively early diagnosis without penetration into adjacent bony structures is responsible for the disease-free interval up to now. Kuruvilla already reported on the curability of leiomyosarcomas by complete surgical excision in case of limited tumor extension (Kuruvilla et al., 1990). Tumor localizations in the maxillary sinuses or nasal cavity are more frequent and have more often been related to evident bone destruction. So recurrences in spite of radical surgery were observed.

Because of the known aggressiveness of leiomyosarcomas a combined treatment of surgery and radiotherapy has to be recommended. More reports about this rare tumor are needed to find strategies in the choice of surgical approaches and radiotherapeutical procedures.

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