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Fulminant Adenoid Cystic Carcinoma of the Maxillary Sinus – A Rare Finding: Case Report

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Abstract. *Background: The most common malignant tumor of the small salivary glands in the head and neck region is adenoid cystic carcinoma (ACK). The most common localization of ACK is the hard palate. ACK does not show any sex predisposition and is mainly diagnosed in middle-aged patients. Case Report: The present case report describes a fulminant ACK in the rare localization of maxillary sinus in a 36-year old male. The subsequent surgical treatment consisted of a radical hemimaxillectomy using an extraoral approach according to Weber-Fergusson-Dieffenbach and ipsilateral neck dissection. A magnetic epithesis was used for initial defect coverage of the maxillary bone accompanied by an obturator prosthesis. The surgical treatment was then followed by adjuvant proton therapy. Conclusion: This case report shows how individual patient care can be provided according to the latest therapy standards of ACK in the rare localization of the maxillary sinus.*

Tumors of the head and neck region are among the most common tumor diseases (1). Squamous cell carcinoma is the most prevalent tumor in the head and neck region (2). In addition to squamous cell carcinoma, there are many other types of tumors, in particular tumors of the salivary glands in this anatomical area. These tumors can be divided into tumors of the large and small salivary glands. In particular, tumors of the small salivary glands are primarily malignant (3) and the most common is adenoid cystic carcinoma (ACK) (4). Overall, ACK accounts for a relatively small proportion of all salivary gland tumors (8%) (2, 4).

The most common location of ACK is the hard palate (5). However, there are studies and individual case reports that describe a variable localization of ACK in the area of the tongue and/or the paranasal sinuses (6, 7). The maxillary sinus is the most common carcinoma localization of all paranasal sinuses with a high percentage of squamous cell carcinoma (8, 9). Nevertheless, the present case report shows that pronounced findings within the maxillary sinus can also be due to ACK.

Case Report

The following case report describes a 36-year-old man who attended the Department of Oral and Maxillofacial Surgery of the Army Hospital Hamburg suffering from a swelling on the right side of his face. The swelling was initially noticed about a year ago and showed consistent progression in size. No previous illnesses were found in the patient. The clinical examination showed an indolent swelling on the paranasal right side of the patient (diameter 5 cm). The swelling extended into the right nasal cavity as well as the right palate including several indurations. Further clinical examination did not show any relevant findings. For further diagnostics, a magnetic resonance imaging (MRI) examination of the head and neck region was carried out with the finding of a massive solid tumor in the right maxillary sinus with bone and soft tissue infiltration/destruction (Figure 1 and Figure 2). No distant metastases were found during tumor staging (positron emission tomography-computed tomography; PET-CT). To confirm the findings, an incisional biopsy was performed with the diagnosis of an adenoid cystic carcinoma. The subsequent surgical treatment consisted of a radical hemimaxillectomy using an extraoral approach according to Weber-Fergusson-Dieffenbach and ipsilateral neck dissection (Figure 3). The histopathological findings showed complete surgical removal of the ACK. No pathological lymph nodes were found. A magnetic epithesis was used for initial defect coverage of the maxillary bone

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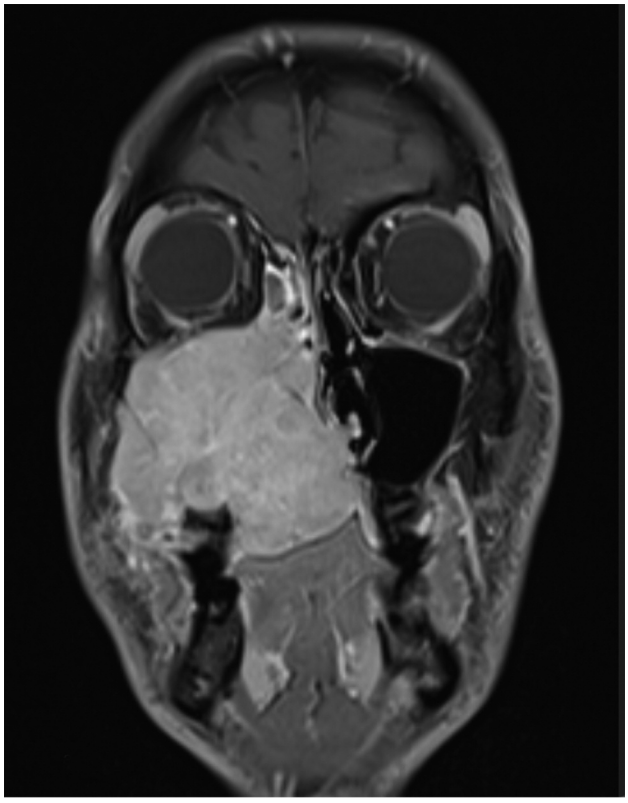

Figure 1. *Preoperative magnetic resonance imaging (MRI) – Coronal view. T1-weighted MRI of a coronal section showing an inhomogeneous, contrast medium-enhancing solid mass in the area of the right maxillary sinus with contralateral midline crossing as well as infiltration of the ethmoid cells and displacement of the nasal septum.*

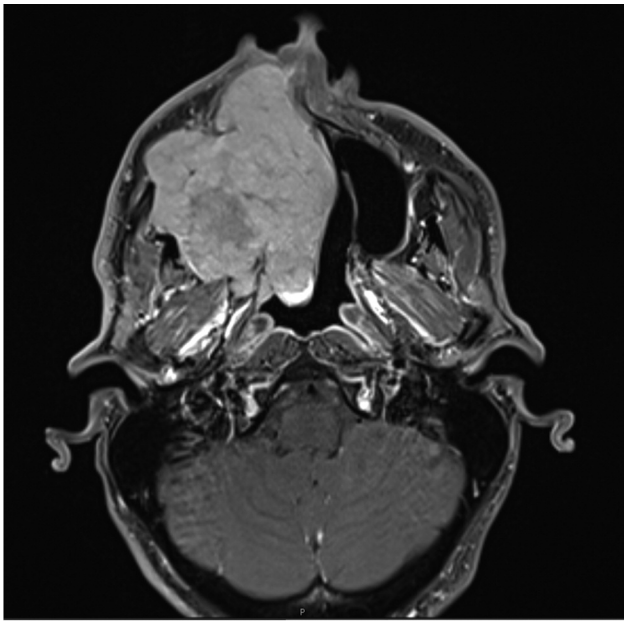

Figure 2. *Preoperative magnetic resonance imaging (MRI) – Axial view. T1-weighted MRI with contrast medium of an axial section showing the length and width extent of the contrast-enhancing inhomogeneous tumor of the right maxillary sinus.*

accompanied by an obturator prosthesis. The surgical treatment was then followed by adjuvant proton therapy. Written informed consent was obtained from the patient.

Discussion

Surgical tumor therapy is an operational focus in a Department of Oral and Maxillofacial Surgery. The treatment of tumors in the paranasal sinuses usually requires an interdisciplinary case discussion, especially with colleagues from the ear, nose, and throat (ENT) Department.

However, tumors of the maxillary sinus are usually operated on by oral and maxillofacial surgeons. As already described, most malignant tumors of the maxillary sinus are squamous cell carcinoma (2, 9). Nevertheless, other malignant tumors such as ACK can also develop in the maxillary sinus. The ACK in particular, does not show any sex predisposition and is mainly diagnosed in middle-aged patients (10).

The symptoms of tumors of the maxillary sinus can be diverse (7). Since the maxillary sinus is a cavity, many tumors in this paranasal sinus grow asymptomatically. Symptoms such as obstructed nasal breathing, chronic sinusitis, epistaxis, or chronic pain can also occur over the course of the disease (7). Also, some patients describe the presence of increased night sweats, recurrent fever, and weight loss (5).

In addition, depending on the growth and extent of the tumor, the displacement of neighboring structures is possible with progressive indolent swelling of the facial region (7). The involvement of swellings, particularly in the area of the palate, as well as the loosening of teeth are also described in the literature (7).

Consequently, many tumors in the area of the maxillary sinus are diagnosed at an advanced stage of the disease (11). However, the ACK shows a special feature due to its perineural growth behavior with intermittent pain and/or sensory disturbances in affected areas (12).

The diagnosis of tumors of the maxillary sinus usually consists of a combination of radiographic imaging and surgical confirmation of the findings by means of a biopsy. In addition to a panoramic tomography, three-dimensional imaging such as CT or MRI can be used for the initial diagnostics (13). Classic two-dimensional imaging such as panoramic tomography or x-rays of the paranasal sinuses often reveal shadowed processes in the maxillary sinus with a partly cloudy structure (13, 14). Diffuse destruction can be observed particularly in the case of infiltrative processes in the adjacent

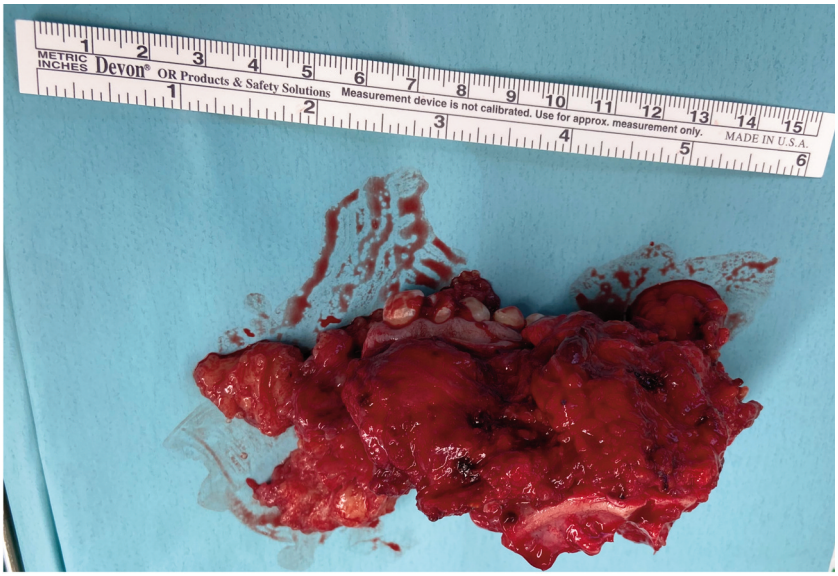

Figure 3. *Intraoperative image showing the tumor after hemimaxillectomy with a width of 11 cm.*

bone structures (14). In particular, three-dimensional imaging (CT, MRI) can make a significant contribution to the diagnostics of tumor expansion and/or infiltration of neighboring structures (14). Increased contrast agent enhancement can often be traced with ACK. In addition, these images are also used for tumor staging, particularly in the head and neck region. Depending on the extent of the tumor, a biopsy is possible via transnasal and/or enoral access.

Different tumor entities can be distinguished histopathologically. The ACK shows three different growth patterns: cribriform, tubular, and solid (15). Most ACK, in the maxillary sinus have a cribriform structure (15, 16). However, the differentiation from other salivary gland carcinomas can be difficult. The use of immunohistochemical markers can help to differentiate between the different carcinomas (17). The radical resection of the ACK of the maxillary sinus with accompanying neck dissection and adjuvant radiotherapy are the decisive therapy pillars (18). In selected cases, especially in the case of pronounced findings, radiation with protons can also be carried out (19).

Studies on ACK show varying survival rates. In the literature, 5-year survival rates vary from 70 to 80% (20, 21). In addition to complete tumor resection (R0), bone infiltrations and the presence of distant and/or lymph node metastases are prognostically relevant (20-22). Especially in the case of metastases, the survival rates drop significantly with equally increased recurrence rates of up to 60% (23).

In the case of the extensive findings of a highly malignant salivary gland carcinoma of the maxillary sinus, as described in the present case, the challenge is not only the complete resection but also the subsequent defect coverage. In particular, epitheses are suitable for intermittent and/or permanent defect repair. However, other surgical procedures are available for maxillary defect repair, such as a fibula or scapula transplant (24).

Conclusion

ACK is a malignant tumor originating from the minor salivary glands. Its localization in the area of the maxillary sinus is rare. This case report demonstrates that even younger patients can suffer from pronounced tumors of this entity albeit of the slow growth of ACK. Fulminant expansion of the ACK in the maxillary sinus requires a radical surgical procedure as well as a special form of adjuvant radiotherapy (proton therapy). In addition to these two challenging therapy pillars, the final defect treatment after tumor resection is a challenge. Nevertheless, the present case report shows how individual patient care can be provided according to the latest therapy standards of ACK in the maxillary sinus.

Conflicts of Interest

The Authors have no relevant financial or non-financial interests to disclose.

Authors' Contributions

KOH, FB, and FD treated the patient and revised the article. FD, CK, and FB researched the scientific literature, provided radiological findings, and wrote the article. All Authors gave final approval for

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