Poorly differentiated sinonasal tract malignancies: A review focusing on recently described entities

Abbas Agaimy

Institute of Pathology, Friedrich-Alexander-University Erlangen-Nürnberg, University Hospital Erlangen, Germany

SUMMARY

Sinonasal tract malignancies are uncommon, representing no more than 5% of all head and neck neoplasms. However, in contrast to other head and neck sites, a significant proportion of sinonasal neoplasms tend to display a poorly/ undifferentiated significantly overlapping morphology and a highly aggressive clinical course, despite being of diverse histogenetic and molecular pathogenesis. The wide spectrum of poorly differentiated sinonasal epithelial neoplasms with small "basaloid" blue cell morphology includes basaloid squamous cell carcinoma (both HPV+ and HPV-unrelated), nasopharyngeal-type lymphoepithelial carcinoma (EBV+), small/large cell neuroendocrine carcinoma, esthesioneuroblastoma, poorly differentiated carcinoma of salivary type (myoepithelial carcinoma and solid adenoid cystic carcinoma), NUT midline carcinoma, the recently described SMARCB1-deficient sinonasal carcinoma, sinonasal teratocarcinosarcoma and, as a diagnosis of exclusion, sinonasal undifferentiated carcinoma (SNUC). On the other hand, a variety of sarcomas, melanoma and haematolymphoid malignancies have a predilection for the sinonasal cavities, and they occasionally display aberrant cytokeratin expression and show small round cell morphology thus closely mimicking poorly differentiated carcinomas. This review summarizes the clinicopathological features of the most recently described entities and discuss their differential diagnosis with emphasis on those aspects that represent pitfalls.

Keywords: sinonasal tract – SNUC – small round cell tumor – NUT midline carcinoma; SMARCB1-deficient carcinoma – esthesioneuroblastoma

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Unlike any other body site or organ, the sinonasal cavities are notorious for being the origin of histogenetically, genetically and biologically highly diverse neoplastic disease entities, an observation that is very surprising given the relatively small proportion of this anatomic region in relation to the total body area (1). Except for conventional squamous cell carcinoma (SCC) and intestinal-type sinonasal adenocarcinoma, the plethora of neoplastic entities in the sinonasal tract and their rarity is only comparable to that of soft tissue sarcomas with every second encountered neoplasm likely being of a different type. This and the fact that sinonasal tract malignancies as a group represents no more than 1% of all malignant neoplasms and 5% or less of head and neck cancers, familiarity with them is generally limited and, thus, the diagnostic workup and exact classification of them pose a real diagnostic challenge in surgical pathology practice necessitating sufficient familiarity with and knowledge of their phenotypic diversity and specific diagnostic criteria. This is further complicated by the fact that poorly differentiated neoplasms at this site frequently display significant morphological and/or phenotypic overlap. Furthermore, diagnosis of some entities relies on demonstration of either specific genetic aberrations (SMARCB1-deficient carcinomas and NUT midline carcinomas) or of a specific infectious agent (EBV or HPV). In this review the pertinent clinicopathological features of the poorly differentiated sinonasal neoplasms and their mimics are discussed with emphasis on aspects that might represent diagnostic challenges or pitfalls.

✉ ***Correspondence address:***
Prof. Dr. med. Abbas Agaimy
Pathologisches Institut, Universitätsklinikum Erlangen
Krankenhausstrasse 8-10, 91054 Erlangen, Germany
telefon: +49-9131-85-22287
email: abbas.agaimy@uk-erlangen.de

SINONASAL BASALOID EPITHELIAL NEOPLASMS

Sinonasal basaloid squamous cell carcinoma and variants

In contrast to conventional SCC which represents the most common carcinoma type in the sinonasal cavities (60%) (2,3), basaloid SCC of the sinonasal tract comprises no more than 5% of head and neck basaloid SCC. The behavior of head and neck basaloid SCC showed site-dependent variation among different studies (4). Although generally considered highly aggressive, recent studies showed survival characteristics comparable to conventional SCC or even paradoxically better than it (4). This variation according to site among other clinicopathological parameters suggests heterogeneity of neoplasms in the generic category of head and neck basaloid SCC. Recent studies illuminated this aspect thereby resulting in splitting of several variants previously included at least in part in this basaloid category (5). Among the latter are HPV-related basaloid SCC with its excellent response to multimodal radiochemotherapy and hence a better outcome compared to conventional basaloid SCC on one hand and the NUT midline carcinoma with its almost universal radio/ chemoresistance and hence dismal outcome on the other hand. Between the two ends of the spectrum are heterogeneous neoplasms including in particular SMARCB1-deficient basaloid carcinoma with generally good but very variable outcome after aggressive treatment regimens. Thus it is mandatory to exactly subtype sinonasal neoplasms traditionally fitting the basaloid SCC line of differentiation. Conventional basaloid SCC is identical to its other head and neck counterparts and is frequently associated with surface epithelial dysplasia (Fig. 1).

A recently reported variant of HPV-associated basaloid sinonasal carcinoma showed a strict sinonasal location among other head and neck sites and adenoid cystic carcinoma-like morphology and/or immunophenotype (6,7). This rare variant can be distinguished from true salivary adenoid cystic carcinoma by