Sinonasal Intestinal-Type Adenocarcinoma
Involvement of the Paranasal Sinuses

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Summary: We present a patient with a biopsy-proved sinonasal intestinal-type adenocarcinoma who presented with moderate confusion. He was found to have bifrontal hemorrhages, which to our knowledge has not been previously described in the literature for this entity. Intestinal-type adenocarcinoma should be in the differential diagnosis of aggressive lesions in the base of the skull with intracranial spread from the paranasal sinuses.

Adenocarcinomas account for 10–20% of all primary malignant neoplasms of the nasal cavity and paranasal sinuses (1). Many of these are of salivary gland origin, but others are less familiar and have histologic patterns similar to those of adenocarcinoma of the colon. These latter ones have been named intestinal-type adenocarcinoma (ITAC) and are responsible for less than 4% of the total malignancies of this region (2). These tumors occur primarily in men aged 55–60 years. The tumors are common in workers in the hardwood and shoe industries. Exposure to wood dust increases the risk of adenocarcinoma by 900 times. Although these neoplasms result in almost 4% of the primary neoplasms of the sinonasal tract, little has been written about them in the radiology literature. We present a case of an ITAC in the sinonasal cavity with intracranial spread, which occurred with bifrontal hemorrhages. To our knowledge, this finding has not been previously described.

Case Report

A 68-year-old man presented to an outside hospital, where he was found to have moderate confusion and evidence of bifrontal hematomas, as demonstrated on a CT scan. He was then transferred to our Veterans Affairs hospital. The patient’s relatives related that he was becoming increasingly confused, disheveled, and inattentive to his personal hygiene. In addition, the patient complained of a 11-kg unintentional weight loss over the last 3 months. He denied any history of trauma. At our institution, neurologic examination showed marked cognitive deficits, but his motor and sensory functions were intact. MR imaging was performed, and the images showed a bifrontal lesion, mostly of high signal intensity on T1-weighted images (Figs 1–3). The lesion extended from the ethmoid sinuses to the skull base and into the parenchyma of the frontal lobes. Erosion of the cribriform plate was highly suspected. The tumor had heterogeneous signal intensity on T2-weighted images, although areas of high signal intensity on T1-weighted images demonstrated corresponding low signal intensity on T2-weighted images. The paramidline portions of the tumor showed enhanced after the administration of contrast material. In addition, substantial edema surrounded the lesion. A preoperative CT scan (Fig 4) was obtained at our institution. This scan demonstrated a bifrontal lesion with attenuation consistent with that of hemorrhage. The ear, nose, and throat (ENT) team was consulted and performed a transnasal biopsy, which revealed ethmoid adenocarcinoma. The patient then underwent bifrontal craniotomy with bilateral orbital osteotomy by means of a skull-base approach involving a neurosurgeon in collaboration with the ENT team. Tumor removal was achieved, and histopathologic analysis revealed a primary ITAC. The patient’s immediate postoperative course was uneventful, and plans were made for postoperative radiation treatment. Subsequently, the patient’s postoperative course was complicated with meningitis, and despite antibiotic therapy, the patient died 3 months after surgery.

Discussion

ITAC of the sinonasal tract may occur sporadically or as an occupational hazard. Exposure to softwood dusts in the logging and milling industries and leather dust in the shoemaking industry has been implicated as risk factors for the development of these neoplasms (3). About 20% of these tumors have historically arisen in individuals with exposure to industrial wood dust (4). The time between the first occupational exposure to wood dust and the development of adenocarcinoma of the sinonasal tract averages 40 years (5).

Findings from recent studies have suggested clinical differences between ITAC arising in individuals with occupational dust exposure and ITAC arising sporadically (3). Tumors related to occupational exposure affect men in 85–95% of cases, and the tumors show a strong tendency to arise in the ethmoid sinuses (6, 7). Sporadic tumors frequently arise in women and involve the maxillary antrum in 20–50% of cases (3). Patients with sporadic ITAC tend to have survival times shorter than those of patients with tumors related to occupational exposure. The reason for this difference is related to the initial stage of the tumor at the time of its discovery. Tumors arising in the maxillary sinus (typically sporadic cases) do not become symptomatic until they are at an advanced stage, unlike those of the nasal cavity and ethmoid tumors, which become symptomatic before they invade local structures (3, 8). Interestingly, our patient had no history of occupational exposure, and although the
lesion seemed to arise in the ethmoid sinus, his symptoms were not related to the sinuses, but rather, to the intracranial spread. Therefore, the tumor was quite advanced at presentation.

Reported sites for the origin of ITACs have been as follows: ethmoid sinuses, 40%; nasal cavity, 28%; maxillary antrum, 23%; and indeterminate, 9% (3). Typical presenting symptoms include nasal obstruction, epistaxis, rhinorrhea, mass in the cheek, and exophthalmos. Less common are symptoms related to facial nerve involvement (3). Symptoms are usually present for less than a year, but they may last as long as 5 years.

The gross appearance of these tumors is similar to that of colonic adenocarcinoma. The most common variant of ITAC resembles typical gland-forming colonic adenocarcinoma, and in many instances, the biopsy specimens are completely indistinguishable from those of a primary colonic neoplasm. Areas of mucosal ulceration are typical and may have associated hemorrhage, as in our case. In poorly differen-

tiated tumors, gland formation is less obvious, and the tumors acquire more-prominent solid components.

The grading of ITAC has been the subject of several studies. Barnes (3) recognized five variants of ITACs: papillary, colonic, solid, mucinous, and mixed. Papillary and colonic tumors have an appearance similar to that of colonic adenomas. They often lack clear-cut features of malignancy, although some of these could be clearly invasive. The literature suggests that the papillary ITAC may have the best prognosis, as it typically behaves as a smoldering, locally destructive lesion with a limited tendency for regional or distant metastases (7, 9).

Initial immunohistochemical results helped confirm the intestinal differentiation of these tumors by documenting the presence of intestinal-type hormones. Interestingly, although carcinoembryonic antigen is strongly expressed in virtually all colonic adenocarcinomas, in one study, only two of 12 ITACs showed strong staining for this antigen (10). In further distinction from colonic neoplasms, nine of 12 ITACs showed numerous chromogranin-positive cells, whereas only three of 12 colonic adenocarcinomas showed rare chromogranin positivity (10).

Pathologists unaccustomed to ITACs may suspect a metastasis from the gastrointestinal tract or even the breast. Metastatic tumors to the nasal cavity and paranasal sinuses do exist and include tumors of the following (in order of decreasing frequency): kidney, lung, breast, testis, gastrointestinal tract, uterus, thyroid, adrenal glands, and pancreas. Also included in this list are melanomas (more common than pancreatic tumors). For this reason, examination of the gastrointestinal tract is important in all patients with a sinonasal ITAC. In our patient, the results of an upper GI series, barium enema study, and CT of the abdomen were negative. Other tests that may be helpful in the differential diagnosis include staining for carcinoembryonic antigen, with which strong positiv-

Fig 1. Axial MR images show a bifrontal lesion.
A, T1-weighted (602/12/1 [TR/TE/NEX]) image shows the lesion with areas of high signal intensity.
B, Postgadolinium T1-weighted (602/12/1) image shows enhancement of the paramidline portion of the lesion (arrows).
C, T2-weighted (5000/96/1) image demonstrates areas of low signal intensity corresponding to the areas of high signal intensity on the T1-weighted images. Note the edema surrounding the lesion.

Fig 2. Sagittal MR images before (left) and after (right) the administration of a gadolinium-based contrast agent demonstrate an enhancing lesion extending from the ethmoid sinuses to the parenchyma of the frontal lobes.
ity suggests the possibility of metastatic disease. Also, strong staining for chromogranin is more typical of primary sinonasal tumors than of others, as stated before. In addition, these tumors must be pathologically differentiated from mucoepidermoid carcinoma and low-grade adenocarcinoma.

The differential diagnosis of anterior skull-base masses extending intracranially includes squamous cell carcinoma, lymphoma, esthesioneuroblastoma, sinonasal melanoma, metastasis, aggressive infection (especially fungal infection), and ITAC. Esthesioneuroblastoma and squamous cell carcinoma usually do not have extreme hypointensity, as seen in our case. Lymphoma may have hypointensity on T2-weighted images but not to the degree seen in our case. An aggressive fungal infection or a sinonasal melanoma could have similar signal-intensity characteristics, especially since these last two entities may have associated hemorrhage. A metastasis may have similar imaging characteristics, especially if it is hemorrhagic.

The signal-intensity characteristics of our patient’s tumor are interesting. T2 hypointensity may be seen in acute hemorrhages, mucin-secreting neoplasms, melanomas, densely cellular tumors, and heavily calcified tumors. The T2 hypointensity of our patient’s lesion is most likely related to hemorrhage, as the pathologic results showed focal areas of hemorrhage and congestion and demonstrated no mucin production or calcification. The preoperative CT scan showed a bifrontal lesion with attenuation consistent with that of hemorrhage and not calcification. In addition, densely cellular tumors would not have had such a high value on CT scans.

Sinonasal ITAC is characterized by one or more local recurrences in more than 50% patients. Regional lymph nodes and distant metastases are less commonly seen, occurring in 8% and 13% of patients, respectively (11). The most frequent sites of metastases are the lungs, liver, and bones.

The optimal treatment for sinonasal ITAC is complete surgical resection with adjuvant radiation therapy to the region of the tumor. In a review by Barnes (3), 60% of patients died of the disease, and 80% of them died within 3 years of diagnosis. Regardless of pathogenesis, the clinical course for these tumors can be protracted. Barnes (3) noted that patients with papillary tumors had a slightly better prognosis, but regardless of the degree of differentiation, all forms of ITAC should be considered locally aggressive. Even tumors deceptively resembling normal intestinal epithelium are known to be locally destructive and ultimately lethal neoplasms (12).

In our patient, several specimens were submitted for pathologic evaluation, including specimens from the base of the skull and the ethmoid region. The final diagnosis was that of a sinonasal ITAC, which was locally poorly differentiated. Our patient’s tumor was locally aggressive, with destruction of the base of the skull and exhibited intracranial extension. To our knowledge, a sinonasal ITAC occurring as intracranial bifrontal hemorrhagic masses has not been reported previously.

**Conclusion**

Intestinal-type adenocarcinoma should be considered in the differential diagnosis of skull base lesions with spread from the paranasal sinuses. These lesions may present with intracranial hemorrhages as in this case. These tumors occur primarily in workers in the hardwood or shoe industry.

**References**


