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Case Report

Sarcomatoid carcinoma involving the nasal cavity and paranasal sinus: a rare and highly progressive tumor

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Abstract: Sarcomatoid carcinoma of nasal cavity and paranasal sinuses is an extremely rare malignant neoplasm. We report a case of sarcomatoid carcinoma arising in the left-side nasal cavity and paranasal sinuses in a 65-year-old man who was hospitalized for left-side nasal epistaxis, odor sensation. Histopathologic examination revealed the tumor was composed of pleomorphic spindle and round cells with frequent mitosis, and no carcinomatous component was recognized. Immunohistochemistry revealed coexpression of cytokeratin and vimentin by the tumor cells, whereas expressions of EMA, S-100, HMB-45, Melan-A, LCA, MyoD1, CD34, CK7, F8 and desmin were negative. The diagnosis was sarcomatoid carcinoma of nasal cavity and paranasal sinuses. The neoplasm was partial resected by nasal endoscopic surgery following chemoradiation therapy immediately. But it was still progressing rapidly, and had a poor prognosis.

Keywords: Sarcomatoid carcinoma, nasal cavity, paranasal sinus

Introduction

Sarcomatoid carcinoma is defined by the World Health Organization as a biphasic tumor consisting of malignant epithelial and mesenchymal elements [1]. The neoplasm has been reported in many sites of the body such as the intestine [2], prostate [3], colon [4], tubulovillous adenoma [5], renal pelvis [6], and lung [7], but it is a rare biphasic malignancy in the nasal cavity and paranasal sinuses [8]. Due to the limited experience, the clinical behaviors of these tumors in the nasal cavity and paranasal sinuses have rarely been specifically addressed, and the treatment and prognosis of these tumors remain ill-defined. Here we present the case of a 65-year-old man with sarcomatoid carcinoma associated with the left-side nasal cavity and paranasal sinuses, and further discuss clinical manifestation, therapeutic options, and prognosis of the disease. The patient's son gave our informed consent for report this case.

Case report

Clinical history

A sixty-five year old man was referred to our ENT department in Nov. 10, 2012. He had left-side nasal epistaxis, odor sensation for two months, and with development of left side nasal obstruction. He already had a nasal endoscopic surgery in local hospital one month ago, with no symptom release, but development of left-side orbit distending pain after surgery. The local hospital's pathologic diagnosis was inverted papilloma. In our department, we found that he had a history of two-side nasal polyp surgery 10 year ago, the physical examination showed many polyp-like masses occupying the left middle nasal meatus and ethmoid sinus zone, the vision of his eyes was unaffected, and no neck nodes were palpable. The MRI scan revealed a mass filled the left-side maxillary sinus and ethmoid sinus, and involving lamina papyracea laterally (**Figure 1A** and **1B**). The neoplasm was partial resected by nasal endoscopic surgery in Nov. 11, 2012. A diagnosis of pure sarcomatoid carcinoma without carcinoma component was made by pathological findings. The patient refused further open operation and went to our radiotherapy department in Dec. 4, 2012. In there, he received 3-dimensional conformal radiotherapy (3-DCRT) and concurrent chemotherapy. The prescribed dose of radiotherapy was 6000 Gy/30 fraction at 2GY/fraction, five fraction in one week. The area of radiotherapy included the tumor, the left-side maxillary sinus,

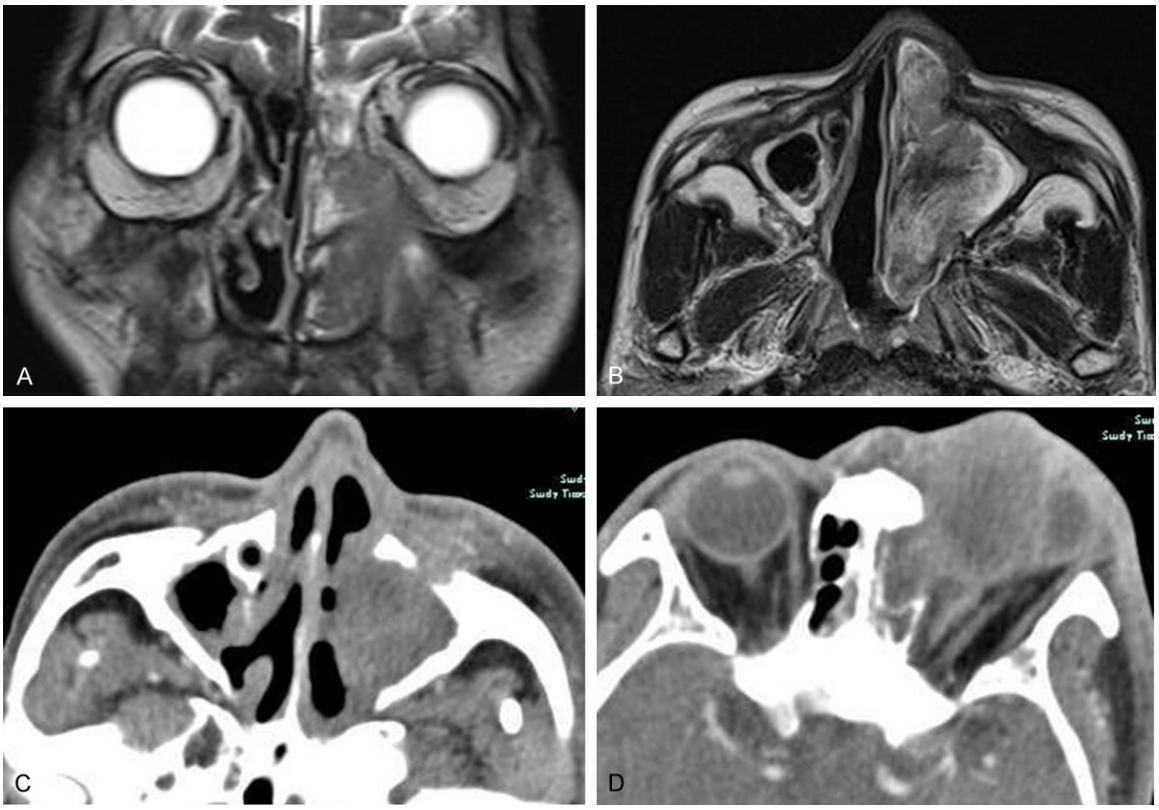

Figure 1. A: The mass occupied the left nasal cavity and ethmoid sinus extending and compressing the left orbital contents. B: The mass origining from left maxillary sinus extending into the nasal cavity anteriorly and posteriorly. C: The computerized tomography showed a soft mass in left-side maxillary sinus with extending into soft tissue of the cheek. D: A 3 × 3-cm mass arising from left-side ethmoid sinus infiltrated the left orbital contents.

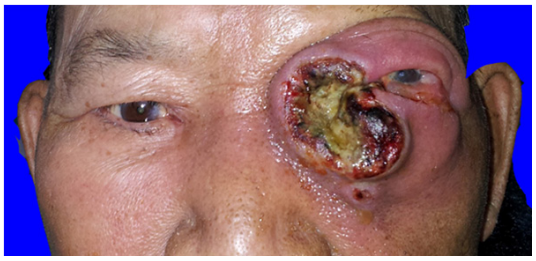

Figure 2. The tumor infiltrated the inner corner skin of the left orbit and the conjunctiva of the left eye, pushing the optic bulb outwardly and upwardly.

the two sides of ethmoid sinuses and nasal cavity. The regimen of chemotherapy consisted of 120 mg/m^2 paclitaxel on Day 1 and 20 mg/m^2 cisplatin on Days 1-3. But to our disappointment, the tumor was still infiltrating peripheral tissue persistently. The clinical examination in Feb. 18, 2013 showed the tumor already involved the skin of left nasal dorsum and eye lids, forming a 4 × 3-cm stink ulcer (**Figure 2**). The CT scan showed the mass infiltrated the left orbital contents and soft tissues of his left cheek (**Figure 1C** and **1D**). But no lymph node was involved and no distant metastasis lesion was found. He was treated with chemotherapy again, consisting of 120 mg/m^2 paclitaxel and 1000 mg/m^2 gemcitabine on Day 1, but no significant effect was obtained. He was recommended to our department for surgery again, and the operation was scheduled after consulting with stomatology department, ophthalmology department and plastic surgery department in Mar. 7, 2013. But the patient refused further surgery, and left hospital, then died a month later.

Pathological findings

Histologically, There were spindle and round sarcomatous cells permeating lamina propria mucosa (**Figure 3A**), and many giant cells and mitotic figures were seen (**Figure 3B**). There was hemorrhagic necrosis in a large segment with rudiment tumor cells in the middle (**Figure 3C**). No carcinomatous component was recog-

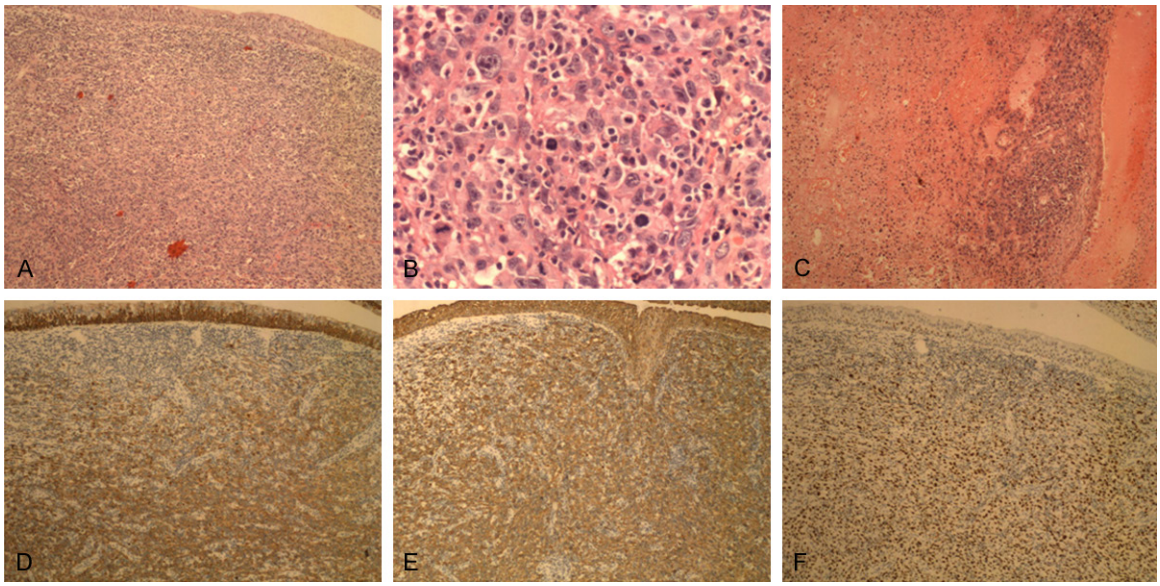

Figure 3. (A) Low-power view of spindle and round sarcomatous cell permeating lamina propria mucosa, normal ciliated columnar epithelium are seen in surface lever. (original magnification, × 40). (B) High-power view of polygonal and round sarcomatous cell of hyperchromatic, pleomorphic nuclei, many giant cells and mitotic figure are also seen. (original magnification, × 200). (C) Tumor are hemorrhagic necrosis in a large segment with rudiment tumor cells in the middle. (original magnification, × 40). Immunohistochemical findings. The tumor cells are positive for cytokeratin 5/6 (D), pancytokeratin Cam5.2 (E), Ki-67 antigen (labeling=80%) (F). (original magnification, × 40).

nized. On immunohistochemistry, the tumor cell were strong positively for cytokeratin (CK), CK5/6, P63, CK19, vimentin, pancytokeratin Cam5.2, and Ki-67 (labeling 80%) (**Figure 3D-F**), but negatively for EMA, S-100, HMB-45, Melan-A, LCA, MyoD1, CD34, CK7, F8 and desmin. So a diagnosis of pure sarcomatoid carcinoma without carcinoma component was made.

Discussion

Sarcomatoid carcinoma is rare, it can occurs in many patterns of body such as the digestive tract, respiratory tract, breast and kidney. Most sarcomatoid carcinoma in hollow viscus are polypoid and pedunculated [9], sometimes it maybe misdiagnosed as benign polyp. In this case, the patient had a history of nasal polyp surgery 10-years ago, and underwent surgery as recurrent polyp in local hospital. For its rare and polypous, making a diagnosis become difficult, the pathologist in local hospital didn't reveal it as sarcomatoid carcinoma.

Morphologically, sarcomatoid carcinomas are mostly biphasic having carcinomatous patterns and sarcomatous components, but occasionally they may be monophasic just having sarcomatous components [10], precisely the present tumor is pure sarcomatoid carcinoma. In such condition, it is difficult to make a diagnosis, and an immunohistochemical study is imperative, if epithelial markers such as cytokeratin, pancytokeratin CAM5.2 are positive in sarcomatous cells, a diagnosis of sarcomatoid carcinoma should be considered [11]. Rarely even epithelial marker are negative in sarcomatous cells, we shouldn't rule out sarcomatoid carcinoma, unless electron microscope reveals no epithelial feature in the sarcomatous cells [7, 12].

Sarcomatoid carcinoma in nasal cavity and paranasal sinus is rare, concrete treatment protocols and prognosis for it have not been reported. The tumor in this case was still persistently infiltrating peripheral tissue after partial resected by nasal endoscopic surgery and chemoradiation therapy subsequently, it showed more tolerated to chemoradiation than squamous cell carcinoma or adenocarcinoma in nasal cavity and paranasal sinus, Terada et al also reported it is tolerated to chemoradiation [13]. Sarcomatoid carcinoma aggressed rapidly in local, but no metastasis lesion was detected. Surgery seems be the first option for primary and recurrent sarcomatoid carcinoma of the nasal cavity and paranasal sinus, and

complete resection in en bloc with larger safety border is needed. Either radiotherapy or chemotherapy seems have little benefit on patient's survival. These treatment protocols were also suggested for treatment of gallbladder sarcomatoid carcinoma [14].

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Disclosure of conflict of interest

None.

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