Sphenoid Sinus Basaloid Squamous Cell Carcinoma Presenting as a Sellar Mass: Report a Case with Review of the Literature

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Abstract Basaloid squamous cell carcinoma (BSCC) is a distinctive variant of squamous cell carcinoma (SCC) with more aggressive behavior. It occurs preferentially in the upper aerodigestive tract. Sinonasal tract BSCC is uncommon, and only limited studies have been reported in literature. In these studies, most BSCCs arose from the nasal mucosa with or without extension to the paranasal sinuses. Rare reported cases of BSCC involved only the paranasal sinus. In this report, we present a case of a female patient with a sphenoid sinus mass. Clinically, the patient had progressively decreasing vision and headache. Magnetic resonance imaging (MRI) and computerized tomographic (CT) scan showed an infiltrating tumor mass involving the sphenoid sinus and the sella with compression of the optic nerve. Pathologic examination revealed an invasive basaloid epithelial neoplasm that was arranged in lobules, nests and cords. The tumor also showed palisading of peripheral cells, focal abrupt squamous differentiation and in situ carcinoma in the surface mucosa. In the immunohistochemical studies, this tumor revealed a strongly positive nuclear staining for p63. The morphologic and ancillary studies indicated a BSCC. To the best of our knowledge, this is the first report of sinonasal tract BSCC that mainly involved the sphenoid bone and sella. In this region, BSCC should be distinguished from benign and

malignant neoplasms that more often affect sella and base of skull, such as pituitary adenoma with extensive necrosis, small cell neuroendocrine carcinoma (SCNC), olfactory neuroblastoma, malignant germ cell tumor, paranasal adenoid cystic carcinoma (ACC), and a variety of metastatic malignancies.

Keywords Basaloid squamous cell carcinoma · Sphenoid sinus · Sellar mass

Introduction

Basaloid squamous cell carcinoma (BSCC) was first reported by Wain et al. [1] and, subsequently, confirmed to be a distinctive morphologic variant of squamous cell carcinoma. BSCC preferentially occurs in the head and neck region. The most common sites to be affected are larynx, hypopharynx, tonsils and the base of tongue [2, 3]. Nose and paranasal sinuses are rarely involved by BSCC. As previous studies indicated, most cases of sinonasal BSCC occur in the nasal cavity with or without paranasal extension [4]. Several case reports have also described BSCC as arising from the maxillary sinus and extending to the frontal portion of the skull base [5, 6]. Primary BSCC as a large sellar mass has not been described. We present a case of BSCC arising from the sphenoid sinus, which presented as a large sellar mass with compression of the apices of the orbits. In this region, BSCC should be distinguished from benign and malignant neoplasms that more often affect sella and base of skull, such as pituitary adenoma with extensive necrosis, small cell neuroendocrine carcinoma (SCNC), olfactory neuroblastoma, malignant germ cell tumor, paranasal adenoid cystic carcinoma (ACC), and a variety of metastatic malignancies.

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Case Report

A 56-year-old female presented with a 1-month history of progressively decreasing vision of the left eye and a mild headache. She had hypertension and type 2 diabetes mellitus for an unknown period of the time and currently was receiving medical treatment. She was a smoker and had consumed about one pack of cigarettes per day for more than 10 years. Physical examination was normal except for decreased left eyesight and a mild elevation of blood pressure. All laboratory tests were within normal limits. Magnetic resonance imaging (MRI) and computerized tomographic (CT) scan showed an infiltrating tumor mass involving the sphenoid sinus and the sella with compression of the optic nerve at the apex of orbits (Fig. 1a, b). The sphenoid bone showed erosion. The sella was enlarged and occupied entirely by a contrast enhancing tumor mass. There were no other lesions in the head and neck region or in the thoracic and abdominal organs by MRI and CT scan studies.

A transsphenoidal resection of the tumor was done and the surgical specimen was submitted for pathologic examination. Under light microscopic examination, both infiltrating and in situ carcinoma components were present. The tumor was composed of moderately pleomorphic basaloid cells with variable growth patterns including nests, cords, trabeculae and large lobules with peripheral nuclear palisading (Fig. 2a, b, c, d). In the large lobules, there were foci of necrosis and abrupt squamous differentiation (single or clusters of cells with eosinophilic keratinizing cytoplasm). Some large lobules showed compacted nests with a jig saw-like pattern. Apoptotic tumor cells and

mitotic figures were easily identified. There was a desmoplastic stromal reaction with focal stromal hyalinization. Perineural invasion and lymphovascular invasion were not seen. Almost all neoplastic cells showed strong positive nuclear staining for p63 (Fig. 2e). The tumor cells were negative for S-100 protein, synaptophysin, CD56, TTF-1, smooth muscle actin, vimentin and p16. Pan keratin (AE1/AE3) was only positive in the focal areas that showed squamous differentiation. The diagnosis of BSCC was rendered and the patient subsequently received radiation and chemotherapy. She was alive with no metastatic disease at the 6-month follow up. CT scan did not show recurrent or progressive disease in the surgical resection site.

Discussion

BSCC was first described as a distinct entity by Wain et al. [1]. Studies have showed that BSCC has a predilection for the larynx, hypopharynx, and base of tongue [2, 3, 7]. Since Wain's first report, BSCC has also been identified at other locations including lung, esophagus, anus, and uterine cervix [8, 9]. Sinonasal tract BSCC is uncommon, and only limited studies have been reported [4, 10, 11]. In the previously reported studies, nasal cavities and paranasal sinus were the most common affected sinonasal sites. In Wieneke et al.'s study, sinonasal tract BSCC arose from nasal mucosa in 11 of 14 cases (4). Nine of them also showed paranasal sinus extension. The other three cases arose from a paranasal sinus (one side or bilateral). One of them extended to the pituitary fossa. All these patients clinically

Fig. 1 Sagittal (a) and cross (b) MRI views revealed a large contrast enhancing mass invading the sphenoid bone, entire sella and compressing the apices of the orbits

Fig. 2 **a** Light microscopy revealed squamous dysplasia and carcinoma in situ of the mucosal surface (Hematoxylin-eosin stain, original magnification 100 $\times$). **b** Light microscopy revealed an invasive BSCC. The BSCC grew as nested lobules in a fibrotic background. Large lobules showed compacted nests with a jagged pattern (Hematoxylin-eosin stain, original magnification 40 $\times$). **c** In this region, the BSCC grew as cords and trabeculae with

peripheral palisading (Hematoxylin-eosin stain, original magnification 100 $\times$). **d** In a high power view, the tumor was composed of closely packed, moderately pleomorphic basaloid cells with hyperchromatic nuclei and scant cytoplasm (Hematoxylin-eosin stain, original magnification 200 $\times$). **e** An immunohistochemical stain for p63 revealed a strongly positive nuclear stain in the tumor cells (original magnification 100 $\times$)

presented with nasal obstruction, nasal bleeding, or both. A few case reports also described sinonasal BSCC arising within the maxillary sinus and extending to the base of skull and sphenoid [5, 6]. A case of primary BSCC of the nasopharynx has also been reported [12]. To the best of our knowledge, BSCC arising from the sphenoid sinus and presenting as a sellar mass has not been described in the literature.

In our case, the patient clinically presented with progressively decreasing vision and headache. The patient had no nasal symptoms. Imaging studies showed a large infiltrating mass in the sphenoid and sella with compression of

the apices of orbits. Preoperatively, pituitary adenoma, metastatic malignancy, chordoma and osteosarcoma arising from the sphenoid were considered. Postoperative pathologic examination revealed an infiltrating carcinoma composed of lobulated nests and cords of closely packed, moderately pleomorphic basaloid cells with nuclear palisading. Foci of necrosis and abrupt squamous differentiation presented in the basaloid lobules. There was a characteristic jagged pattern with a desmoplastic, occasionally hyalinized, stroma. These findings are typical morphologic features of BSCC. Further immunohistochemical studies that show strong nuclear staining for p63

supported the diagnosis of BSCC arising from the sphenoid sinus.

Since BSCC has a tendency to invade locally with regional metastasis, direct extension and metastatic BSCC need to be considered in the differential diagnosis. In this case, there was no evidence of concurrent or subsequently identified primary BSCC developing in the head and neck region or in other locations.

BSCC in the sphenoid sinus needs to be differentiated from tumors that may arise from sinonasal tract, such as a solid variant of adenoid cystic carcinoma (ACC), small cell neuroendocrine carcinoma (SCNC) and sinonasal undifferentiated carcinoma. BSCC may have microcystic spaces containing PAS-positive material similar to punched out spaces in classic ACC, and central comedonecrosis mimicking the solid variant of ACC. However, ACC often has less nuclear pleomorphism, fewer mitosis and a lack of prominent peripheral palisading. BSCC lacks the characteristic features of molding and crush artifacts seen in SCNC. In addition, the presence of in situ carcinoma and abrupt epidermoid squamous differentiation also helps to distinguish BSCC from ACC and SCNC, since ACC and SCNC are rarely connected to the mucosa surfaces. Similar to BSCC, tumor cells of sinonasal undifferentiated carcinoma are often arranged in nests, lobules and sheets. However, sinonasal undifferentiated carcinoma has no squamous differentiation. Compared to BSCC, tumor cells in sinonasal undifferentiated carcinoma show more prominent nuclear pleomorphism, hyperchromasia and high nuclear to cytoplasmic ratio. Other tumors that may involve the sella, such as primary germ cell tumor and metastatic malignancies also need to be considered in the differential diagnosis.

Application of immunohistochemical studies may be necessary to differentiate BSCC from other neoplasms, particularly when transsphenoidal biopsy specimens are small or biopsy artifacts are present [13, 14]. BSCC are usually negative for neuroendocrine markers such as CD56, synaptophysin and chromogranin although S-100 protein may be weakly positive in some case. The immunoprofile for BSCC, ACC and SCNC may overlap. A variable degree of positive immunoreactions for p63 were reported in these tumors. In general, BSCC typically reveals a strongly positive nuclear reaction for p63, while the reaction for p63 in ACC and SCNC was usually weak and variable. This feature, when combined with application of other immunostains, is helpful in distinguishing BSCC from ACC and SCNC.

In the past, BSCC was thought to be a morphologically distinctive high-grade variant of SCC. However, currently BSCC is not considered to be a high-grade carcinoma, particularly in the group of BSCC that show human papillomavirus (HPV) infection. Studies indicate that HPV

plays an etiologic role in a subset of BSCC, specifically BSCC in the oropharynx [15]. In this region, up to 75% of BSCC contain high-risk HPV DNA and show elevated expression of p16. Compared to HPV-positive nonkeratinizing SCC in oropharyngeal region, HPV-positive BSCC have similar clinical outcomes. HPV-positive BSCC have significantly better overall survival when compared with HPV-negative BSCC. In the larynx, hypopharynx and other head and neck regions, rare BSCC show positive HPV DNA in the tumor cells.

In conclusion, sinonasal tract BSCC is uncommon. The most commonly affect sites are the nasal cavity and paranasal sinuses. Primary BSCC may directly arise from the sphenoid sinus, present as a large mass and affect the sella and base of skull. Sphenoid BSCC needs to be differentiated from the lesions that more commonly affect the skull base. Since BSCC has a tendency to metastasize, it is necessary to rule out metastatic BSCC before diagnosing a primary sphenoid sinus BSCC.

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