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Case Report

Coexistence of Neuroendocrine Neoplasm (Carcinoid Type) and Rathke's Cleft Cysts in the Sella; A Case Report and Review of Literature

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Abstract

Intracranial Neuroendocrine Neoplasm (NET) is extremely rare. Herein, we report a very rare case of collision of primary NET (carcinoid type) and Rathke's cleft cysts (RCCs) in the sellar region. A 57-year-old man received a medical checkup and notified a possibility of pituitary neoplasm. However, he did not have any symptoms suggesting intracranial neoplasm, and kept follow up observation. About 5 years later, the patient started consistent headache. Magnetic resonance imaging revealed a cystic mass 25 mm in diameter in the sellar region. Mass removed by an endoscopic endonasal transsphenoidal surgery from the neoplasm located nearby RCCs and pituitary gland displayed morphology of NET (predominant trabecular pattern) with strong immunohistochemical activity of chromogranin A and synaptophysin. The neoplasm was frequently attached to the ciliated columnar or cuboidal epithelium of the cyst wall of RCCs. Such case has never been reported in the literature. Clinicopathological features of the collision case is described and a presumption regarding the possible histogenesis of the NET in the brain is discussed.

Introduction

Carcinoid (neuroendocrine neoplasm) is a rare type of neoplasm. Most of these tumors derive primarily from the gastrointestinal tract (55%) and the bronchopulmonary segments (30%) [1] but are capable of arising throughout the body. Occurrence of the intracranial carcinoid is very rare, only 4 cases have been reported in the literature [2-5]. Of them, first case was recognized as a mass with a prominent dural-based tail that compressed the right frontal lobe. Second case was confirmed as a mass located at the foramen jugulare extending into the cerebello-medullary angle cistern. Third case was revealed as a tumor in the cerebello-pontine angle. For the fourth case, the tumor was present at a cavernous sinus extending into the infratemporal fossa. Thus, location of these carcinoids was variable and they were not collision types. However, considering present case in addition of these reports, skull base area will be one of preferable sites for the occurrence of carcinoid tumors. In this case, carcinoid coexisted with Rathke's cleft cysts (RCCs) known as pars intermedia cysts, represent benign lesions formed from remnant of the embryologic Rathke's pouch. The incidence of RCCs varies from 2 to 26% as seen in autopsy series [6,7]. Coexistence of RCCs and pituitary adenomas is already notable [8-10].

Concerning the pathogenesis of collision of RCCs and pituitary adenoma, Kepes [11] exhibited a transitional cell tumor of the pituitary gland developing from a RCCs, considering that

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the tumor was derived from "transitional" cells between the lining cells of RCCs and the glandular cells of the anterior pituitary. However, the theory was rejected by Ikeda et al [12] who proved that the tumor shown by Kepes [11] corresponds to an early development stage of the pituitary anterior lobe and that a cyst within a pituitary adenoma differs from cyst found in the embryonic stage of the pituitary gland. Thus, pathogenesis of the collision of pituitary adenoma and RCCs is still controversial. Presently, we report a collision case of carcinoid tumor and RCCs. So far as we concerned, such collision case has never been described. Certainly, the relationship between the two lesions is now unknown. Nevertheless, our report will be important for understanding of sellar lesions including neoplasms and non-neoplastic cystic lesions.

Case presentation

A 57-year-old man received a medical checkup at medical checkup center of our hospital. After the CT examination as a course of the checkup, the patient was notified a possibility of neoplasm of pituitary gland, although hormonal inbalance was not present. Neurological examination including visual activity and visual fields showed no abnormality. Since he did not have any symptoms suggesting intracranial neoplasm, a follow up of observation by the group of neurosurgery of our hospital was decided. About 5 years later, he started to develop consistent headache. Magnetic resonance imaging revealed a cystic sellar mas which was slightly hypertense with sparse hypointence in the lesion on T2-weighted image (Figure 1A) and was enhancing after the administration of gadolinium (Figure 1B). These evidences suggest to indicate that cystic mass gradually grew up from 20 mm to 25 mm in 5 years.

The patient underwent an endoscopic endonasal transsphenoidal surgery (eTSS). During the procedure of eTSS, jerry-like fluid was discharged implying the presence of RCCs. Histologically, the removed tissues were solid neoplasm together with cyst walls and tissue of the pituitary gland with non-neoplastic changes indicating that the neuroendocrine neoplasm was present nearby RCCs and the pituitary gland in the sellar region. The cysts had lining by ciliated columnar or cuboidal epithelium (Figure 2). Squamous cells or goblet cells were not recognized. The cysts were diagnosed as RCCs. The tumor cells showed the predominance of a trabecular pattern, often admixed with tubuloacini or broad, irregular trabeculae with rosettes, and only occasionally with solid nests (Figure 3). Cytologically, the tumors possessed uniform round to oval nuclei with indistinct nucleori and coarsely granular chromatin pattern. Cellular mitosis was few and labeling index of KY67 was <2%. Immunohistochemically, the cells appeared strong activity of chromogranin A and synaptophysin (Figure 4). The neoplasia was diagnosed as a neuroendocrine neoplasm (carcinoid type) of low grade. The lining epithelium of the cysts was frequently attached to the margin of the neoplasm (Figure 3). After the operation, the patient has been healthy without any symptoms of the sellar lesion.

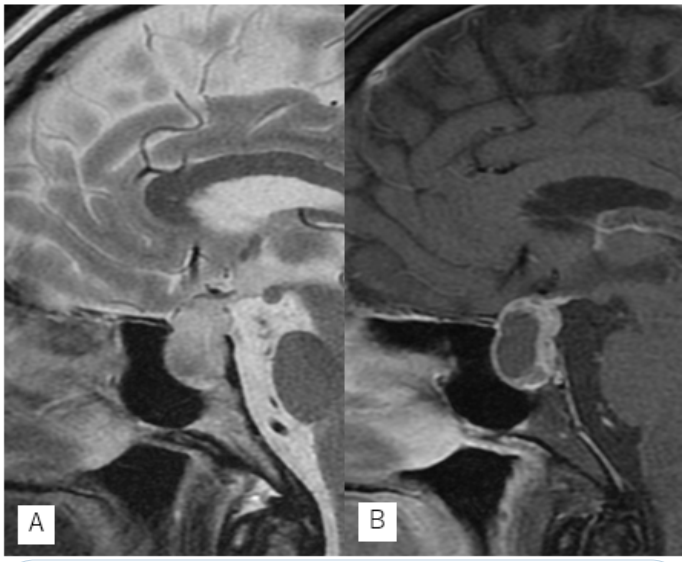

Figure 1: Magnetic resonance imaging brain showing a cystic mass in the sella.

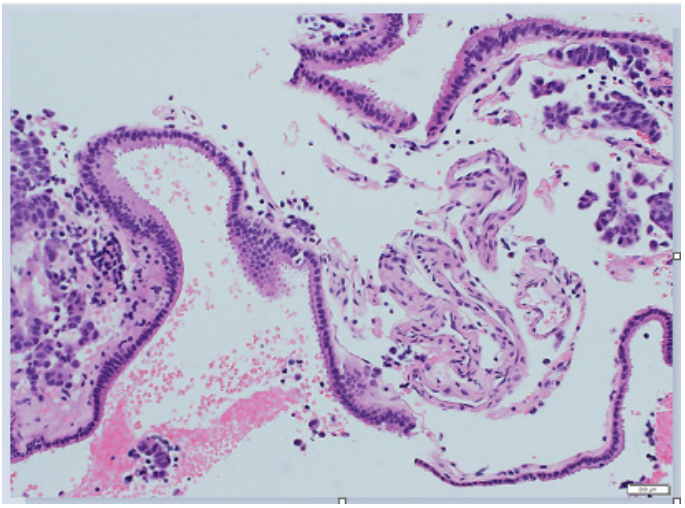

Figure 2: Histology of the cyst walls with ciliated columnar or cuboidal cells of RCCs.

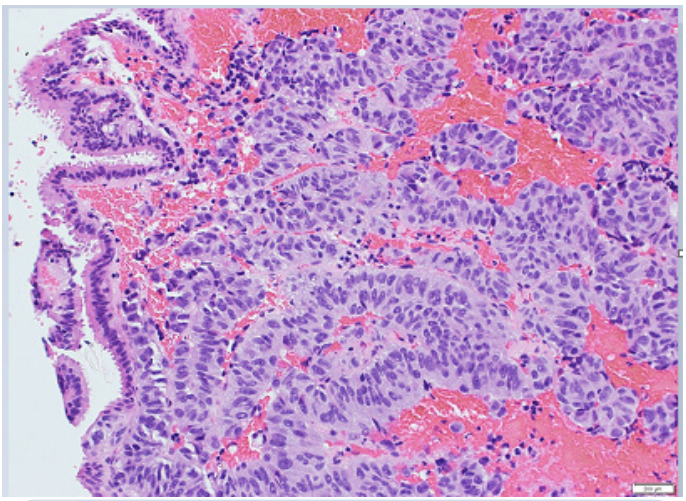

Figure 3: Histology of neuroendocrine neoplasm (carcinoid type). Trabecular pattern is clear. Ciliated epithelial cells are attached to the neoplasm.

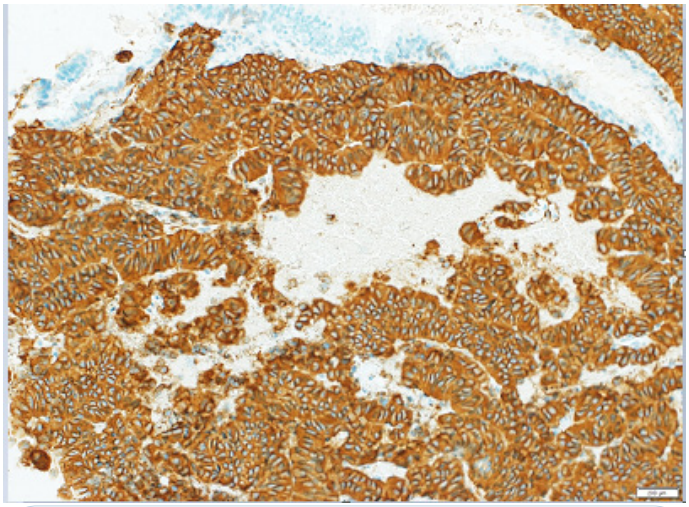

Figure 4: Positively stained by chromogranin A suggesting that the tumor is neuroendocrine neoplasm.

Discussion

In the present case of neuroendocrine neoplasm, histological evaluation revealed that the tumor was a neuroendocrine neoplasm of low grade with carcinoid-like pattern. Furthermore, no other primary or metastatic sites were detectable suggesting that the neoplasm was primary. Up to date, only a few cases of intracranial carcinoids have been reported [2-5]. Interestingly, these carcinoids were mainly dural-based tumors. Furthermore, reported evidences suggest that carcinoid tumors may have an affinity to dura mater on metastasis of other organs into the brain [13,14]. Thus, it is presumed that some of the embryonic cells developing into the neuroendocrine tumors may locate in the stromal tissues surrounding cerebrum including those of skull base. Furthermore, it is also suggested that the neuroendocrine neoplasm like this case starts to grow from the stromal cells in the sellar area.

Another possibility regarding the origin of the present neuroendocrine neoplasm would be certain potential cells migrating in the cyst wall of RCCs. According to Batt et al [15] with 30 cases RCCs, 56.6% of them had pituitary acini in their walls, and 5.6% of them constituted all the non-neoplastic cysts of central nervous system including neuroepitheial element. Accordingly, RCCs may have a role for development of neuroendocrine neoplasm as well as pituitary adenoma. Present case of collision of carcinoid and RCCs could be a key issue for understanding of clinicopathology of neuroendocrine neoplasm of the brain and significance for the relationship between non-neoplastic cystic lesions and neuroendocrine neoplasm in the central nervous system.

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