

A Case Report of Left-Sided Headache with Rhinocleisis

Hailing Hou^{1,2}
Zhiyong Yuan²
Ping Wang²
Li Zhu²

¹ Key Laboratory of Cancer Prevention and Therapy, Tianjin 300060, China.

² Department of Radiotherapy, Tianjin Medical University Cancer Institute & Hospital, Tianjin 300060, China.

Correspondence to: Hailing Hou
E-mail: hhl0528@sina.com

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CJCO <http://www.cjco.cn> E-mail: cocr@eyou.com
Tel(Fax): 86-22-2352 2919

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Case Report

A patient, with a complaint of a left-sided headache plus a rhinocleisis for a month, was admitted to our hospital. The clinical diagnosis showed that it was a primary adenoid cystic carcinoma (ACC) of the nasopharynx. At another hospital she had undergone 17 ordinary external irradiations, with a test dose (DT) of 200 cGy/t, and a cranial gamma knife treatment, with a central dosage of 35.6 Gy/t, however the local lesion recurred. After hospitalization, a whole-body chemotherapeutic regimen with DDP+THP+FUDR was administered, but her symptomatic relief was not effective. Then the patient received 5 X-knife treatments, at a dose of 400 cGy/t, resulting in a slight alleviation of her pain.

The case discussion of our study involves some aspects, such as the pathology, imaging, surgery and radiochemotherapy of this disease, etc., with an aim of instituting a standard plan of treatment to relieve patient suffering, to improve the quality of life, and to devise clinical therapy. Primary ACC of the nasopharynx is rare, with most patients being female. The tumor growth presents an evident invasion. Because of the complexity of the excision and infiltration of ACC into the vessels and nerves, surgery is not appropriate. So radiotherapy is the treatment of choice. The 5-year local control and survival rate of patients with this tumor is 21% and 31%, respectively.

Case history briefing

General patient data

A 45-year old female was admitted on the 15th of July, 2006 in the Radiotherapy Department of our hospital, with the complaint of left-sided headache plus a rhinocleisis for one month.

Medical exam after hospitalization

The left nasolabial groove was slightly shallow, and the left-hand side of the nasal cavity was obstructed, without abnormal secretions. There was an absence of tenderness in all of the accessory nasal-cavity areas, and a positive preauricular tenderness. No intumescent lymph nodes were found upon bilateral palpation of the neck and other superficial areas.

Laboratory and auxiliary examinations

Laboratory results indicated that the indices for blood and uriner-

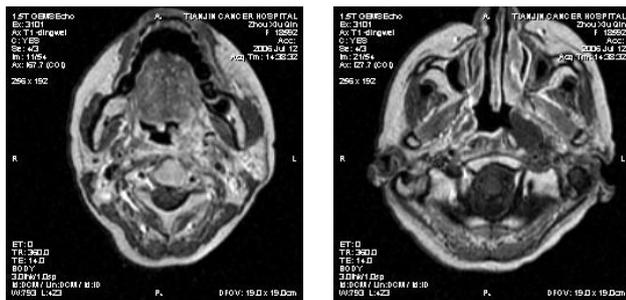


Fig.1. CT image of oral pharynx, nasopharynx and cranial base of the patient.

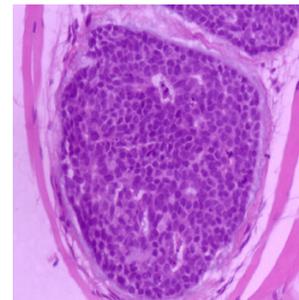


Fig.3. A pathological section (10 × 40).

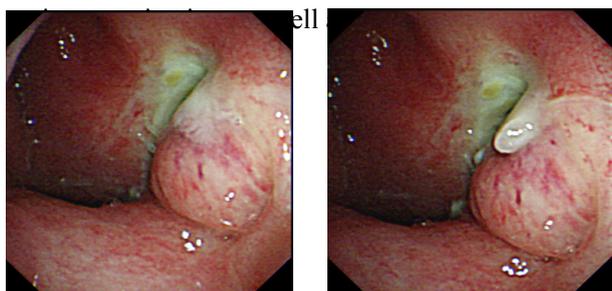


Fig. 2. Nasopharyngolaryngoscopic images.

roughly normal; the physical state scoring indicated a KPS of 80. The results of the imaging examination were as follows: MRI showed that there was a thickening on the left-side of the nasopharynx and lateral wall of the oropharynx, with an abnormal signal and a clearly inhomogenous reinforcement. Left pharyngeal recess disappeared, and in the parapharyngeal space an abnormal fortified image could be seen. The left basilar region presented an inordinate abnormal intensification, with an approximate scope of 5.2×3.5 cm and an intrusion into the middle cranial fossa. No obvious abnormal signal was found in the accessory nasal cavity (Fig.1); endoscopy (nasopharyngoscope): the left pharyngeal recess was covered by a yellow white necrotic tissue, and the left torus was rough, with congestion and tumescence. A few yellow and white sphacluses were seen on surface of the lesion, and the biopsy tissue had a tenacious texture. There was an engorgement at the left-side opening of the auditory tube, which was not sharp. The right torus and pharyngeal recess as well as the opening of the auditory tube was well-defined and smooth (Fig.2). Pathological results of the biopsy (nasopharynx) showed chronic inflammation and necrotic tissue.

Patient's case file in another hospital

Prior to admission of our hospital, the patient was admitted in a hospital in Tianjin on the 1st of September, 2004, with a complaint of the left-side rhinoclei-

sis for 3 months and then a nasopharyngeal tumor for 1 month. Biopsy of the nasopharyngeal tumor was conducted, with an initial report of the pathologic results showing the ACC (Fig.3); performance of local radiotherapy: 17 treatments were completed, at a dose of 200 cGy/t. The patient withdrew from the treatment and left the hospital on account of intolerance to the adverse reactions. On the 9th of January, 2006, she was admitted into another hospital in Tianjin, with a final diagnosis of the nasopharyngeal carcinoma for over one year, and a complaint of pain on the left-side of her face and head after radiation therapy. A cranial gamma knife treatment was conducted with local anesthesia on January 10, 2006; localization film showed that there was a disorderly structure on left-side of the nasopharyngeal space, and the intrafat space vanished. An irregular massive abnormal signal shadow could be seen, with displacement owing to a compression on the adjacent tissular structure. The left pharyngeal recess had become shallow, and the internal carotid artery and internal jugular vein were encased by the lesions. There was a well-defined boundary between the lesion and the external pterygoid muscle, without seeing visible intumescent lymph nodes in the neck. There were 9 targets in the TPS, with a weekly dose of 16 Gy and an isodose curve of 45%; central dose: 35.6 Gy. The patient left the hospital following the postoperative symptomatic treatments, such as pain relief, dehydration and hormonal therapy, etc.

Patient's condition and treatment after admission in our hospital

After admission, a regimen of whole-body chemotherapy (DDP+THP+FUDR) was administered for one week, but with no obvious symptomatic relief. Five additional localized X-knife treatments were conducted using a dose of 400 cGy/t. The symptom of pain was slightly alleviated compared to previous treatments.

Topics for next discussion

1. Further treatment plan of the patients.
2. Important significance of standard treatment.

Clinical discussion, excerpts from a discussion of intractable cases in the Department of Radiotherapy, Tianjin Cancer Hospital, Tianjin

Prof. Zhang Lianyu, Pathology Department: Head and neck ACC most frequently occurs in the tissues of the salivary gland. Based on the WHO histological typing in 1991, ACC was divided into 3 types: tubiform, screen opening and substance types. There is an obvious correlation between the pathologic typing and prognosis. The prognosis of the substance type is poor, and the screen opening type has the best prognosis among all of the ACC cases^[1]. Primary minor salivary gland tumors of the nasopharynx are rare, comprising about 2% of the minor salivary gland tumors, with ACC as the most common histological subtype. The major basis for histopathological diagnosis of the nasopharyngeal ACC includes the following: a) more dark staining of the cancer cell nucleus and less cytoplasm, presenting a basal cell form; b) a homogeneity or/and muciform material between the cancer cells; c) a characteristic screen mesh-like structure^[2].

Dr. Liu Peifang, MRI Section: CT and MRI respectively possess their own superiority over all diagnostic methods for nasopharyngeal ACC. The bony-wall destruction and the neoplasm in the capsular space can be easily observed by CT scans, thus clearly demonstrating the extent of the disease. However, the CT scans fail to judge whether there is tumor infiltration around the orbit, and can not identify the soft tissues from retention liquid. It also can not judge a specific scope of tumorous invasion. However MRI can clearly display an image of the lesion, especially when there is a progression of the tumor along the nerve fiber bundle, and antegrade or regressive thickening nerves can be shown on MRI. Owing to the profuse necrotic tissues in the lesion of our patient a half year after gamma knife treatment, a clear display of the lesion could only be seen 9 months following radiotherapy, even though MRI has an advantage over CT scans in observation of the basicranial lesions.

Dr. Hu Jianzhang, Endoscopy Section: Half a year after a gamma knife treatment of the patient, the surface of the lesion was covered with profuse necrotic tissues, which became an obstacle in the biopsy, increasing the difficulties for pathological confirmation of the local recurrence. So diagnosis of

the lesion should be closely involved with the clinical manifestations, as well as the sections, imaging and pathology.

Dr. Wang Xudong, Department of Head and Neck Surgery: It has been proposed in the past that surgery should be the first choice for treatment for head and neck ACC, with a combined therapy of postoperative conventional radiotherapy and/or chemotherapy. A sufficient surgical margin is needed for surgical treatment of the primary focus. Since the local lymphatic metastasis of ACC is very low, it is unnecessary to conduct a prophylactic radical neck dissection (RND). Regional RND will be beneficial for treating doubtful cases, and the RND should be conducted once the intraoperative frozen section shows a metastasis of the jugular lymph nodes^[3]. However, since there is a complicated anatomic structure in the nasopharynx, with frequent encroachment of the ACC upon the vessels and nerves, the surgical treatment is unfit and radiotherapy should be the first choice.

Dr. Xie Guangru, Department of Integrated Chinese-Western Therapy: At present postoperative chemotherapy has been conducted in only some of the patients with a distant-metastasis of the head and neck ACC in China and overseas. Therefore, assessment of the role of chemotherapy for treatment of distant-metastasis from ACC is unavailable^[4]. However, in view of a nonstandard treatment of the patient over the previous days, nedaplatin can be used as a basic chemotherapeutic agent for prevention of distant metastasis, in accordance with the physical state of the patient, as the radiotherapy on local relapse is performed.

Prof. Li Ruiying, Radiotherapy Department: Induced chemotherapy, with paclitaxel as the principal agent, can be tentatively applied. In addition, fractionated stereotactic radiotherapy for head and neck ACC is safe and effective^[5].

Prof. Wang Ping, Radiotherapy Department: The irradiation field for nasopharyngeal carcinoma should comprise the tumor bed and should be close to the sub-clinical foci. The irradiation scope of the gamma knife treatment might be too limited resulting in a basicranial, oropharyngeal or parapharyngeal progression. Moreover, only if the one-time quantity-sufficient radiotherapy is completed, can the local control of recurrence be preferably managed.

Summary

Primary ACC of the nasopharynx is rare, compris-

ing approximately 1.5% of head and neck ACC^[6]. Females make up the majority of cases. ACC differs from squamous cell carcinoma of the nasopharynx in that there is not a very close relationship between nasopharyngeal ACC and EB viral infection^[2]. The tumor growth presents an evident invasion, with a frequent infiltration into the nasopharynx and its peripheral tissue. Its growth and differentiation may depend on the laminin-5 basal membrane^[7]. Positive expression was found in the p53 protein in about 38.1% of the ACC patients. In addition, distant metastasis in these patients is very high, and has a low 10-year survival rate^[8].

Owing to the complicated anatomic structure of the nasopharynx and frequent encroachment of ACC upon the peripheral vessels and nerves, surgical treatment is inadvisable. Therefore radiotherapy becomes the major therapeutic means. Entire first-course treatment is the key to success. The exposure field should comprise the tumor bed and neighboring subclinical focus, with a radiotherapeutic dose of 70 Gy or higher^[9]. During the treatment, special attention should be paid to the areas where there are basicranial nerves etc. Fast neutron^[10] and carbon particle therapy may raise the therapeutic efficacy^[11]. The prognosis of ACC has a close correlation with some factors, such as age, duration of the symptoms, paraesthesia, histopathology types, tumor size, regional lymph node metastasis, distant-metastasis, postoperative residue and clinical stages etc^[6]. The 5-year local control and survival rates of ACC patients were reported to be 21% and 31%, respectively^[12]. Concerning the occurrence of primary ACC in other sites, such as trachea,^[13] bronchus,^[14] esophagus^[15] and external auditory canal etc.^[16], surgery has always been the treatment of choice. Patients with a total resection of these tumors have a relatively favorable prognosis.

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