Naopharyngeal Notochondroma- A Case Report and Literature Review

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Abstract

Background: Nasopharyngeal notochordoma is a rare congenital low-grade malignant tumor which is very rarely seen in clinical practice. We describe a case of nasopharyngeal notochordoma, the patient admitted in our department for surgery for fifth time.

Case report: The patient complained of left sided nasal obstruction, snoring, with left fronto-temporal headache and progressive decline of vision especially of left eye for 2 months. She had been performed surgery for the removal of nasopharyngeal notochordoma for four times. Every time she was performed surgery postoperative imaging examination showed no any residual tumor mass. But unfortunately there was always recurrence of mass. This time (fifth time) the patient was admitted for surgery, latter after the CT scan and MRI examination, showed bone destruction so patient denied for surgical treatment.

Conclusion: In this study we describe the clinical features, diagnostic evidence, treatment and discuss the possible reason of recurrence. Nasopharyngeal notochordoma is impossible to remove completely by surgery so other radiotherapy will help to increase prognosis and survival rate. As clinical feature of nasopharyngeal notochordoma is quite similar to nasopharyngeal carcinoma so it should not be confused in diagnosis and treatment.

Keywords: Notochordoma; Congenital; Malignant tumor; Recurrence

Introduction

Notochondromas are low-grade malignant tumors arising from embryonic remnants of notochord in craniopharyngeal axis, usually scro-coccygeal and spino-occipital region. Remnants of notochord form the nucleus pulpous of the intervertebral disks in adult. A remnant of notochord persists in skull base, sacro-cccygeal area and body of vertebra. Nasopharyngeal notochordoma is a rare case in otologyngology department, so it is often misdiagnosed as other diseases. It exhibits slow invasive growth and locally destructive nature. Because nasopharyngeal notochondromas present with new growth of mass in nasopharynx, done destruction of skull base so sometime it may be misdiagnosed as nasopharyngeal carcinoma especially in those reason where nasopharyngeal carcinoma is common. We describe the uncommon case of recurrent nasopharyngeal notochordoma.

Case Report

A 22 years old female, without any obvious inducing factors patient complained about left sided nasal congestion, snoring, with left fronto-temporal headache and progressive decline in vision especially left eye for 2 months, no nasal bleeding, no purulent nasal discharge, no bloody sputum, no eye movement abnormalities, no dysphasia, no limbs movement disorder. Six months ago she underwent resection of nasopharyngeal chordoma in our hospital. For the further diagnosis and treatment, patient come to out-patient department of our hospital, diagnosed as “post-operative recurrence of nasopharyngeal chordoma?” Since the onset of the disease, the general mental state, sleep and appetite is normal, no significant changes in body weight. From 2000-2014 4 times surgery was performed. Four previous surgeries in 2000 March; performed endoscopic nasopharyngeal notochordoma resection surgery, in 2007 January and 2009 September performed combined endoscopic palatal approach notochordoma resection surgery, and in June 2014 performed endoscopic trans-sphenoidal approach nasopharyngeal notochordoma resection which was not performed in our hospital. This time when she was admitted in our department, all the preoperative tests were normal. Nasopharynx computed tomography scan (CT scan) (2015-1-20) (Figure 1A, B and C) and magnetic resonance image (MRI) (2015-1-23) (Figure 2A, B and C) revealed postoperative changes in skull base notochordoma, recurrence?

Figure 1: CT scans of fifth time preoperative (2015-1-20). (A) Axial (B) Sagittal (C) Coronal.
During routine nasopharyngeal endoscopy examination found polypoid growth in nasopharynx (Figure 3) biopsy of that mass showed inflammatory polyps (Figure 3). Patient was planned for surgery again after the discussion with the patient’s family member and consultation with department of radiotherapy, patient denied to perform surgery this time but rather try local radiotherapy and the patient was discharged from our department.

Discussion

Nasopharyngeal notochordomas are slow-growing, can continue to grow for several years to more than 10 years. Nasopharyngeal chordoma occurs mainly in nasopharynx (basisphenoid or basiocciput or tissue around eustachian tube orifice), but also invade to the middle cranial fossa, saddle area, clivus, petrous apex and parasella [1]. Chordoma can occur at any age. Which can range from 1 year to 80 years old, but mostly occurred in the 40s. Male sexes are predominant, between the ages of 40-60 [2]. Nasopharyngeal chordoma usually presents with nasal obstruction, purulent nasal discharge, decreased sense of smell, nasal obstruction, snoring, tinnitus, hearing loss, decline of vision, headache, and sometimes may have nose bleeding and difficult in swallowing [3].

In our case the patient also complain of left sided nasal obstruction, snoring, with left fronto-temporal headache and progressive decline of vision especially left eye. Form 200-2014 the patient had been operated for 4 times with different surgical approach but there was reoccurrence of the tumor. Figure 4(A) showed the MRI preoperative images which were taken before the surgery for second time, mass in the skull base and bone destruction confirming the reoccurrence of the mass. Every time the patient underwent surgery postoperative CT scan or MRI showed tumor free, but the mass reoccur again and again within few months.

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In generally it is believed that notochordomas are less sensitive to radiotherapy, but is also increasingly used as postoperative treatment which has better effect than the surgery alone.

Skull base notochordomas sometime could be very challenging and, frustrating tumors to treat because of their critical location, invasive nature and reocurrence rate. Ossama Al-Mefty and Luis Borba suggested both radical surgical removal and high-dose radiation therapy, particularly proton beam therapy, reportedly are effective in tumor control and improve survival rates. The mean disease-free interval was 14.4 months [7].

Nasopharyngeal chondroma when invade the surrounding tissue complete resection of the tumor become difficult. Most of the tumor most should be treated with radiotherapy (RT). It is believed that the risk of severe late complications and occurrence are low and cure rate high. Proton beam therapy alone or combined photon – proton beam therapy offers the advantage of improved dose distribution and the ability to treat the tumor to a higher dose without exceeding normal tissue tolerance. The 10-year local control rate after proton/photon RT is approximately 40% to 50% [8].

In another study by Feigl GC in 13 patients with 15 tumors first underwent maximal tumor resection. Within 2 to 10 months after surgery they were treated with Gamma knife surgery, GKS (mean treatment dose was 17 Gy and the mean dose was 52%), after the follow up for 17 months only one reocurrence was at the radiation margin so he encouraged the use treatment combining surgery and early GKS [9].

Prognosis of Nasopharyngeal notochordoma depends largely on the extent of surgical resection, postoperative recurrence is common, and so the prognosis is poor. In a study conducted by Georges Noël, Jean-Louis Habrand et al. in 67 consecutive patients chordoma and chondrosarcoma of the skull base and the cervical spine treated with irradiation combined high-energy photons and protons. Photons represented two thirds of the total dose and protons one third. The median total dose delivered within gross tumor volume was 67 Cobalt Gray Equivalents (CGE; range: 60–70 CGE), found that within a median follow-up the 3-year local control rates was 71% for chordomas the 3-year overall survival rate was 88% [10]. Complete resection is very difficult to achieve, reocurrence rate and metastasis after surgery is high. Complete resection is difficult to achieve due to tumor size, infiltrative nature, central location, and proximity to vital structures. Residual tumor is often unavoidable and invariably leads to recurrence. Tumor slippage during surgery is another cause of recurrence and may metastasis to lymph nodes, skin, lungs, liver and bones [2]. Nasopharyngeal notochordoma is a low-grade malignant tumor, selective postoperative radiotherapy is an effective method to increase the time interval to recurrence, radiation therapy offers excellent chances of cure.

Conclusion

Nasopharyngeal notochondroma is a rare disease in the department of otolaryngology. Because of it clinical features and site of occurrence is similar to nasopharyngeal carcinoma so some time the disease may be mis diagnosed so otolaryngologist should pay attention to this disease. The main stay of treatment is believed to be surgery although complete surgical resection of mass is difficult to achieve. In order to resect the tumor completely and reduce the complication and possible malformation, according to the radiological findings, pathologica changes and area involved different surgical approaches must be designed. Skull base chordoma is a rare neoplasm that is rarely cured after surgery alone. So to get better prognosis after surgery the surgeon should design the shortest approach, avoid damaging the important nasopharyngeal and intracranial structure, and resecting the total mass as possible.

From the above literature review and case report study we derived a conclusion that nasopharyngeal notochordoma are not completely resectable so the best possible treatment method should be surgical resection of mass along with radiotherapy (such as GKS or proton radian therapy, etc).

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Conflict of Interest

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References