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Case Report

Chordoma in the Nasopharynx- Treated with Radiotherapy after Surgical Resection

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Abstract

Chordoma is very rare bony tumour which arises from the notochordal remnants .Nine cases of Chordoma in the nasopharynx were identified in Rajshahi Medical College Hospital since January 1997 to December 2009. All the patients were non-diabetic and 2 of them were hypertensive. Their renal function tests and liver function tests were normal.

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Introduction

Chordoma is a malignant bone tumour (1). It arises from the embryonic remnants of the notochordal tissue. This tissue is normally situated within the vertebral bodies and intervertebral disc. It is found at any point along the axial skeleton. It presents mostly in the fifth and sixth decades of life but the disease starts in younger age group and both sexes are affected with male predominance. Fifty present of the chordoma arises form the sacrococcygeal area, 35% form the spheno-ocipital and rest form the cervico-thoracic spine (2). It is very rare below 40 years of age. It takes about five to ten years to develop the symptoms. Memorial Sloan-Kettering Institute identified 53 cases of chordoma in the sacrum, 24 in vertebral bodies and three in the spheno-occipital region. A high percentage [8.4%] of primary malignant tumours were reported to the National Cancer Institute's Surveillance Epidemiology and end Results [SEER] as chordoma between 1979 to 1987(3).

In Queen Mary Hospital, tumours in and around nasopharynx were identified and removed in 26 patients. Among them 18 suffered from nasopharyngeal carcinoma, three had Chordoma, two had Schwanoma and one had adenocarcinoma and one had malignant fibrous histeocytoma(4). Vollrath in Germany reported two cases of chordoma in the nasopharynx and categorized the chodoma as clival, cervial and sacrococcygeal(5).

Spheno-occipital chordoma may appear with nasal, paranasal or nasophsryngeal mass. It is hard on palpation and may create pressure symptoms. Multiple Cranial nerves may be involved. Chordoma is gelatinous and contains areas of haemorrhages. Microscopically it resembles normal notochordal tissue. Histologically the physeliferous cells are pathognomonic. The tumor in the spine destroys vertebral bodies and arches and can bulge into subdural space.

Clinical diagnosis is made by symptoms, signs and involvement of the surrounding structures and finally by the x-rays, CT scan. A soft tissue mass is essential to the radiological diagnosis, with a variable degree of destruction of the vertebrae. CT or MR Scans are invaluable for the demonstration of the extent of bone destruction and the extent of soft tissue mass(7).

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Patients and Methods

A total of 9 patients reported to Rajshahi Medical College Hospital since January,1997 to December,2009 with the complaints of headache, nasal obstruction, feeling of mass in the throat and occasional bleeding from the nose. Patients also complain that something was coming out from behind the soft palate which obstructed the nasal passage. A thorough clinical examination was done of the patients and focal neurological signs detected. X-ray of the nasopharynx lateral view showed a mass in the nasopharynx completely obstructing the air passage. CT scan of the base of the skull showed a tumuor is attached to the body of C1 and C2 vertebrae in all the cases. All other investigations were normal. All the patients were non-diabetic and 2 of them were hypertensive. Their renal function tests and liver function tests were normal.

Results

Excision of the tumuor of all the patients was done under general anesthesia. Specimens were sent for histopathology and was reported as chordoma.

The patients were then treated with radiotherapy in the department of Radiotherapy at Rajshahi medical college hospital. A total of 6000 cGy was given in 30 fractions to all the patients and the patients were tolerated well. After 6 months of radiotherapy 3 of the patients developed recurrence and they were treated with chemotherapy.

Discussion

Chordoma is a low potentiality malignant embryonal bone tumor. It arises from the notochordal remnants. The ends of the spines are the most common site of its origin. They look like soft, gray colured and multiloculated masses by posterior rhinoscopic examination. Cranio-cervical chordoma frequently presents as a nasopharngeal mass. Most of the tumour in the nasopharynx are thought to be carcinoma. But it must be distinguished form chordoma or chondrosarcoma. Incase of carcinoma of the nasopharynx metastatic lymphnodes in the neck is an early presentation whereas chordoma has characteristic of late matastases. Invasion of spinal canal by chordoma may cause neurological complictions. In one series the frequency of metastases was 43%(7). The most common sites are the skin and bones but they may occur in any place in the body (8). Treatment of chordoma consists of complete resection of the tumour followed by radiation therapy.

German Society Vollrath compared the result of operation and radiation therapy and found that each alone fails because of the high recurrence rate. Hence they preferred combination of surgery & radiotherapy. One of his patients who was treated with both radiation therapy and surgery had a survival period of 14 years (9). Thirteen patients were treated in the department of Radiotherapy ,University of Cologne, Koeln, Germany histopathology revealed carcinoma arising from the nasopharynx ; 8 chordoma 1, rabdomyosarcoma-1, chordosarcoma-1 and haemangiopericytoma-2. All patients had repeated tumour resection or irradiation, hindering any further conventional fractionated radiotherapy or surgery (10). But overall prognosis was poor.

Because of the location of chordoma in the base of the skull, removal of the tumour is very difficult and usually partial removal is done. Transcervical and transmandibular approach to the skull base can be employed in removing this tumour (11). In Russian Academy of Medical Sciences, Moscow a transoral approach was used in patients with tumours of the clivas, a chordoma and another chordosarcoma. Choice of the approach was based on data provided by clinical and radiographic examinations(12).Cryosurgery with liquid nitrogen is occasionally used when complete removal of the tissue is not possidle (13). Orthopaedic surgeon should be included in the surgical team. Help of neurosurgeons asked for when there is intracranial extension. Digital palpation is sometimes helpful in differentiating chordoma from other spinal tumours. X-ray of nasopharynx lateral view, CT scan, MIR, FNAC and finally excision biopsy are essential requirement for accurate evaluation of chordoma. Vertebral angiogaphy is helpful in demonstrating the tumours by vessel displacement, encasement and vascular staining (14).

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