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Chordoma in the Nasopharynx- Reports of Two Cases

MA SHAIK[a], AKM N ISLAM[b] , T CHAKRAVORTY[c] , MS ISLAM[d], A CHOUDHURY[e]

Summary

Chordoma is a very rare bony tumour which arises from the notochordal remnants .Two cases of Chordoma in the nasopharynx were identified in the ENT Department of Bangladesh Medical College Hospital January 1992 to December 2002.Both the patients were old women. Tumour from nasopharynx were removed through the oral route. The Diagnosis was confirmed by the histopathology examination which showed chordoma. A full course of radiotherapy was given in both the cases and the patients improved rapidly.

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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 interverteibral disc. It is found at any point along the axial skeleton. It is present mostly in the fifth and sixth decades of life and in both sexes equally. Fifty present of the chordoma arises from the sacrococcygeal area , 35% from the spheno-ocipital and rest from 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 Surveillence Epidemiology and End Results[SEER] as chordoma between 1973 to 1987[3].

a. Dr. Md. Alauddin Shaik DLO, FAMS, Professor and Head of the Dept. of ENT and Head Neck Surgery, Bangladesh Medical College and Hospital, Dhaka.
b. Dr. AKM Nazrul Islam FCPS, Assistant Professor. of Surgery, Bangladesh Medical College and Hospital, Dhaka.
c. Dr. Tapas Chakravorty DLO, Registrar, Dept. of ENT and Head Neck Surgery, Bangladesh Medical College and Hospital, Dhaka.
d. Dr. Md. Sirajul Islam MBBS, Assistant Registrar, Dept. of ENT and Head Neck Surgery, Bangladesh Medical College and Hospital, Dhaka.
e. Dr. Azim Choudhury, MBBS, Assistant Registrar, Dept. of ENT and Head Neck Surgery, Bangladesh Medical College and Hospital, Dhaka.

Address of Correspondence : Dr. Md. Alauddin Shaik MBBS, DLO,FAMS, Prof and Head of the Dept. of ENT and Head Neck Surgery, Bangladesh Medical College and Hospital, Dhaka.

In Queen Mary Hospital, Hongkong, 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 chordoma as clival, cervical and sacrococcygeal[5].

Spheno-occipital chordoma may appear with nasal, paranasal or nasopharyngeal 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, MRI and myologram. 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].

Case reports

Case1:

Mrs. JB, 50 years of age, was admitted in ENT department of Bangladesh Medical College Hospital on 19.3.2001 with the complaints of nasal obstruction, feeling of a mass in the throat and occasional bleeding from the nose. She experienced

these problems since previous one year. She observed that something was coming out from behind the soft palate which obstructed her nasal passage. She had snoring during sleep. She also experienced bleeding from the nose three times in previous one year. A thorough clinical examination was done. X-ray of nasopharynx lateral view showed a mass in the nasopharynx completely obstructing the air passage. CT scan of base of the skull showed a tumour attached to the body of C1 and C2 vertebrae. All other routine investigations were normal. She was non-diabetic and non-hypertensive. Her renal and liver function tests were normal.

Excision of the tumour was done under general anaesthesia. The patient was put in tonsillar position by applying a sand bag below the shoulder. The tumour was firm on palpation. The soft palate was lifted up and the tumour was dissected from the body of the vertebrae. The bleeding was controlled by diathermy and a post nasal pack was applied. The specimen was sent for histopathology and was reported to be a chordoma.

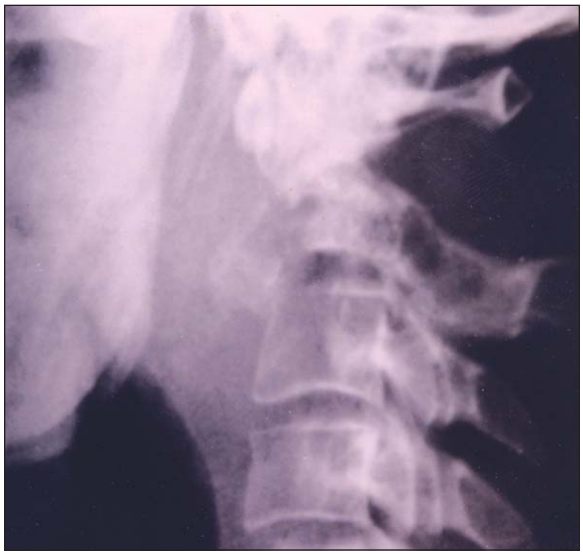

Fig.-1 : *Chordoma of 2nd cervical vertivra body of C2 is destroyed and large paravertileral mass is seen.*

Case- 2:

Mrs. DB, 45 years, was admitted in the ENT ward of Bangladesh Medical College Hospital on 24.10.1999 with history of nasal obstruction and occasional bleeding from nose for the previous six months. A thorough clinical examination and routine investigations were done. X-ray of nasopharynx lateral view showed a tumour in the nasopharynx. All other investigations were normal. She was non-diabetic and non-hypertensive. Her liver and kidney functions were normal.

Excision of the tumour was done through the oral route under general anaesthesia. The tumour was 2.5 x 3.5 cm in size and there was some erosion over the surface. The specimen was sent for histopathology and was reported to be chordoma. The wound healed smoothly within three weeks.

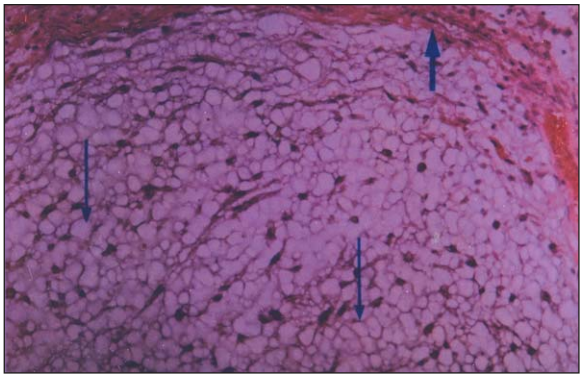

Fig.-2 : *Miroscopic picture of chordoma. the this arrow show large vacides with pale grey maleviales.*

Discussion:

Chordoma is a malignant bone tumour. It arises from the notochordal remnants. The ends of the spines are the most common site of its origin. They look like soft, gray coloured and multiloculated masses. Cervical chordoma frequently presents as a pharyngeal mass. Most of the tumour in the nasopharynx are thought to be carcinoma. But it must be distinguished from chordoma or chondrosarcoma. In case 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 complications. 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 & rediotherapy. 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, chordrosarcoma-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 two 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 possible [13]. Orthopaedic surgeon should be included in the surgical team. Help of neurosurgeons is 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, MRI, FNAC and finally excision biopsy are essential requirement for accurate evaluation of chordoma. Vertebral angiography is helpful in demonstrating the tumours by vessel displacement, encasement and vascular staining [14].

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