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Thoracic Chordoma of Thoracal 7th-8th: A Case Report

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Abstract. Spinal chordomas are rare, locally invasive, malignant neoplasm, representing 5% of all malignant tumors of the skeleton. This publication is based on a case of a thoracal chordoma in a 53 year old man at Haji Adam Malik General Hospital Medan. Patient with a major complaint of back pain accompanied by weakness in both lower limbs, beginning with numbness and tingling sensation. Based on physical examination, thoracal CT-Scan and Thoracal MRI, diagnosis of a thoracal chordoma was made. Laboratory was normal. There was no other comorbidities. Surgical treatment was applied in this case. The patient performed laminectomy tumor removal on throracal 7th-8th, followed with posterior stabilization on thoracal 5th-6th and thoracal 9th-10th. We managed to achieve near complete macroscopic tumor resection in this case. Tumor samples were taken for Histopathologhy analysis. From the microscopy finding we found a typical cell of a chordoma.On the 7th day the patient was discharged. The patient then controls for follow up and radiotherapy. Patient neurologically improved, no complications and no surgical implants failures in this case. After surgery, the patient underwent radiotherapy. The main goal of therapy in this chordoma case is total eradication, spinal decompression without neurologic impairment and prevention of recurrence.

Keyword: Chordoma, Neurosurgery, Spine, Thoracal

Abstrak. *Kordoma tulang belakang bersifat jarang,cenderung merupakan neoplasma ganas dan invasif lokal, mewakili 5% dari semua tumor ganas kerangka. Publikasi ini didasarkan pada kasus chordoma thoracal pada pria berusia 53 tahun di Rumah Sakit Umum Haji Adam Malik Medan. Pasien dengan keluhan utama nyeri punggung disertai dengan kelemahan pada kedua tungkai bawah, dimulai dengan mati rasa dan kesemutan. Berdasarkan pemeriksaan fisik, CT-Scan thoracal dan Thoracal MRI, diagnosis chordoma thoracal dibuat. Laboratorium normal. Tidak ada komorbiditas lain. Perawatan bedah diterapkan dalam kasus ini. Pasien melakukan pengangkatan tumor laminektomi pada throracal 7-8, diikuti dengan stabilisasi posterior pada thoracal 5-6 dan thoracal 9-10. Kami berhasil mencapai reseksi tumor makroskopik yang hampir lengkap dalam kasus ini. Sampel tumor diambil untuk analisis histopatologi. Dari hasil mikroskop kami menemukan sel khas chordoma. Pada hari ke 7 pasien dipulangkan. Pasien kemudian kontrol untuk tindak lanjut dan radioterapi. Pasien membaik secara neurologis, tidak ada komplikasi dan tidak ada kegagalan implan bedah dalam kasus ini. Setelah operasi, pasien menjalani radioterapi. Tujuan utama terapi dalam kasus chordoma ini adalah eradikasi total, dekompresi tulang belakang tanpa gangguan neurologis dan pencegahan kekambuhan*.

Kata Kunci: *Bedah saraf, Kordoma, Thorakal, Tulang belakang,*

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1 Introduction

Chordomas were first characterised microscopically by Virchow in 1857. He described unique, intracellular, bubble-like vacuoles that he referred to as physaliferous, a term now synonymous with their histopathology. These physaliferous features of chordoma remain a distinguishing, if not pathognomonic, feature. Virchow hypothesised that chordomas were derived from cartilage; however, more contemporary evidence suggests that they are derived from undiff erentiated notochordal remnants that reside within the vertebral bodies and throughout the axial skeleton. In fact, Ribbert first introduced the term chordoma in the 1890s, in view of the notochord hypothesis. [1][2][3]

Examination of human embryos and fetuses and cell-fate-tracking experiments in mice showed that notochordal cell nests topographically correspond and distribute to the sites of occurrence of chordoma. Although there is little direct evidence that cells transform to chordoma, molecular phenotyping of these primitive rests compared with neoplastic lesions suggests they are indeed the likely source for transformation**.** Spinal chordomas are rare, locally invasive, malignant neoplasm, representing 5% of all malignant tumors of the skeleton.The male:female prevalence ratio is 2:1 with an increasing incidence after the fourth decade. These lesions arise from notochordal remnants within the vertebral bodies The main affected segment is the sacrum, followed by the spheno-occipital region and only in rare cases the mobile spine. [1][3][4]

Chordomas are slow-growing tumors with aggressive local behavior that probably originate from embryonic remnants of the notochord. This is a midline structure along the neural tube that presents involution and fragmentation as ossification of the axial skeleton occurs, in such way that in the second month of intrauterine life, it is restricted to the intervertebral discs, where the nucleus pulposus will appear. Other remnants of the notochord originating outside the discs and within the bones can be found in any topography long the vertebral column but they are preferably found in the sacrococcygeal region (50 to 60%) or in the sphenobasilar region (25 to 40% of the cases). In the remaining portion of the vertebral column (15%) the main localization is the cervix, especially the high cervical spine, followed by the lumbar spine. [2][3][5]

Chordomas of the thoracic spine are rare and usually present as extradural bone tumors that lead to complaints of pain by instability and intercostal radiculalgia or neurological changes in the lower limbs due to spinal cord compression. [3][6]

2 Case Report

This publication is based on a case of a thoracal chordoma in a 53 year old man at Haji Adam Malik General Hospital Medan. Patient with a major complaint of back pain accompanied by weakness in both lower limbs, beginning with numbness and tingling sensation. Based on

physical examination, thoracal CT-Scan and Thoracal MRI, diagnosis of a thoracal chordoma was made. Laboratory was normal. There was no other comorbidities. Surgical treatment was applied in this case.

3 Results

The most compelling evidence of the notochordal hypothesis was the discovery of a gene duplication in the transcription factor T gene (brachyury) in familial chordoma. An important transcription factor in notochord development, brachyury is expressed in normal, undiff erentiated embryonic notochord in the axial skeleton. High-resolution array comparative genomic hybridisation showed unique duplications in the 6q27 region in tumour samples from patients with familial chordoma. This duplicated region contained only the brachyury gene, which was known to be uniquely overexpressed in almost all sporadic chordomas compared with other bone or cartilaginous lesions.Brachyury regulates several compelling stem-cell genes and has recently been implicated in promoting epithelial–mesenchymal transition in other human carcinomas. [7][8][9]

The clinical manifestation of the thoracic chordomas is usually insidious and progressive, with signs of pain by spine instability, a strip of radiculalgia by compression of the thoracic roots or syndromes by compression of the spinal cord by the tumor. These pictures of myelopathy may worsen suddenly when a pathological fracture of the vertebra happens, with sudden reduction of the spinal canal. Sympathetic involvement in upper thoracic lesions was described in an 18-year old patient whose expression of a chordoma between T1 and T2 was a Claude-BernardHorner syndrome . Recurrent dry cough was the clinical manifestation of a chordoma in a 14-year old patient, with a cystic form of the tumor growing from T3 and T4 which extended to the upper mediastine, posteriorly. Eight similar cases are reported in the literature. [7][8][9]

Treatment options for chordomas include surgical resection, radiotherapy, and chemotherapy. Radiotherapy is most often used in patients with local recurrence, while chemotherapy is usually reserved for patients with metastasis. Surgical resection is associated with a significant difference in survival compared to those who do not undergo surgery. En bloc surgical resection has been the optimal treatment modality since it was first introduced by Stener and Gunterberg in the 1970s. In 2016, Lee et al., using the SEER database, found that the overall survival of 1,593 patients was 61% at five years and 41% at 10 years. [8][9][10]

The patient performed laminectomy tumor removal on throracal 7^{th}-8^{th}, followed with posterior stabilization on thoracal 5^{th}-6^{th} and thoracal 9^{th}-10^{th}. We managed to achieve near complete macroscopic tumor resection in this case. Tumor samples were taken for Histopathologhy analysis. From the microscopy finding we found a typical cell of a chordoma with intercellular myxoid matrix and polygonal cells with abundant cytoplasm.On the 7th day the patient was

discharged. The patient then controls for follow up and radiotherapy. Patient neurologically improved, no complications and no surgical implants failures in this case

4 Conclusion

A Rare case of Thoracal chordoma in male 53 years old was reported. Patient is treated with a posterior approach and posterior stabilization. After surgery, the patient underwent radiotherapy. The main goal of therapy in this chordoma case is total eradication, spinal decompression without neurologic impairment and prevention of recurrence.[10][11]

Surgical resection remains the first choice for the skull base with the appropriate surgical approach based on tumor size and location. Given the irinvasive nature with spread along critical bony and neural structures, and large tumor burden, complete resection of these tumors is often difficult. Surgery should aim towards maximally safe cytoreductive surgery with wide enbloc resection with preservation of neurological function and quality of life, even at the price of postoperative residual tumor. Within the constraints to safety and minimizing complications, a particular effort should be made to obtain the maximal surgical reduction of the lesion and clearance from eloquent structures even to repeating further surgery. The reduction of the burden of tumor and the abutting to critical structures can also favor the safer delivery of high doses of irradiation.[12][13]

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