

Lumbar spine chordoma in adolescent: Case report.

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Abstract

Chordoma is a rare slow-growing neoplasm that arises from primitive notochordal remnants with a high rate of recurrence. It can occur anywhere within the spine or the base of the skull. Chordoma involving lumbar spine are rare, approximately 6% of spinal chordomas originate in the lumbar vertebrae. Prognosis is better than adults and depends on the extent of surgical resection, age and histology subgroup. We report a case with solitary L2/L3 vertebral body lesion causing caudal compression and bilateral lower limb weakness treated with L2/L3 right hemilaminectomy and tumor decompression.

Keywords: Chordoma, Lumbar spine, Surgical treatment.

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Introduction

Chordomas are rare low grade malignancies that represent approximately 1% of all intra cranial tumors and 4% of bony primaries [1-3]. They develop from notochordal embryonic residues [4]. Only 5% of cases have been described in the pediatric age [5-7]. We report in this paper a clinical case of lumbar spine Chordoma in an adolescent.

Case Report

A 15-year-male patient came to our consultation department as he was referred to us by neurosurgeon and histopathologist for further assessment and management.

The mother gave a history of sudden onset of inability to walk and severe bilateral lower limb pain for 2 months duration that made him awake from sleep which was associated with paresthesia and numbness that was not responding to regular analgesics. He was under treatment with a rheumatologist, but without much benefit. An X-ray was done followed by Magnetic Resonance Imaging (MRI) which revealed a lesion in L2/L3 vertebral body (Figure 1).

He was assessed by a neurosurgeon and underwent right hemi laminectomy and tumor decompression in India (due to unavailability of advanced neurosurgical center in Iraq).

His histopathology and immunohistochemistry revealed chordoma (CK AE1+AE3 positive, S-100 patchy positive).

Regarding the antenatal care there was no problem but the patient was premature and low birth weight which needed admission in NICU. There is a consanguinity of parents with non-significant family history, and middle to low socioeconomic status. The patient has a history of appendectomy and some hearing problem and no known drug history.

On post-operative examination the patient was conscious, alert, afebrile not bed ridden and well looking with vital signs. Normal patient was reevaluated with an MRI (Figure 2).

The patient underwent postoperative radiotherapy within 34 days after surgery for 27 fractions (2000cGy/fraction) of total

5400 cGy and was under regular close follow up (Figure 2).

The patient underwent Computerized Tomography (CT) scan of brain to exclude any metastasis which was normal (Figure 3), also there were no sign of metastasis in his chest X-ray at his first evaluation and last follow up.

On regular follow up and after approximately 3.5 months he developed the sign and symptoms as before, but the severity was less. There was mild to moderate lower limb weakness and back pain with inability to walk.

But because the surgery is not done in our country so we put him on supportive treatment (short course of dexamethasone and Non-Steroidal Anti-Inflammatory Drugs (NSAID)) and follow up.

A new MRI was done which showed recurrence; he was put on supportive treatment (short course of dexamethasone and NSAID) and follow up (Figure 4).



Figure 1. Large cystic mass at level of L2/L3 vertebral body.



Figure 2. Postoperative and post radiation MRI (No residual mass only postoperative changes).

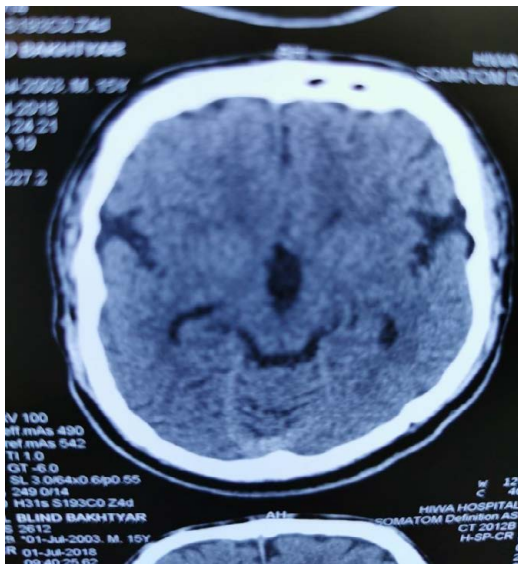


Figure 3. Normal CT scan of brain.



Figure 4. MRI of lumbosacral spine showing postoperative laminectomy at level L1/L2 and extra dural enhancing soft tissue extending to right neural foramina without nerve compression.

Discussion

Paediatric chordomas are rare malignant tumors which involve the cranial base and spinal column. They seem to be different from their adult counterparts. They develop from notochordal embryonic residues [4]. Only 5% of cases have been described in the pediatric age [5-7]. Mean age varies according to series between 8 and 12 years, most approaching 10 years [7,8]. Sex ratio is close to 1, although some series evidence a slight female predominance [9,10]. In our case, L3 and L4 vertebra bodies were involved.

Their anatomopathological classification is the same for adults and children and distinguishes “classic” (“chordoma NOS” Not Otherwise Specified), “chondroid” and “undifferentiated” chordomas. These tumors are slowly growing and the symptoms are closely related to the site of tumor and tumor compressing the root, spinal cord or cauda equina and paravertebral tissues.

Paresthesia and pain are most common complaints in patients with lumbar chordomas and usually take long time before any other neurological symptoms occur. The rates of local recurrence and metastasis depend on the quality of the initial surgery, pathological features, and age of the patient.

Because of their location, lumbar chordomas may easily be confused with more common tumors in the lumbar spine such as aneurysmal bone cyst, giant cell tumor, hemangioma, myeloma and metastasis, and preoperative diagnosis can be difficult [11]. CT and MRI are currently used in the diagnosis of chordomas. MRI signals of the mass are commonly isointense or hypointense on T1-weighted images and hyperintense on T2-weighted images.

Metastatic dissemination primarily occurs *via* the blood circulation, but also *via* the cerebrospinal fluid, either *via* the subarachnoid spaces [12,13] or through ventricular shunting [14]. The most common site of metastasis is lung and lymph nodes remove also, bone and liver are common sites of metastasis.

Surgical resection, radiation therapy, chemotherapy are the current proposed therapy modalities for the spinal chordomas. Prognosis and survival are better in children than in adults except for the aggressive form of chordomas occurring in children under 5 years of age. Prognosis depends on histological subtype, site of tumor and finally age at onset of tumor.

Our patient was complaining of sudden onset of inability to walk and severe bilateral lower limb pain for 2 months duration. Patient underwent complete surgical resection of tumor with laminectomy and high dose irradiation, but unfortunately the tumor recurred with the same symptoms but less aggressively and neurosurgeon suggested managing conservatively because of the difficulty to re-operate. Low dose chemotherapy or targeted therapies as adjuvant treatment was advised but not implemented because of rarity of disease and both lines being refused by patient’s family.

Conclusion

Pediatric chordomas are rare malignant tumors with a high rate of recurrence. The surgical removal of the tumor still is the first step of the treatment followed by radiotherapy and the use of

adjuvant chemotherapy or targeted therapies are controversial and have not been proven. Our patient underwent surgical resection of tumor and received irradiation but unfortunately the mass recurred and we proposed him chemotherapy or adjuvant target therapies but the family refused so we put him on close regular follow up hop.

Consent

Written informed consent was obtained from patient's parent for this case report.

Acknowledgments

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Authors' Contributions

Consultant Dr. Basil Kadhim Abdallah Al oada has been involved in clinical diagnostic evaluations and management.

Conflict of Interest

The authors declare that they have no conflict of interests.

Ethical Consideration

Ethical approval for this case report was obtained from NCI ethical committee. Because of the retrospective nature of this case report, the ethical committee waived the requirement of informed consent.

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