Magnetic Resonance Imaging Profile of Chordomas - A Retrospective Evaluation from a Tertiary Institution of Eastern India

Bhaskar Bhattacharyya[1], Amit Kumar Das[2], Arijit Roy[3], Aprateem Mukherjee[4], Tapan Dhibar[5], Kiran Sankar Saha[6], Amitabha Chattopadhyay[7]

[1, 2, 4] Department of Radio Diagnosis, R.G. Kar Medical College, Kolkata, West Bengal, India. [3]Department of Neuro Medicine, Bankura Sammilani Medical College, Bankura, West Bengal, India. [5]Department of Radiodiagnosis, Bangur Institute of Neurosciences, Institute of Post Graduate Medical and Research, Kolkata, West Bengal, India. [6]Department of Radiodiagnosis, Institute of Post Graduate Medical Education and Research, Kolkata, West Bengal, India. [7]Department of Pathology, Institute of Post Graduate Medical Education and Research, Kolkata, West Bengal, India.

ABSTRACT

BACKGROUND

Chordomas are tumours presumed to originate from the remnants of embryonic notochord. They present a diagnostic challenge as they can occur in any region within the craniospinal axis and are rare with an incidence of less than 0.1 / 100,000 per year. Imaging is required for their evaluation and magnetic resonance imaging (MRI) is the imaging modality of choice due to excellent soft tissue resolution, multiplanar imaging capabilities and precise anatomical delineation.

METHODS

MRI scans of 10 patients, who had proven chordomas histologically, were evaluated retrospectively in the Department of Radiodiagnosis, Bangur Institute of Neurosciences from July 2012 to June 2018. Patients without histological proof of chordomas and those lost to follow-up were not included in the study. Clinical information such as age, sex, presenting symptoms were noted. Imaging parameters assessed included tumour location, bone destruction, extraspinal soft tissue component, spinal canal and neural foramina encroachment, arterial encasement, signal intensity, morphology and enhancement pattern by MRI. All the cases were scanned in a 1.5 Tesla MRI machine. Intravenous contrast were used in all cases.

RESULTS

Of the 10 cases, 7 were male and 3 were female. There were 7 cases of sacrococcygeal chordomas and 3 cases of clival chordomas. The tumours appeared as multilobulated masses with bone destruction. All sacrococcygeal lesions involved more than one vertebral segment with extraspinal soft tissue component encroaching the pelvic cavity and showed cystic spaces containing hypointense septae. All 3 cases of clival chordomas showed compression of the pons and basilar artery. In one case there was compression of optic chiasma with displacement of internal carotid arteries laterally. On T1 weighted imaging, the chordomas were isointense in 4 and hypointense in 6 cases. These tumours were hyperintense in T2WI in 8 and intermediate to high in signal intensity in 2 cases. All demonstrated heterogenous enhancement on contrast with moderate enhancement in 8 patients and mild enhancement in 2 cases.

CONCLUSIONS

Chordomas are rare tumours that can occur anywhere in the craniospinal axis. MRI is the modality of choice for imaging of chordomas. Characteristic findings in MRI include low to intermediate signal intensity on T1w images and high signal intensity on T2w images. Sacral chordomas have T2 hyperintense cystic masses with hypointense septa. Enhancement is heterogenous ranging from mild to moderate. MRI is invaluable for a pre-operative diagnosis, delineation of tumour extent and as roadmap for surgery and radiotherapy.

KEYWORDS

Chordomas, Magnetic Resonance Imaging, Clivus, Sacrococcygeal

Corresponding Author:
Dr. Aprateem Mukherjee,
Flat - 2A, 62A Peary Mohan Roy Road,
Kolkata -700027, West Bengal, India.
E-mail: aprateem@gmail.com

DOI: 10.18410/jebmh/2020/561

How to Cite This Article:
Bhattacharyya B, Das AK, Roy A, et al. Magnetic resonance imaging profile of chordomas: a retrospective evaluation from a tertiary institution of eastern India. J Evid Based Med Healthc 2020; 7(46), 2730-2734. DOI: 10.18410/jebmh/2020/561

Submission 25-08-2020,
Peer Review 31-08-2020,
Acceptance 09-10-2020,
Published 16-11-2020.

BACKGROUND

Chordomas are rare malignant tumours of the axial skeleton and skull base, having an incidence of less than 0.1 / 100,000 per year.[1] It is presumed that chordomas develop from the embryonic remnants of the primitive notochord.[2] They are considered to be locally aggressive tumours with propensity to invade adjacent structures and cause bone destruction. Magnetic resonance imaging is the imaging modality of choice as it allows precise delineation of the tumour and its relationship to adjacent soft tissue structures, thus helping in the diagnostic work-up and in the planning prior to surgery, which is the mainstay of treatment. In this paper we describe a series of cases of chordoma involving the craniospinal axis and discuss its imaging features.

We wanted to study the MRI characteristics of chordomas and evaluate their anatomical location, and demographics.

METHODS

This retrospective study evaluated case records of MR imaging in Dept. of Radiodiagnosis, Bangur Institute of Neurosciences**,** between July 2012 to June 2018. A total of 10 patients of histologically proven chordomas who underwent surgery on the basis of preoperative MRI were included in this study. Histological proof of chordoma was taken as inclusion criteria. Patients without histological proof of chordomas and those lost to follow-up were excluded from the study. Clinical information such as age of presentation, sex of patient, presenting symptoms were also noted.

All the patients were evaluated in a 1.5 Tesla MRI scanner. Pre-contrast T1 and T2 weighted sequences followed by post-contrast T1 weighted sequence with fat saturation using 0.1 mmol / kg intravenous gadolinium were used in all cases.

Image Analysis

On imaging the parameters evaluated were: location of involvement, morphology (uni-lobular or multi-lobular), bone destruction, extraspinal soft tissue component, encroachment of adjoining sacral spinal canal and neural foramina, arterial encasement, signal intensity and enhancement pattern by MRI. Signal intensity (SI) in MRI was compared with that of spinal cord, fat and cerebrospinal fluid (CSF) on T1 and T2 weighted images. On T1-weighted images (T1WI), it was considered as hypointense if the SI was less than SI of spinal cord, intermediate if SI was between SI of spinal cord and fat or hyperintense if SI was similar to fat. On T2-weighted images (T2WI), it was classified as hypointense if SI was less than SI of spinal cord, intermediate if SI was between spinal cord and CSF, hyperintense if SI was similar to CSF. Cystic spaces with hypointense septa was evaluated on T2-weighted sequences. On post contrast T1 fat saturated images, the degree of enhancement was evaluated as being mild, moderate or marked by comparing SI to that of muscle. The patterns of contrast enhancement were also observed as uniform or heterogenous.

RESULTS

In our study we had 7 male and 3 female patients. The patients' age were in the range of 27 years to 53 years. The mean age was 40.3 years. Chordomas were located in the sacrococcygeal region in 7 cases and in clivus in 3 cases.

Of the 3 patients with clival chordomas, 2 of them presented with headache and diplopia. One of them presented with bitemporal hemianopia and dysphagia. All the 7 cases of sacrococcygeal chordoma presented with low back pain. 2 of them had radicular pain in lower limbs additionally, while 1 patient also presented with constipation and urinary incontinence.

On imaging all 10 cases had a multilobulated appearance morphologically. Evidence of bone destruction were detected in all. Two cases had a CT (Computed Tomography) scan which also confirmed the nature of bone destruction. All 7 sacrococcygeal lesions involved more than one vertebral segments, the most extensive lesion involving both sacrum (S2 downwards) and coccyx. Extraspinal soft tissue component was seen predominantly in the ventral aspect, encroaching the pelvic cavity in all 7 cases of sacrococcygeal chordomas. The adjoining pelvic organs were compressed and displaced but not involved. In addition, the encroachment of adjoining sacral spinal canal and neural foramina were well demonstrated in MRI in 3 cases. The posterior paraspinal muscles were involved in 3 cases. In one case the sacrococcygeal lesion extended through sciatic foramen into gluteal region on left side. No intrapelvic enlarged lymph node was seen in these patients. Cystic spaces with hypointense septae were seen all cases of sacrococcygeal chordomas.

Age Range	27 - 53 years
Sex	Males = 7, Females = 3
Location	Clival (n = 3) Sacrococcygeal (n = 7)
Presenting clinical features Clival chordomas	Headache, diplopia (n = 2) Bitemporal hemianopia, dysphagia (n = 1)
Sacral chordomas	Low back pain (n = 7) Radicular pain (n = 2) Incontinence, constipation (n = 1)
Bone destruction	n = 10
Imaging T1	Hypointense (n = 6) Isointense (n = 4)
T2	Hyperintense (n = 8) Intermediate-hyperintense (n = 2)
Post contrast	Heterogenous moderate (n = 8) Heterogenous mild (n = 2)
Cystic spaces and septae	n = 7

Table 1. Summary of Findings

In the 3 cases of clival chordomas the pons and basilar artery were compressed as the mass encroached into prepontine cistern. In one patient the clival lesion was larger which compressed the optic chiasma, displaced the adjoining internal carotid artery segments laterally on either

side and destroyed the floor of anterior cranial fossa. However, there was no invasion of brain and pituitary. There was no cystic change or septation in these clival lesions.

On T1WI the chordomas were isointense in 4 and hypointense in 6 cases. These tumours were hyperintense in T2WI in 8 and intermediate to hyperintense in signal intensity in 2 cases. All of them demonstrated heterogenous enhancement following contrast administration. The enhancement was moderate in 8 patients, while 2 showed mild enhancement.

A presurgical diagnosis of chordoma was concluded based on MRI features in all the cases. All of the cases were managed surgically and histologically proven as chordomas.

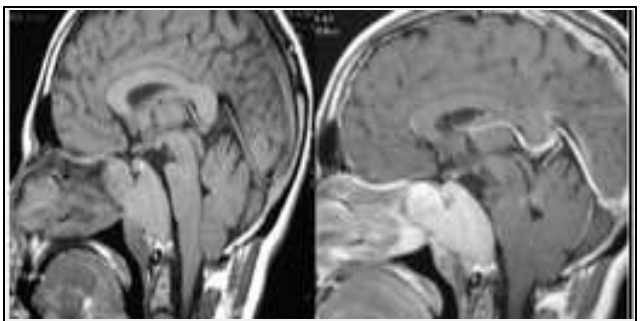

Figure 1. Sagittal T1 (Pre-Contrast) and T1 (Post-Contrast) Images Showing a Lobulated Clival Chordoma with Moderate Enhancement

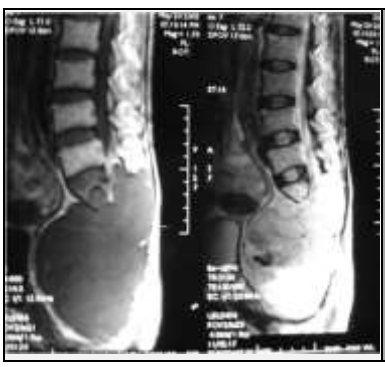

Figure 2. Sagittal T1 and T2 Weighted Images of Lumbosacral Spine Showing a T1 Hypointense, T2 Hyperintense Mass in the Sacrococcygeal Region with Destruction of Sacrum – A Chordoma

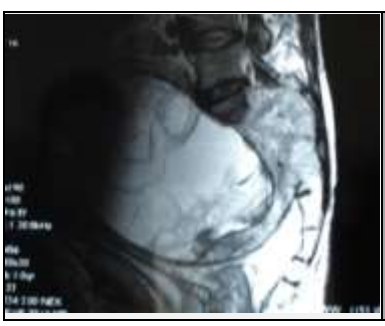

Figure 3. A Sagittal T2 Weighted Image of the Sacral Region Showing a High Signal Intensity Mass. Not the Presence of Hypointense Septa within It. (Sacrococcygeal Chordoma)

DISCUSSION

Chordomas are relatively rare bone tumours that are presumed to arise from the embryonic notochord, which is a primitive cell line around which the vertebral column and skull base develops. Although chordomas are slow growing, they are invasive and locally destructive.

The male to female ratio is 2:1.[3] Chordomas are found mainly in adults and uncommon below the age of 30 years. The average age at diagnosis is 48 years. Sacrococcygeal region is the most common site where 50 % chordomas occur. About 35 % occur is clivus and 15 % in the vertebrae involving cervical, lumbar and thoracic spine in descending order of frequency.[4,5] Of the vertebrae axis (C2) is most commonly affected.[6]

Chordomas represent 3 % - 4 % of primary bone tumours.[4] Clival chordomas constitute less than 0.2 % of all intracranial tumours.[7] Chordoma is the most common primary sacral neoplasm.[8] More than one vertebral segment is often involved.

The clinical presentation depends on location, local pain being the most common symptom.[9] Constipation is the next most frequent symptom in sacrococcygeal chordomas which may also present with radicular leg pain and urinary complaints. Features of cranial nerve deficits, most commonly diplopia, may occur in clival chordomas.[10] Sixth cranial nerve is involved most commonly. Clinical manifestations were similar in our study group, with 2 patients with clival chordomas presented with headache and diplopia. Another one presented with bitemporal hemianopia and dysphagia. All the cases of sacrococcygeal chordoma presented with low back pain, while 2 of them had radicular pain in lower limbs additionally. One patient with sacral chordoma also presented with constipation and urinary incontinence.

Radiological evaluation of chordomas is crucial, not only for the diagnosis but also as part of pre-treatment workup. Radiological evaluation has improved with modern imaging methods such as CT scan and MRI, however MR has been demonstrated to the single best imaging modality for imaging evaluation of intracranial chordomas.[11] Tumour morphology, extension into surrounding structures, relationship of tumours to surrounding vital structures in brain and spine is very well delineated with MRI. This helps not only the surgical team in planning the resection of the lesion, but also the radiation oncologist who needs to know the tumour extent and neighboring vital structures such as nerves and vessels, for planning further therapy.

MRI provides excellent soft tissue contrast and anatomical information. The multiplanar capabilities of MR helps accurately determine the extent of the tumour. Sagittal images depict the posterior extension of tumour, relationship with brainstem and basilar artery and nasopharyngeal extension in case of clival chordomas, levels of vertebrae involvement, disc involvement, cord involvement in case of spinal chordomas. Coronal MR images help to determine cavernous sinus involvement and the relationship of the tumour to optic chiasma. MRI even without contrast is able to demonstrate major blood vessels such as basilar and internal carotid arteries, as the vessels appear as flow voids.

The clivus is the sloping part of skull base, posterior to dorsum sellae and anterior to skull base.[12] On T1WI, clival chordoma appears as a hypointense to intermediate intensity mass, being easily recognized as normal clivus has high SI due to fatty marrow. On T2WI, it has a high SI as a result of its high fluid content or vacuolated cellular

elements. There may be areas of intratumoural heterogeneity that likely represents calcification, haemorrhage or mucin contents. Majority of chordomas demonstrate moderate to marked enhancement.[11] Morphologically, the tumour has a lobulated appearance. Low SI septations may be seen within the lobulations. Gadolinium enhanced images better depict the tumour margin, adjoining soft tissue invasion and presence of enlarged lymph nodes if any. All our cases of clival chordomas presented as multilobulated masses centred at the clivus destroying it without invasion into brain or pituitary. They were seen encroaching the prepontine cistern compressing the pons and basilar artery. In one case the large lesion displaced internal carotid arteries laterally, compressed the optic chiasma and destroyed the floor of anterior cranial fossa.

Spinal chordomas usually develop within the vertebral body. Soft tissue mass with bone destruction and spanning across multiple vertebral levels is common. Soft tissue masses in the sacrococcygeal chordomas typically grow anteriorly into the pelvic cavity, with posterolateral extension as well. On MRI, they have intermediate to low SI in T1WI and heterogeneously high signal in T2WI Multilobulated mass with high signal on T2 with hypointense septa is characteristic in chordomas and has been reported in 70 % of tumours.[13]

In our study, all the tumours were low to intermediate signal intensity on T1WI. On T2WI 8 chordomas were hyperintense, and 2 showed intermediate to high SI. Following contrast administration all the tumours demonstrated heterogenous enhancement, which was moderate in 8 cases, while 2 showed mild enhancement. Morphologically, all the tumours in our cases were multilobulated and demonstrated bone destruction which was corroborated with previous CT scans done in two cases. All our cases of sacrococcygeal chordomas destroyed more than one vertebral segments and had large pre-vertebral soft tissue component expanding anteriorly into the pelvic cavity. Spinal canal and neural foraminal encroachment were also demonstrated in 3 cases. Characteristic T2 hyper intense cystic spaces with hypointense septa were seen in all our cases.

Based upon the typical appearance of the lesions as compared with available literature, a primary diagnosis of chordomas was suggested in all our cases.

A few lesions resemble chordomas and may present as a diagnostic challenge in some cases. Most often, chondrosarcomas are confused with clival chordomas. Both the lesions have similar SI patterns MRI.[14] Chondrosarcomas usually arise off midline along petro-occipital fissure, while chordomas arise from the midline. Chondrosarcomas classically have arc like, linear or globular calcifications, best seen on CT.

Clival meningiomas do not usually have a destructive appearance, instead they can cause bone sclerosis. Meningiomas may have a dural attachment and show homogenous enhancement. Nasopharyngeal carcinomas grow anteriorly into nasopharynx and show regional lymphadenopathy. Plasmacytomas and lymphomas of the skull base cause lytic bone destruction and may mimic clival chordomas if they are located in midline. Paediatric rhabdomyosarcomas can have a similar appearance although they originate anteriorly from the nasopharynx appearing as a bulky tumour with lytic bone destruction.[15] Other rare differentials of clival chordomas include aggressive pituitary adenoma, fibrous dysplasia, histiocytosis X.[16] The differentials of sacral chordomas include chondrosarcoma, giant cell tumour, plasmacytoma and metastasis. Chondrosacromas, although may have a similar imaging appearance on MRI, have an off-midline location and have characteristic calcification patterns similar to chondrosarcomas elsewhere. Giant cell tumours are eccentric in location with extension across the sacro-iliac joint.[17] They may also show fluid-fluid levels.

Although, MR imaging is invaluable in determining tumour location, morphology, extension and its relationship to adjacent structure, it has limited ability in the evaluation of bone and calcification which appear hypointense on all sequences due to scarcity of mobile protons.[18] Subtle bone involvement or erosion maybe difficult to demonstrate on MRI alone, although contrast enhanced MRI improves visualisation of bone involvement. CT scan is highly sensitive for depiction of bony abnormalities and also for picking up intratumoural calcifications. However, CT is less accurate for delineation of tumour extent and anatomical details. Hence, these two imaging modalities may be complementary in imaging evaluation of chordomas. Motion related image quality degradation due to patient's restlessness and claustrophobia are also some of the disadvantages of using MRI, albeit in a small number of patients. Fortunately, no such problems were encountered in our study.

With the advent of newer high field strength magnets and improved software and hardware configurations, new applications of MR imaging have emerged. MR angiography can used to demonstrate relationship of tumour to major vessels, vessel narrowing, encasement or occlusion especially in intracranial chordomas, without the use of intravenous contrast media.

Diffusion Weighted Imaging (DWI) is based on Brownian motion of water and its restriction. Studies have demonstrated, DWI can help to differentiate benign and malignant musculoskeletal lesions.(19) Thus DWI may be used to differentiate between a chondrosarcoma and a chordoma. Dynamic Contrast Enhanced (DCE) imaging is another emerging application of MRI to differentiate chondromas from other skull base tumours. Lang et al used DCE to distinguish between giant cell tumour and chordoma.[20]

Treatment of chordomas consists mainly of surgical resection, the more complete the removal the better the prognosis. With improved imaging qualities, the surgeon can now have more detailed information to choose the best surgical approach. Radiotherapy is offered with aggressive surgical resection to treat any residual lesion. Here also imaging plays an essential role in planning for radiotherapy. It helps tumour localization, delineation, volume measurement and provides information about adjacent vital structures such as nerves and vessels.

Nowadays, proton beam therapy is used with precision in clival chordomas avoiding damage to surrounding structures. Chordomas are notorious for recurrence which eventually is the cause of death in most cases.[9] Recurrences are best evaluated with MRI.

CONCLUSIONS

Chordomas are rare, locally aggressive tumours of notochordal origin. MR is the imaging modality of choice for these tumours as it provides excellent anatomical information with superb soft tissue contrast and ability to image in any desired plane. Chordomas are commonly located in clivus and sacrococcygeal region. Characteristic MRI features include a multilobulated appearance, hypointensity to intermediate intensity on T1WI, hyperintensity on T2WI with hypointense septations. MRI is invaluable not only to narrow down a preoperative diagnosis, but helps to determine tumour extent, relationship with adjacent structures and provides important roadmap for treatment planning including surgery and radiotherapy.

Data sharing statement provided by the authors is available with the full text of this article at jebmh.com.
Financial or other competing interests: None.
Disclosure forms provided by the authors are available with the full text of this article at jebmh.com.

REFERENCES

[1] Stiller CA, Trama A, Serraino D, et al. Descriptive epidemiology of sarcomas in Europe: report from the RARECARE project. Eur J Cancer 2013;49(3):684-695.
[2] Beaugie JM, Mann CV, Butler CE. Sacrococcygeal chordoma. Br J Surg 1969;56(8):586-588.
[3] Post MJD. Primary spine and cord neoplasms. In: Categorical course on spine and cord imaging. American Society of Neuroradiology 1988;8:58-70.
[4] Krol G, Sundaresan N, Deck M. Computed tomography of axial chordomas. J Comput Assist Tomogr 1983;7(2):286-289.
[5] Firooznia H, Pinto RS, Lin JP, et al. Chordoma: radiologic evaluation of 20 cases. AJR Am J Roentgenol 1976;127(5):797-805.
[6] Sze G, Uichanco LS 3rd, Brant-Zawadzki MN, et al. Chordomas: MR imaging. Radiology 1988;166(1 Pt 1):187-191.
[7] Kaye A, Laws E. Brain tumours. 2nd edn. Edinburgh: Churchill Livingstone 2001: p. 857-868.
[8] Wetzel LH, Levine E. Pictorial essay. MR imaging of sacral and presacral lesions. AJR Am J Roentgenol 1990;154(4):771-775.
[9] Atlas S. MRI of the brain and spine. 2nd edn. Philadelphia, Pa: Lippincott Williams & Wilkins 1998.
[10] Haaga J. CT and MRI of the whole body. 4th edn. Philadelphia, PA: Mosby/ Elsevier 2003.
[11] Erdem E, Angtuaco EC, Van Hemert R, et al. Comprehensive review of intracranial chordoma. Radiographics 2003;23(4):995-1009.
[12] Laine FJ, Nadel L, Braun IF. CT and MR imaging of the central skull base. Part 1: techniques, embryologic development and anatomy. Radiographics 1990;10(4):591-602.
[13] Cui JF, Hao DP, Chen HS, et al. Computed tomography and magnetic resonance imaging features of cervical chordoma. Oncol Lett 2018;16(1):861-865.
[14] Weber AL, Brown EW, Hug EB, et al. Cartilaginous tumours and chordomas of the cranial base. Otolaryngol Clin North Am 1995;28(3):453-471.
[15] Weber AL, Liebsch NJ, Sanchez R. Chordoma of the skull base. Radiologic and clinical evaluation. Neuroimaging Clin North Am 1994;4(3):515-527.
[16] Doucet V, Peretti-Viton P, Figarella-Branger D, et al. MRI of intracranial chordomas: extent of tumour and contrast enhancement - criteria for differential diagnosis. Neuroradiology 1997;39(8):571-576.
[17] Llauger J, Palmer J, Amores S, et al. Primary tumours of the sacrum: diagnostic imaging. AJR Am J Roentgenol 2000;174(2):417-424.
[18] Larson TC 3rd, Houser OW, Laws ER Jr. Imaging of cranial chordomas. Mayo Clin Proc 1987;62(10):886-893.
[19] Razek AA, Mossad A, Ghonim M. Role of diffusion weighted MR imaging in assessing malignant versus benign skull-base lesions. Radiol Med 2011;116(1):125-132.
[20] Lang N, Su MY, Xing X, et al. Morphological and dynamic contrast enhanced MR imaging features for the differentiation of chordoma and giant cell tumours in the axial skeleton. J Magn Reson Imaging 2017;45(4):1068-1075.