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**CASE STUDY**

# ROLE OF IMAGING IN CHORDOMA WITH HISTOPATHOLOGICAL CORRELATION

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**ABSTRACT**

**Aims and Objectives:** Chordoma are midline tumours originating from embryonic remnants of the primitive notochord. They are low-grade neoplasms, locally aggressive, slow-growing, but highly recurrent. Chordomas have 4 pathognomonic characteristics on plain film evaluation: expansion of the bone, rarefaction, trabeculation, and calcification. The usual radiographic pattern is lytic, with frequent calcification or sequestered bone fragments. CT scanning is essential, highly sensitive, and accurate for evaluating bony integrity, bone destruction, and calcifications or bone fragments within the lesion. Evaluation of the precise extent of the tumour and the degree of involvement of adjacent tissues is best performed by MRI. The aim of our study is to highlight the role of imaging in chordomas and to correlate imaging diagnosis with histopathological findings.

**Materials and Methods:** The study was conducted over a period of one year on patients who came to MIMS general hospital with clinical and radiological suspicion of chordoma. This is a prospective study done on a total of 15 patients who were radiologically suspected as having chordoma and evaluated on Seimens 16 slice CT and Siemens essenza 1.5 tesla MRI with contrast.

**Results:** In our study of 15 cases, most common age group was found to be 20-40 years and chordomas had slightly greater female predilection. Out of 15 cases, 9 were histopathologically proved as chordomas and the remaining 6 turned out to be false positive. Based on these results the sensitivity and specificity of CT and MRI in diagnosing chordoma is 100% and 60% respectively. Though there remains a question of controversy, in our study most common location is spheno-occipital region.

**Conclusion:** In our study, we found out that both CT and MRI have good accuracy and are complementary to each other in diagnosing chordomas, however each one has certain superior characteristics over the other. Though radiological imaging plays an important role in diagnosing chordoma, histopathological examination is essential for confirmation. A rare presentation of chordoma with orbital extension was also included in our study, which was histopathologically proven. So, the possibility of chordoma should be suspected even if we found the lesion in an uncommon location, when typical radiological features of chordoma are encountered.



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## INTRODUCTION

Chordoma are midline low-grade neoplasms, locally aggressive, nevertheless highly recurrentbut uncommonly metastasise. They originate from embryonic remnants of the primitive notochord (earliest fetal axial skeleton, extending from the Rathke's pouch to the coccyx). Since chordomas arise in bone, they are usually extradural and result in local bone destruction. They are slow growing tumours and often present clinically when there is mass effect on adjacent structures. Chordomas are found along the axial skeleton and a relatively evenly distributed among three locations – sacro-coccygeal (30-50%), spheno-occipital (30-35%), vertebral body (15-30%) (c). On imaging chordoma features on CT include well-circumscribed, centrally located expansilede structive lytic lesion, sometimes with marginal sclerosis, an extraosseous soft-tissue component (usually hyper-attenuating relative to the adjacent brain; however, inhomogenous areas may be seen due to cystic, necrosis or haemorrhage; the soft-tissue mass is often disproportionately large relative to the bony destruction) with irregular intratumoral calcifications (thought to represent sequestra of normal bone rather than dystrophic calcifications) and withmoderate to marked enhancement. MRI features include intermediate to low signal intensity on T1 weighted images and small foci of hyperintensity (intratumoral haemorrhage or a mucus pool) may be seen. Most chordomas exhibit very high signal on T2 sequences. On contrast administration, heterogeneous enhancement with a honeycomb appearance corresponding to low T1 signal areas within the tumour are noticed.

### Aims and Objectives

1. To confirm the diagnosis of chordoma
2. To compare and correlate CT and MRI findings.
3. Correlation of radiological (CT & MRI findings) with histopathological findings.
4. To assess sensitivity and specificity of imaging in diagnosing chordoma.

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## MATERIALS AND METHODS

The patients included in our study were those who were referred to Department of Radio- diagnosis of MIMS general hospital with clinical and suspicion of chordoma. This prospective study was conducted over a period of one year. A total of 15 patients were included who were radiologically suspected as having chordomas,out of which 9 were females,6 were males. All patients were subjected to plain and contrast studies of CT and MRI. CT was done on Seimens 16 slice and MRI on Siemens essenza 1.5 tesla machines. After surgical excision, biopsy for histopathological examination has been sent for correlation.

### Inclusion criteria

All the patients with typical clinical and radiological features of chordoma.

### Exclusion criteria

Patient who are contraindicated to MRI were excluded from study. In patients in whom histopathological diagnosis could not be obtained.

## RESULTS

In our study of 15 cases, majority frequency of chordomas are noticed in the third to fifth decade (60%), followed by sixth and seventh decades(33.3%) as depicted in Table 1 and figure 1. In my study female predilection was more compared to male in all types of chordomas. Out of 15 cases, 9 were females comprising 60% of study population and 6 cases were males (40%) as shown in Figure 2. The most frequent site of chordoma in our study is found to be spheno-occipital (clival) region which comprises of 53.5% of the study group, the next common site being sacro- coccygeal (40%). The less common or rare presentation of chordoma with orbital invasion was noted in a single case in the present study as depicted in Figure 3.

**Table 1. Age distribution of patients in study group**

| AGE GROUP | NUMBER OF CASES |
|---|---|
| 20-40 YEARS | 9 CASES |
| 41-60 YEARS | 5 CASES |
| 61-80 YEARS | 1 CASE |

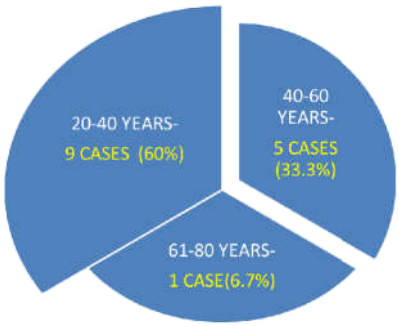


**Figure 1. Depicting the age distribution in percentage**

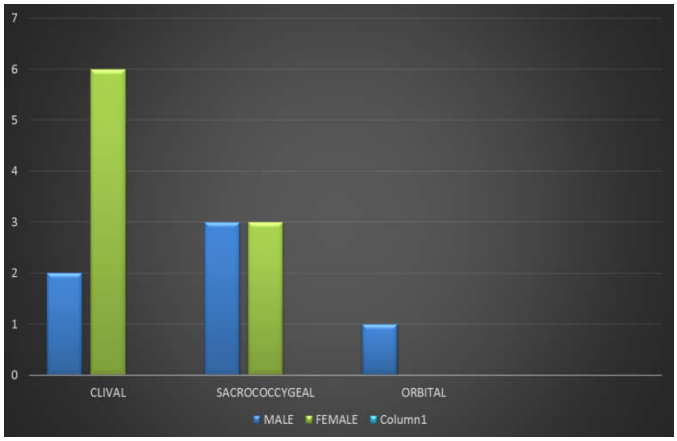


**Figure 2. Bar diagram depicting the sex distribution of various chordomas**

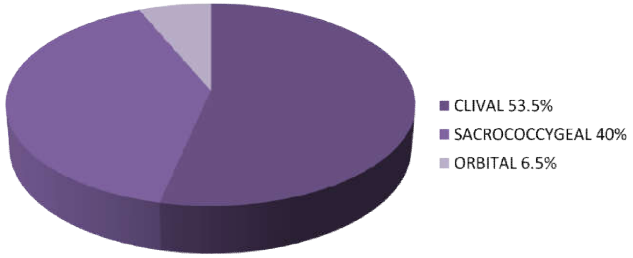


**Figure 3. Pie chart depicting location wise distribution of chordomas in the study**

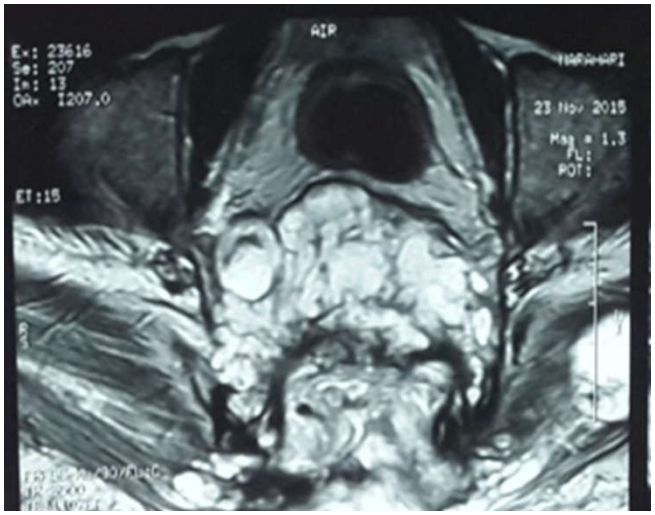

**Figure 4. Sacro coccygeal chordoma**

## DISCUSSION

Chordomas are rare malignant neoplasms that arise from remnants of the embryological notochord. The incidence of chordomas is approximately 1 per 2 million. The major site for chordomas is the sacrococcygeal region, which accounts for 50% of all chordomas. Other sites include the skull base (35%) and cervical, thoracic, or lumbar vertebral bodies (15%) (Baratti *et al*., 2003). Chordomas occur at any age but are usually seen in adults (30-70 years). Those located in the spheno-occipital region most commonly occur in patients 20-40 years of age, whereas sacrococcygealchordomas are

typically seen in a slightly older age group (peak around 50 years) (Farsad *et al*., 2009). Chordomas have 4 pathognomonic characteristics on plain film evaluation: expansion of the bone, rarefaction, trabeculation, and calcification.

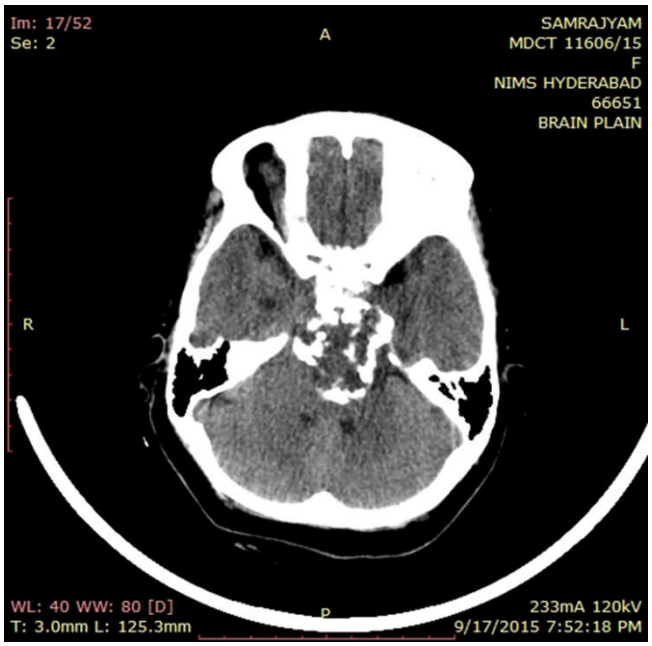


**Figure 5. Clivalchordoma**

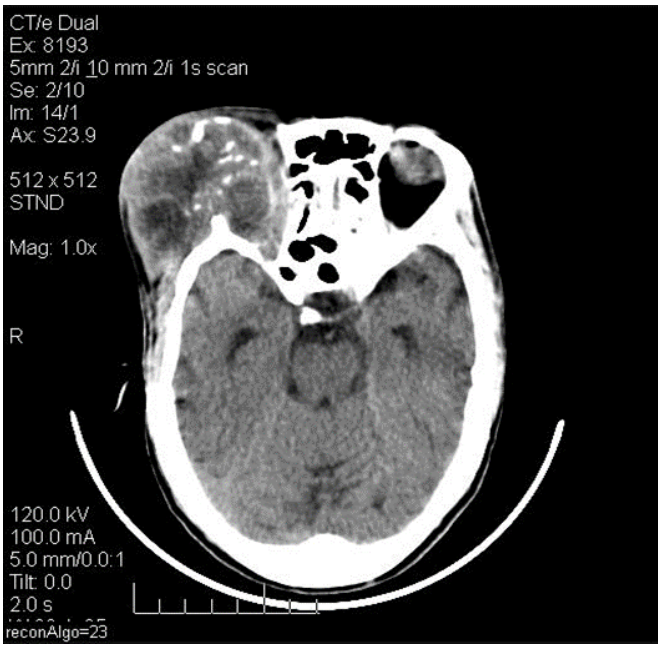


**Figure 6. Orbital chordoma**

The usual radiographic pattern is lytic, with frequent calcification or sequestered bone fragments. Plain radiograph of the pelvis showing an expansion of the sacrum, bone rarefaction, and large mass of soft tissue with some trabeculations indicates a sacro-coccygeal chordoma. However, radiographs are neither specific nor sensitive for detecting chordoma; for intracranial chordomas, plain films are no longer used. In addition, although plain films are often the first examination for sacrococcygeal and spinal chordomas, CT scanning and MRI are necessary for the diagnosis. Finally, even if a destructive clival lesion is observed on plain films, the size of the tumor may be grossly underestimated not only because portions of a chordoma may have little or no calcification present but also because the soft-tissue component is not visualized. CT scanning is essential, highly sensitive, and accurate for evaluating bony integrity, bone destruction, and calcifications or bone fragments within the lesion. On CT scans, the chordomas appear homogeneous, with a density comparable to that of muscles. The tumor appearance on contrast enhancement is heterogeneous. Calcification is found in less than one half of patients, and differentiation from sequestered bone fragments is difficult. CT findings of different types of chordoma include – Intracranial chordomas - The most characteristic appearance of an intracranial chordoma is centrally located expansile destructive mass lesion with large soft tissue component, arising from the clivus which primarly extends posteriorly, if massive, extension into sellaanterosuperiorly and nasopharynx and middle cranial fossa antero inferiorly is common. (Meyer *et al*., 1986) Calcification is common, and areas of low attenuation within the soft-tissue mass — representing the myxoid and gelatinous material found on pathologic examination — are occasionally found on CT scans. CT scanning reliably demonstrates petrous apex involvement and lysis of the skull base foramina. (Moore *et al*., 2015)

Sacrococcygealchordomas - Chordomas are often massive, well-delineated tumors that shift the fatty tissue of the pelvis and involve bone structures and the epidural area. Peripheral sclerosis may be observed in approximately 50% of patients, and frequently, a discrepancy is found between a large soft-tissue component and the area of bone involvement. In addition, regional lymph nodes are usually invaded. The most reliable sign of sacral chordomas is the destruction of several sacral vertebrae associated with a tissue mass anterior to the sacrum. However, the association of osteolytic lesions and soft masses involving the discs and the vertebrae suggests other diagnosis, such as neurofibromas, lymphomas, metastases, and plasmacytomas. Spinal chordomas - Infrequently, chordomas arise in the mobile (i.e, cervical, thoracic, lumbar) spine (15%). (Meyer *et al*., 1986) The cervical spine is the most common site for these tumors, with a predominance in the C2 vertebra; the thoracic (Taki *et al*., 1996) and lumbar areas of the spine are involved less frequently. Initially, the presentation of chordoma on CT scan is of bone destruction centered in the vertebral body, with an associated anteriorly or laterally situated, paraspinal soft-tissue mass that may contain calcification. Epidural extension of the tumor is usual. Among imaging methods that contribute to the diagnosis, MRI is particularly reliable; this modality is highly accurate in assessing the soft-tissue extent of chordomas and in evaluating involvement of adjacent tissues (Wetzel and Levine, 1990). The best tool for demonstrating tumoral site and extension and for selecting the surgical approach is 3-dimensional (3-D) MRI. (Stephens and Schwartz, 1993) Indeed, for clivalchordomas, 3-D gradient-echo T1-weighted sequences are helpful, because they visualize the tumor in 3 planes within a short time and with a good analysis of tumoral signal. (Mehnert *et al*., 2004). Coming to individual MRI characteristics:

Intracranial chordomas - MRI specifically shows tumour extension which is primarily along the anteroposterior axis rather than laterally. The expansion of the bone in the early stage indicates that the tumour arises from bone and not from adjacent structures. Skull base chordomas are well delineated at the outset, as they displace adjacent structures. Most chordomas are iso-intense or demonstrate low signal on T1-weighted images. Most chordomas exhibit high signal on T2-

weighted images, which is also nonspecific. Following contrastchordomas usually show lobulated areas with a honeycomb appearance corresponding to low signal areas within the tumor. (Leproux *et al*., 1993) Chordoma signal is described as heterogeneous after gadolinium injection and on T1- and T2-weighted images. Sacrococcygealchordomas - On MRI, these are lobulated tumours, typically with low to intermediate signal intensity on T1-weighted images and heterogeneous high signal intensity on T2-weighted images. The pattern of gadolinium enhancement is the same as for clivalchordomas.

**Differential diagnosis of Chordomas include**

**Chondrosarcomas -** They originate from embryonic remnants of the primitive cartilage and tend to arise off-midline from petro-occipital fissure and extend more laterally. Hemorrhage is uncommon

**Craniopharyngiomas–** Thesetumours are suprasellar, sellar, or infrasellar and are rarely at the level of the nasopharynx. Generally, the site is more anterior and superior, and extension is almost always posterosuperior (interpeduncular cistern).
Aggressive Pituitary macroadenomas - usually invade the sphenoid sinus.

**Clivalmeningiomas-** They have a large dural attachment and do not appear similar to bone tumours. Homogeneity of their signal is an additional element.

**Sacral schwanoma-** these are anteriorly located well circumscrined lesion with intratumoral cysts causing adjacent bone erosions, vertebral body scalloping .It often extends paraspinally causing foraminal widening and giving adumbell shaped appearance.

In our study frequency, chordomas was found to be more in females compared to males, which is similar to findings in the studies done by Young *et al*. (2008) and Hoch *et al*. (2006) who conducted their study on skull base chordomas on 30 and 73 patients respectively. According to the study conducted by Smolders *et al*. (2003), the mean age of presentation was found to be 45 years, which was very much coinciding with ours being 41 years. In the study conducted by Elefante *et al*. (2013) both spinal and clivalchordomas had similar incidence, whereas in our study clivalchordomas was found to be more common followed by sacrococcygeal and spinal chordomas. In our study, out of 15 radiologically suspected cases of chordomas, 9 were proven histologically. Hence, the sensitivity and specificity of radiological imaging in diagnosis of chordomas was considered to be 100% and 60% respectively.

**Conclusion**

In our study, we found out that both CT and MRI have good accuracy and are complementary to each other in diagnosing chordomas, CT-being faster, greater availability and lower cost makes it a method of choice in most of the cases. Moreover, it is better in depicting bone destruction and patterns of intratumoral calcifications. MRI is capable of good depiction of the tumour margins, nerve roots, neural foramina and vascular involvement. The overall sensitivity and specificity of radiological diagnosis of chordoma is 100% & 60% respectively, which indicates that, though radiological imaging plays an important role in diagnosing chordoma but histopathological examination is essential for confirmation.

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