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Case Report

# Primary spinal chordoma: A rarity in itself, with a rarity of its own

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ABSTRACT

Chordoma accounts for 1-4% of all primary malignant tumours of bone. Besides advances in rgical techniques and radiation therapy, tumour may recur locally or metastasize to distant sites with local recurrence being predominant form. Here we present a case report of primary spinal chordoma which recurred locally within a year of surgical excision. Chordoma of spine is a rare low grade malignant tumour with strong propensity for local recurrence. Long term follow up is usually recommended as there is high chance of recurrence and malignant transformation even after surgery followed by radiation therapy.



## 1. Introduction

Chordomas are locally aggressive malignant tumours of notochordal origin. Besides advances in surgical techniques and radiation therapy, tumour may recur locally at the site of primary or metastasize to distant sites with local recurrence being predominant form.[1] Here we present a case report of recurrent spinal chordoma which recurred within a year of surgical excision.

## 2. Case Report

A 40 year old male came on November 2016 with complaints of weakness of left lower limb for past 8 months and difficulty in passing stools and urine for past 7 months.

CT spine showed D11 – D12 intradural intramedullary tumour? Ependymoma /? Glioma.

MRI dorsolumbar spine showed intradural and intramedullary contrast enhancing lesion from D10-D11.

Patient was operated on November 2016- D11- D12 laminectomy and subtotal excision of SOL was done. Biopsy proved to be chordoma which is confirmed by immunohistochemistry and then the patient remained symptom free for next 5 month.

Again he presented on May 2017 with complaints of persistent weakness of left lower limb with tightness of left lower limb while squatting.

MRI dorsal spine showed T1 hypo intense and T2 heterointense lesion of 3.1x2.2 cm noted in spinal canal at D 11- D12 level showing heterogenous contrast enhancement-known case of chordoma involving D11-D12 segments-post operative status- features suggestive of residual lesion with syringomyelia.

Re-exploration and excision of SOL was done in May 2017.

### *2.1. November 2016.*

Gross: received multiple grey brown soft tissue fragments measuring 0.5 cm.

MICROSCOPY [Figures 1 and 2]: Section shows fragments of neoplasm composed of sheets of polyhedral cells with vacuolated cytoplasm and eccentrically placed dark staining nuclei (physaliferous cells). Stroma showed

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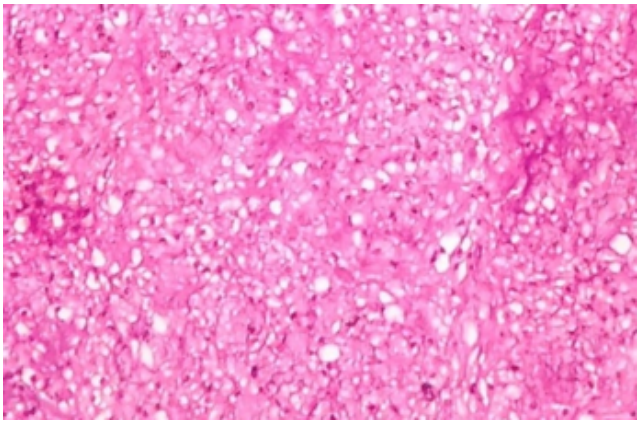

**Fig. 1:** H & E 100 X

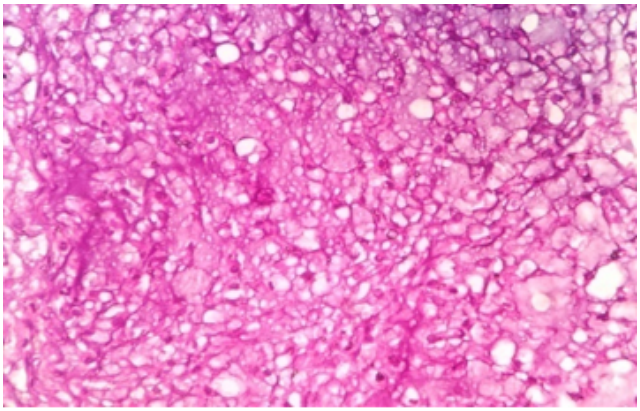

**Fig. 2:** H & E 400X

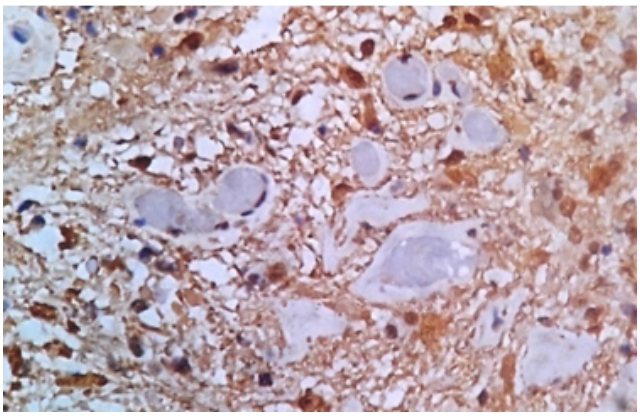

**Fig. 3:** S 100, 100X

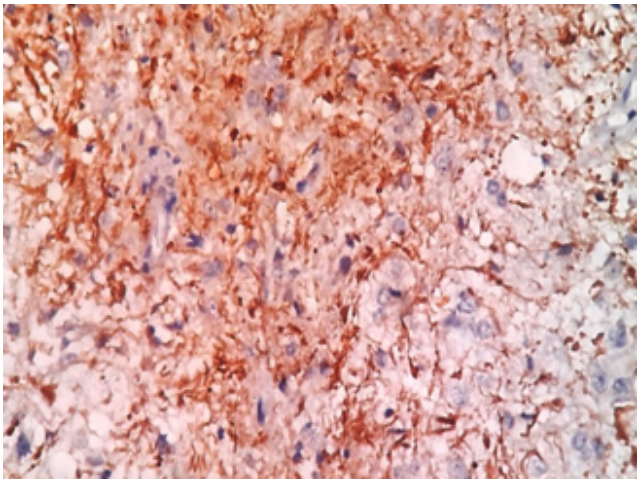

**Fig. 4:** CK 100 X

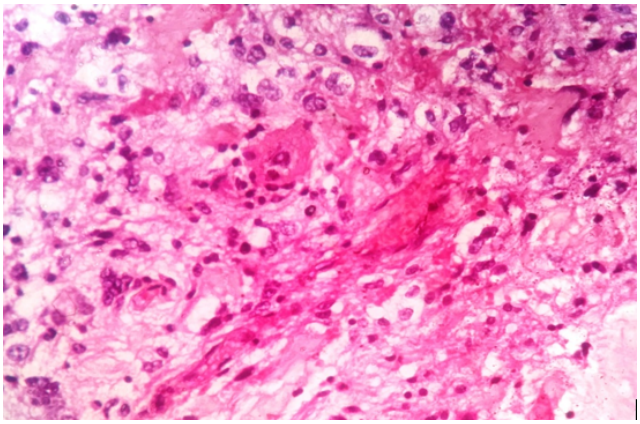

**Fig. 5:** H & E 100X.

extravasated myxoid and mucin material. Stroma showed congested vessels and foci of hemorrhagic infarction.

IMPRESSION was given as features suggestive of CHORDOMA. IHC markers were under study.

S-100 and CK [Figures 3 and 4] showed strong positivity confirming the diagnosis.

### *2.2. May 2017*

GROSS: Received multiple grey white soft tissue fragments measuring 0.2 cc in aggregate.

Microscopy: [Figure 5 ] Sections showed a neoplasm arranged in small clusters and sheets. The tumor cells were large oval to polyhedral with vacuolated cytoplasm and eccentrically placed vesicular nuclei. Surrounding stroma showed extravasated mucinous material with few congested blood vessels and scattered chronic inflammatory infiltrate.

With this the impression was given as recurrent chordoma.

**Table 1:** Differential diagnosis for chordoma.

| Lesions | Age group | Site | Gross | H&E | IHC |
|---|---|---|---|---|---|
| **Chordoma** [2,3] | 40-70 | Sacrococcyx, clivus, spine | Lobulated firm mass with mucoid cut surface | Well defined lobules of epithelial cells separated by bands of fibrous tiisue with mucoid background. Physaliferous cells. nuclear pleomorphism is rare. | Vimentin:+ve. Cytokeratin:+ve. EMA-+ve. S 100 protein-variably+ve. GFAP- negative. Brachyury- positive. D2-40- positive. |
| **Chondrosarcoma** | 20-30 | Lesion of dural origin or directly arises within brain parenchyma | Well circumscribe, firm, lobulated mass | Biphasic appearance with hyaline cartilage and sheets of round to oval small cells. staghorn vascular spaces are seen. | Vimentin-strong positivity. S-100 -positivity Cytokeratin-negative. GFAP- negative EMA-negative D2-40 – NEGATIVE. Brachyury-negative |
| **Myxopapillary ependymoma** | | Sacrococcygeal region,Filum terminale | Tumor cells were grouped around blood vessels. | Pseudopapillary architecture with elongated monomorphic cells. | GFAP- positive. Keratin-negative. S 100 -positive |
| **Chondroid Chordoma** | Younger age | Spheno-occipital region | | Chordoid areas admixed with low grade chondrosarcoma like areas. | CK-positive EMA-positive |
| **Chordoid Glioma** [4] | 30-60 years | Anterior wall or roof of third ventricle | Well circumscribed tumor with central cyst or necrosis | Cords and clusters of epithelioid cells within a mucinous background with lymphoplasmacytic infiltrates. | GFAP – diffuse positivity. EMA – focal positive.vimentin – positive D2-40-positive S-100- negative .pan ck and synaptophysin-negative. |
| **Chordoid meningioma** [5] | No age predilection | Supratentorial region | | Chordoid areas mixed with typical meningothelial area | S-100- negative.EMA – PositiveD2-40- positive |

## 3. Discussion

Chordoma of spine is a rare slow growing primary bone tumor that arises from the remnants of the notochord, can occur anywhere along spine from skull base to coccyx.[6] Chordoma accounts for 1-4% of all primary malignant tumours of bone and around 20% of the primary tumors of spinal column. Incidence in spine is 0.08 per 1,00,000 people.

Most common sites involved are sacrococcygeal region (50%), base of skull involving sphenooccipital region (35%) and vertebral column (15%).

Most common age group is 40-70 years with a male – female ratio of 2:1.[7–9] Usually presents with pain as the cardinal symptom. Neurological deficits tend to vary based on the location of the lesion.[9]

### *3.1. Radiological features*

Chordomas are usually extradural and induce bone destruction. CT scan of spine shows bone destruction centered in vertebral body with paraspinal soft tissue mass that may contain calcification. MRI of spine shows hypo or isointense on T1 and hyperintense signals on T2 weighted images with heterogenous contrast enhancement.[10]

A sclerotic and osteolytic lesion in a vertebral body with a large paraspinal soft tissue mass in an older patient with long standing back pain should raise the suspicion of chordoma.[11]

### *3.2. Gross & microscopic features*

Chordoma is gelatinous and soft and contains areas of haemorrhage.[2] It shows epithelioid cells in cords and lobules separated by variable amount of mucoid material and fibrous septa. Physaliferous Cells which are extremely large with vacuolated bubbly cytoplasm and prominent vesicular nucleus. Some of the cytoplasmic vacuoles contain glycogen. Mitotic figures are absent. Areas of cartilage and bone may be present.[3]

### *3.3. Immunohistochemical findings*

Physaliferous cells shows positive reactivity for PAS, S-100 protein, keratin and epithelial membrane antigen.

Matrix is alcian blue and mucicarmine positive.[12]

CEA and GFAP- shows negativity.[13]

### *3.4. Treatment*

The most common accepted approach is aggressive surgery to resect tumour as far as possible.

### *3.5. Recurrence*

Recurrence is common after surgery about 28-68% and is due to surgical violation of tumour margins at the time of surgery. Metastasis is seen in 3-60% of cases.[14,15]

## 4. Conclusion

Chordoma usually appears extradurally but in our case it was presented for its rarity and as an intradural mass and also thoracic vertebral chordoma is relatively rare. The incidence in our instituition for chordoma among total CNS tumours is 0.2%. Long term follow up is usually recommended as there is high chance of recurrence and malignant transformation even after surgery followed by radiation therapy.

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## 6. Conflicts of Interest

None to declare.

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