

## **Sacrococcygeal Chordoma**

By

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## **Sacrococcygeal Chordoma**

Malignant Tumors with Notochord differentiation

### **Types**

1) Conventional Chordoma -95%

2) Poorly Differentiated Chordoma

-Loss of INI 1

-Brachyuri positive

3) Dedifferentiated Chordoma-1%

-Biphasic. - Conventional Chordoma + High Grade Sarcoma like High Grade Undifferentiated Pleomorphic Sarcoma or Osteosarcoma

### **Sites**

1 Sacrococcygeal

2 Clivus

3 Spine. Vertebrae

1 to 5 %

Age -40 to 60 years

More common in males

### **IHC**

Positive for CK ,EMA ,S100

Brachyuri

Loss of INI 1

Children and young adults - Cranial Chordoma

Poorly Differentiated Chordoma - Clivus. Cervical

T Gene

Brachyuri duplication

T box Transcription factor

Rare Tuberous Sclerosis

Biallele I activation of TSC1 and TSC2

DD

1 Chondrosarcoma

2 Metastatic Carcinoma

3 Myxopapillary ependymoma

4 RCC

5 Chondroid Meningioma

6 Benign Notochordal tumor

7 Myoepithelioma

**Bad prognosis**

Large size

Positive Surgical margins

Tumor Necrosis

High Ki 67. MIB 1

Dedifferentiated Chordoma

