

# Chordoma of the Clivus

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## Citation

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## Abstract

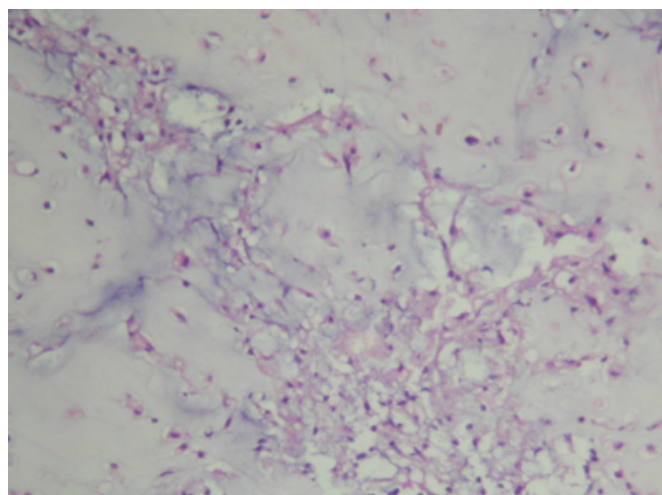
Sir,

Chordomas of the clivus are aggressive lesions which arise from remnants of primitive notochord and pose unique diagnostic and management challenges<sup>1</sup>. They account for 1% of all intracranial tumours and present with cranial nerve palsies, orbito-frontal headache, visual disturbances and intracranial hemorrhages<sup>2,3</sup>. They are therefore usually discovered only after having undergone long periods of undisturbed growth. Clinical diagnosis is made by interpretation of patient's presenting symptoms and objective findings on physical examination. Radiographic studies supplement this information. True diagnosis can only be made histologically.

A 50-year-old male presented with the chief complaints of headache, vomiting, and decreased vision for 8-9 months. Headache was continuous, severe, and present all over the head and associated with vomiting. History of increased sweating, and intolerance to heat was also present. Pituitary hormone levels (human growth hormone, TSH, cortisol) were within normal limits. CT scan revealed a hyperdense mass within the sphenoid sinus extending into the nasopharynx, abutting the posterior choana. MRI showed a sellar tumour invading the clivus. The patient underwent a right frontal craniotomy via sub-frontal approach and tumour decompression was done. The specimen was sent for histopathological examination. Grossly, multiple grayish-white to grayish-brown soft tissue pieces were received that all together measured 0.6x0.4x0.2cm. Microscopy showed a tumour arranged in nests, cords and sheets. Tumour cells were present in a chondromyxoid background and had round nuclei with abundant eosinophilic cytoplasm (Fig. 1). Physaliferous cells were also seen. Some tissue bits showed compressed pituitary glands and areas of hemorrhage (Fig.2). Tumour cells stained positively with PAS (Fig.3) and S-100. The final diagnosis was chordoma.

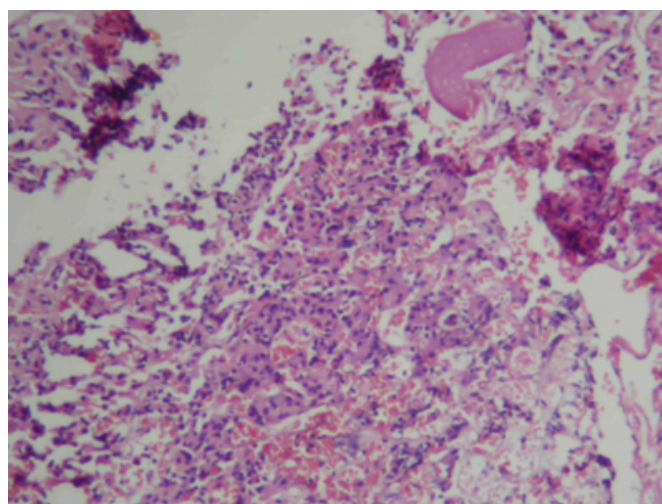
## Figure 1

Fig. 1 - Photomicrograph showing tumour cells in a chondromyxoid background having round nuclei with abundant eosinophilic cytoplasm. (H&E, x100)



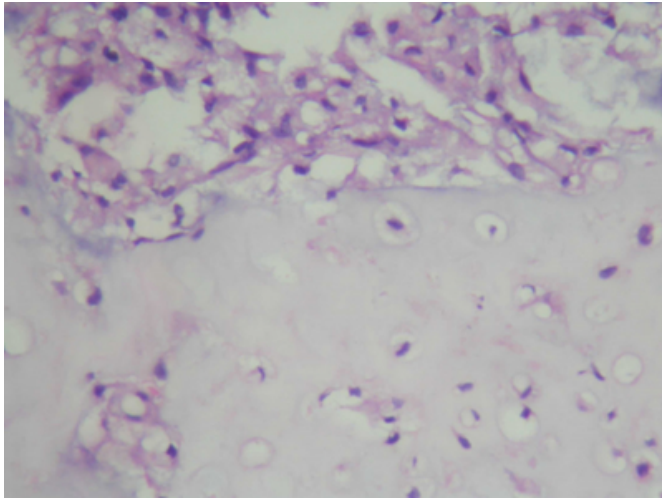
## Figure 2

Fig. 2 - Photomicrograph showing compressed pituitary glands and areas of haemorrhage in the vicinity of the tumour. (H&E, x400)



### Figure 3

Fig. 3 - Photomicrograph showing cytoplasmic positivity of tumour cells with PAS stain. (PAS, x400)



Chordomas are rare tumours that are unevenly distributed along the craniospinal axis; approximately half arise in the sacrum, one-third in the spheno-occipital region or clivus, and the remainder in the articulating vertebrae. In the spheno-occipital region or clivus, these tumours involve the posterior nasal cavity, sphenoid bone, nasopharynx and the base of skull. Most are histologically benign without evidence of nuclear mitotic figures. They produce marked destruction of the sphenoid bone and extend into the sphenoid sinus and the nasopharynx.

Cranial nerve palsies are a common presentation due to extension of the tumour into the neural foramina. Other common presenting symptoms are diplopia and headache<sup>4</sup>. CT and MRI are both used to visualize the bony and soft

tissue anatomical relationship of the critical structures in the region of the skull base.

Some chordomas growing in this area contain cartilaginous tissue having the features of chondroma or well differentiated chondrosarcomas. These tumours have been diagnosed as “chondroid chordomas”. If physaliferous cells are found in the lesion, a diagnosis of chordoma should be made even though cartilaginous or sarcomatous features are identified. By immunohistochemical stains, cells in the chondroid component of chordoma express cytokeratin, epithelial membrane antigen and S-100 protein. This is in contrast to negative immunoreactivity for cytokeratin and epithelial membrane antigen and positive expression for S-100 protein in low-grade chondrosarcomas<sup>1</sup>. These immunostains appear useful for diagnosis.

Clival chordomas grow slowly, infiltrate locally, and do not metastasize. Proton beam therapy followed by surgical debulking promises long-term survival despite multiple recurrences<sup>5</sup>.

### References

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