A Comprehensive Review of Intracranial Chordomas

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Introduction

Chordoma is an extremely rare primary bone malignancy of clival origin. It is a soft tissue mass within the bone, often having a considerable mass effect on the bony calvarium, cranial nerves, and other surrounding structures. It is a slow-growing tumor, with an average doubling time of 10 years. Chordoma accounts for 0.01% of all bone cancers and 2% of primary bone malignancies. It is a rare disease, with approximately 200 new cases diagnosed each year in the United States. The overall diagnosis rate has not changed significantly over the last decade.

Imaging

Chordomas are characterized by a variable signal intensity on T1- and T2-weighted MRI images. The signal intensity on CT imaging may vary from hypodense to isodense to hyperdense, but typically, it is hypodense. Bone erosion and destruction are common findings. MRI is the imaging modality of choice for chordoma, as it provides excellent soft tissue contrast and allows for the assessment of the extent of the disease and its relationship to surrounding structures.

Histopathology

Chordomas are characterized by a mass of chondroid matrix and a glial-like stroma. The tumor cells are arranged in a circumscribed, cartilage-like mass, giving the tumor a characteristic “chicken wire” pattern. The cells are often positive for vimentin and S100 protein, and negative for desmin and actin. The tumor cells also stain positively for cytokeratin and mesenchymal markers, such as CD99.

Tumor Spread

Chordomas can spread through the bone, through the soft tissue, and through the cranial nerves. The most common site of recurrence is the skull base, followed by the spine and the orbit. However, spread to other sites, such as the lungs, liver, and lymph nodes, has been reported.

Treatment

The treatment of chordoma is multimodal, and it depends on the location and extent of the disease. The standard treatment consists of surgical resection, followed by radiation therapy. The goal of surgery is to achieve a complete or near-complete resection of the tumor. Postoperative radiation therapy is recommended to reduce the risk of local recurrence. Chemotherapy is not effective against chordoma.

References


Conclusion

Intracranial chordomas are rare, midline tumors of clival origin. MRI and CT imaging are the modalities of choice for diagnosis, treatment planning, and follow-up. Bone destruction and the presence of intraparenchymal calcification are useful on CT imaging. MRI is superior to CT for the assessment of soft tissue invasion and interventional spaces. Surgical resection followed by radiation therapy is the standard treatment, and achieving the best results. Local recurrence is not uncommon with an unfavorable prognosis.