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Commentary

Chordoma: The Pathology, Clinical Presentation, and Advances in Treatment

Liang Qifeng*

Department of Pathology, Shandong University, Shandong, China

DESCRIPTION

Chordoma is a rare and slow-growing type of cancer that originates in the bones of the skull base and spine. Despite its rarity, chordoma presents significant clinical challenges due to its location, potential for recurrence, and resistance to conventional therapies. This study explains about the epidemiology, pathology, clinical presentation, diagnostic methods, and treatment strategies for chordoma, as well as ongoing research and future directions in its management. Chordomas account for about 1%-4% of all primary bone tumors and are diagnosed in approximately one in one million people annually. They occur equally among males and females and can develop at any age, though they are most commonly diagnosed in adults between the ages of 40 and 70. Chordomas are thought to arise from remnants of the notochord, an embryonic structure that contributes to the formation of the spinal column.

Histologically, chordomas are characterized by the presence of large, vacuolated cells known as physaliphorous cells, set in a myxoid stroma. They are generally divided into three subtypes: conventional (or classical), chondroid, and dedifferentiated. Conventional chordomas are the most common, while chondroid chordomas, which exhibit features of both chordoma and chondrosarcoma, have a slightly better prognosis. Dedifferentiated chordomas are rare but highly aggressive, often associated with poor outcomes. Molecularly, chordomas frequently exhibit alterations in the T (brachyury) gene, a transcription factor important for notochord development. Brachyury expression is a attribute of chordoma cells and is used as a diagnostic marker. Other genetic alterations, including mutations in genes involved in cell cycle regulation and growth factor signaling pathways, have also been implicated in chordoma pathogenesis.

The symptoms of chordoma depend on the tumor's location. Skull base chordomas can cause headaches, cranial nerve deficits, and visual disturbances, while spinal chordomas often present with pain, neurological deficits, or dysfunction in nearby organs due to spinal cord compression. Because chordomas grow slowly, symptoms may be present for months or even years before a diagnosis is made. The diagnosis of chordoma typically involves a combination of imaging studies and histopathological examination. Magnetic Resonance Imaging (MRI) is the preferred modality for visualizing chordomas, providing detailed information about the tumor's size, extent, and relationship to surrounding structures. Computed Tomography (CT) scans can complement MRI by highlighting bone involvement. Biopsy and subsequent histological examination confirm the diagnosis, with immunohistochemistry for brachyury aiding in differentiating chordoma from other neoplasms.

Pathology clinical treatments

The treatment of chordoma is challenging due to its anatomical location and tendency to invade critical structures. Multimodal approaches, including surgery, radiation therapy, and, in some cases, systemic therapies, are often employed.

Surgery: Surgery is the primary treatment for chordoma is surgical resection, aiming for complete removal of the tumor with clear margins. However, achieving this is often difficult because of the tumor's proximity to vital structures like the brainstem, spinal cord, and major blood vessels. En bloc resection, where the tumor is removed in one piece, is associated with better outcomes but is not always feasible.

Radiation therapy: Radiation therapy is an important adjunct to surgery, particularly when complete resection is not possible. High-dose radiation, including proton beam therapy and stereotactic radiosurgery, offers precise targeting of the tumor while sparing surrounding healthy tissue. Proton beam therapy has shown promise in treating chordomas due to its ability to deliver high doses of radiation with minimal damage to adjacent structures.

Systemic therapies: Chordomas are generally resistant to conventional chemotherapy, necessitating the investigation of targeted therapies and immunotherapies. Recent advances in understanding the molecular biology of chordoma have led to the investigation of agents targeting specific pathways, such as tyrosine kinase inhibitors, mTOR inhibitors, and PD-1/PD-L1 inhibitors. Clinical trials are ongoing to evaluate the efficacy of these novel treatments. The prognosis for chordoma patients varies based on several factors, including tumor location, size, extent of resection, and histological subtype. Generally, skull

Correspondence to: Liang Qifeng, Department of Pathology, Shandong University, Shandong, China, E-mail: qfenglianf@1548.com

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base chordomas have a better prognosis than those located in the sacrum or mobile spine. Recurrence is common, even after aggressive treatment, underscoring the importance of long-term follow-up.

Regular imaging studies are pivotal for monitoring recurrence or progression. MRI is typically performed every 6-12 months for the first few years post-treatment, with intervals extended based on the stability of the disease. Any new or worsening symptoms should prompt immediate investigation. Genetic and molecular research, further elucidation of the genetic and molecular underpinnings of chordoma could identify new therapeutic targets. Studies on the role of brachyury and other molecular pathways in chordoma pathogenesis are critical for developing targeted therapies. The potential of immunotherapy in treating chordoma is being actively exhibited. Checkpoint inhibitors, cancer vaccines, and adoptive cell therapies represent potential method, particularly for cases resistant to conventional treatments.Participation in clinical trials offers chordoma patients access to innovative therapies and contributes to the collective understanding of the disease. Trials investigating various targeted therapies, combination treatments, and novel radiation techniques are ongoing.

Advanced imaging techniques: Improved imaging modalities and techniques could enhance the accuracy of diagnosis, surgical planning, and monitoring. Functional MRI, PET/CT, and advanced MRI sequences are being studied for their utility in chordoma management. Given the complexity of chordoma treatment, multidisciplinary care involving surgeons, radiation oncologists, medical oncologists, radiologists, and pathologists is vital. Collaboration among specialists ensures comprehensive care and optimizes treatment outcomes.