ORIGINAL ARTICLE

Challenges in Diagnosis and Management of Pediatric Presacral Tumors: A Radiologist's Perspective

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ABSTRACT

Presacral tumors in pediatric patients pose unique diagnostic challenges due to their rarity, diverse histopathology, and complex anatomy of the presacral region. Accurate diagnosis and management of these tumors require a multimodal imaging approach and specialized expertise. This article explores the challenges encountered in imaging pediatric presacral tumors and emphasizes the importance of a comprehensive imaging strategy. The rarity and diverse histopathology of these tumors make accurate diagnosis challenging. The complex anatomy of the presacral space necessitates advanced imaging techniques such as magnetic resonance imaging (MRI) and computed tomography (CT) to visualize the relationships between the tumor and adjacent structures. Differentiating between benign and malignant tumors is crucial for treatment planning, but it can be challenging due to overlapping imaging features. Assessing the treatment response and detecting tumor recurrence require regular follow-up imaging, but post-treatment changes can mimic or mask tumor recurrence. A multidisciplinary approach involving radiologists, pediatric surgeons, pediatric oncologists, and pathologists is essential for accurate diagnosis and treatment planning. Effective communication and coordination among team members are necessary to overcome the challenges associated with managing these complex cases. Future research and advancements in imaging technology will further enhance the ability to diagnose and manage pediatric presacral tumors, ultimately improving patient outcomes.

Keywords: diagnostic imaging, neoplasms, pediatric, presacral

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INTRODUCTION

Presacral tumors are rare neoplasms that develop in the presacral space located between the rectum and sacrum. In pediatric patients, presacral tumors present unique diagnostic challenges due to their rarity, diverse histopathology, and complex anatomy of the presacral region. Imaging plays a crucial role in the diagnosis and management of these tumors, enabling accurate characterization, localization, and preoperative planning. However, the evaluation of pediatric presacral tumors requires a multimodal approach, combining various imaging techniques and expertise to overcome the diagnostic complexities. This article explores the diagnostic challenges encountered in imaging pediatric presacral tumors and highlights the importance of a comprehensive imaging strategy.

Pediatric presacral tumors are extremely rare, accounting for less than 1% of all pediatric tumors [1–3]. The wide range of histopathological subtypes further complicates their diagnosis. Common types include sacrococcygeal teratomas, neuroblastomas, Ewing sarcomas, rhabdomyosarcomas, and primitive neuroectodermal tumors [2]. Each subtype presents unique imaging features, making accurate diagnosis challenging. The radiologist's familiarity with these entities and their

imaging characteristics is essential for accurate interpretation.

Complex anatomy of the presacral space:

The presacral space contains various vital structures, including the rectum, sacrum, nerves, blood vessels, and adjacent pelvic organs [1]. The proximity of these structures to presacral tumors can influence the extent of disease and surgical planning [4]. Accurate delineation of tumor borders and their relationship with adjacent structures is crucial to determine the feasibility of complete resection and the potential for functional morbidity. Advanced imaging techniques, such as magnetic resonance imaging (MRI) and computed tomography (CT), with multiplanar reconstructions, help visualize these anatomical relationships.

The precise location of the tumor within this region provides important clues about its origin, relationship with nearby structures, and potential for invasion or compression of vital structures like the rectum, bladder, or blood vessels. Understanding the anatomical context helps in formulating a differential diagnosis and determining the appropriate diagnostic approach. Primary presacral tumors are more likely to arise from developmental remnants or embryonic tissues within the presacral region [1], whereas secondary tumors may indicate a primary tumor elsewhere in the body. Different types of tumors can occur in the presacral region, each with its own characteristic location and epicenter [4]. For example, sacrococcygeal teratomas primarily originate from the coccygeal region, whereas neuroblastomas tend to arise from the sympathetic ganglia. Identifying the specific location and epicenter of the tumor can help narrow down the possible tumor subtypes and guide further diagnostic tests. The location and epicenter of the tumor provide critical information about its extent of infiltration and potential involvement of nearby structures. For example, a tumor originating from the anterior presacral region may have a higher propensity to invade the bladder, while a tumor arising from the posterior presacral region may extend into the sacrum or coccyx. Evaluating the extent of infiltration and extension is essential for accurate staging and treatment planning. Certain tumor subtypes in the presacral region are associated with specific syndromes or congenital anomalies. For example, sacrococcygeal teratomas are often associated with chromosomal abnormalities or anomalies of midline structures [4]. Knowledge of the tumor's location and potential associated syndromes can aid in formulating the differential diagnosis and guide further genetic or syndromic evaluations.

Imaging modalities:

a) Ultrasonography: Ultrasonography is often the initial imaging modality used to evaluate pediatric presacral tumors due to its accessibility and lack of ionizing radiation. It aids in the assessment of tumor vascularity, tumor composition (solid vs. cystic components), and presence of calcifications. However, ultrasound has limited ability to evaluate deep pelvic structures and may not provide comprehensive anatomical information.

b) Computed Tomography (CT): CT is a standard tool for the evaluation of bony involvement and extent of the tumor. It is useful for evaluating bony structures and calcifications associated with presacral tumors. It provides detailed information on bone erosion, invasion, and potential complications such as pelvic abscess formation. CT is particularly valuable in cases where MRI is contraindicated or inaccessible. However, CT involves ionizing radiation and should be used judiciously, especially in pediatric patients.

c) Magnetic Resonance Imaging (MRI): MRI is considered the modality of choice for evaluating pediatric presacral tumors. It provides superior soft tissue contrast, multiplanar imaging capabilities, and the ability to characterize tumor components. MRI can accurately determine tumor extension, invasion of adjacent structures, and potential involvement of neural structures, aiding in surgical planning. Diffusion-weighted imaging (DWI) and dynamic contrast-enhanced MRI may provide additional information on tumor aggressiveness and vascularity.

d) Nuclear Imaging: PET-CT and PET-MRI play a significant role in the evaluation of pediatric presacral tumors. They provide functional information by detecting metabolic activity, aiding in differentiation between benign and malignant tumors, detecting lesions and metastases, assessing treatment response, differentiating residual tumor from post-treatment changes, and assisting in radiation therapy planning. These advanced imaging techniques provide valuable information that complements anatomical imaging, helping in accurate diagnosis, treatment planning, and monitoring of pediatric presacral tumors.

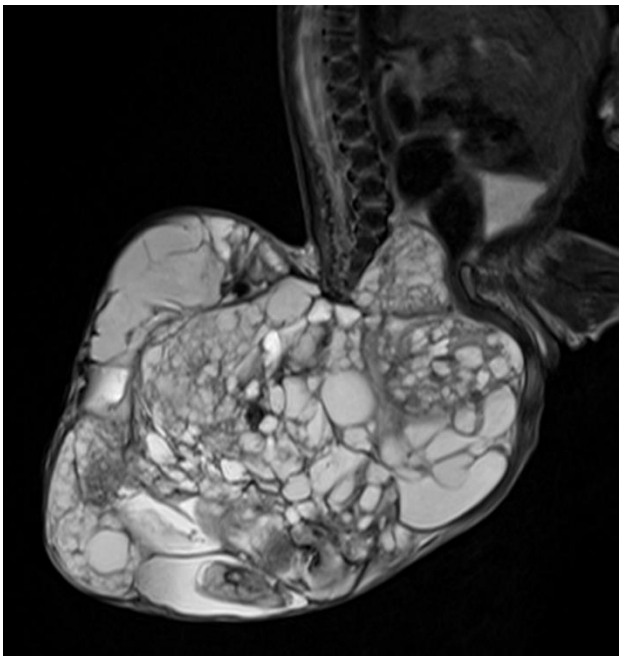
Differentiating benign and malignant tumors:

Accurate differentiation between benign and malignant presacral tumors is crucial for appropriate treatment planning [2]. Benign and malignant presacral tumors can often have similar imaging appearances, making it difficult to differentiate them based solely on imaging findings. Both types of tumors can present as well-defined

masses with heterogeneous internal characteristics, such as cystic or solid components, enhancing soft tissue, or necrotic areas. While certain imaging characteristics, such as invasion into adjacent structures or metastatic spread may suggest malignancy, these findings are not always present or definitive [5]. Additionally, some benign tumors can mimic malignant features on imaging, further complicating the differentiation.

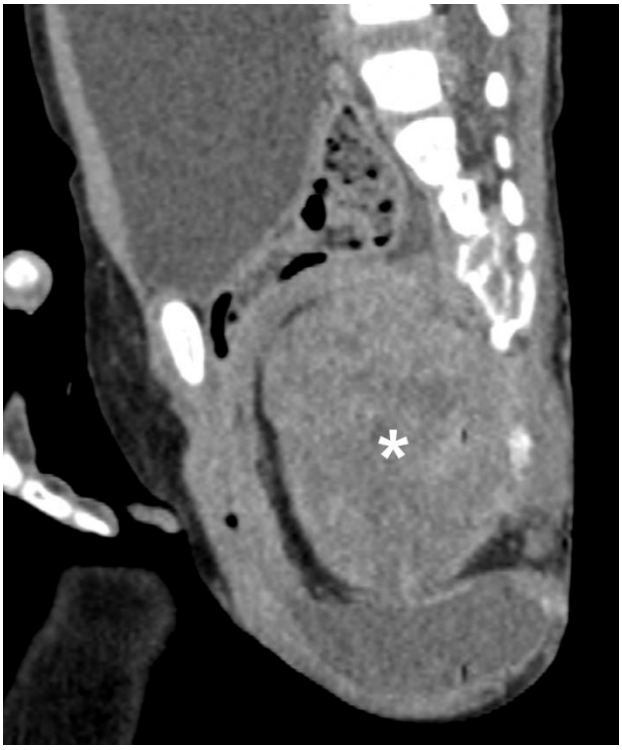
Both benign and malignant presacral tumors can exhibit heterogeneity in terms of their internal composition, vascularity, and enhancement patterns. This heterogeneity can make it challenging to accurately characterize the tumor based on imaging alone. While larger tumors may raise suspicion for malignancy, small malignant tumors or benign tumors with aggressive features can be challenging to detect or differentiate from surrounding structures using imaging techniques. Some pathologic examples are presented in Figures 1 to 7.

Fig. 1 Immature sacrococcygeal teratoma in a 24-day-old neonate. Sagittal T2-weighted MR image shows a complex solid cystic tumor in the presacral space with extrapelvic extension and containing intralesional fat, soft tissue and hemorrhage



Advanced imaging techniques such as diffusion weighted imaging (DWI), dynamic perfusion imaging and nuclear imaging techniques such as positron emission tomography (PET) may aid in distinguishing between benign and malignant tumors by assessing cellular activity and metabolic characteristics [6].

Fig. 2 Malignant sacrococcygeal teratoma in a 5-year-old child. Sagittal reformatted contrast enhanced CT image shows a presacral tumor (asterisk) with heterogeneously enhancing solid component, intralesional fat and cystic areas. Associated destruction of the sacral vertebrae is also seen

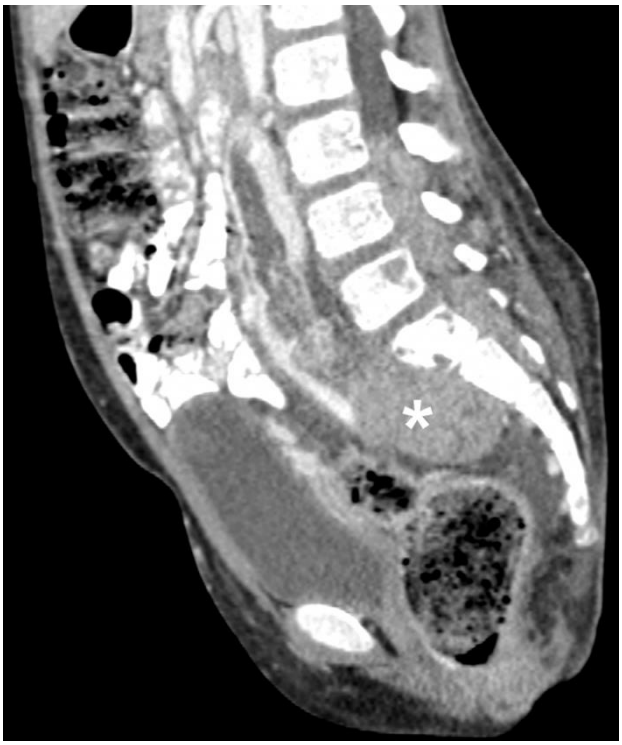


Assessment of tumor response and recurrence:

Monitoring the response to treatment and detecting tumor recurrence are crucial aspects of managing pediatric presacral tumors. Imaging modalities, particularly MRI, play a vital role in assessing treatment response by evaluating changes in tumor size, vascularity, and necrosis after neoadjuvant therapy. In addition, regular follow-up imaging helps in early detection of recurrent disease, enabling prompt intervention and improving patient outcomes.

Presacral tumors are located deep in the pelvis, which makes them difficult to access and evaluate using conventional imaging techniques. Complex anatomical structures in the pelvic region, such as the bladder, rectum, and blood vessels, can further obscure the tumor boundaries and hinder accurate assessment. They can vary significantly in size and shape. Some tumors may be large and well-defined, while others can be small and irregularly shaped. This variability can make it challenging to accurately measure the tumor size and assess the response to treatment over time.

Fig. 3 Rhabdomyosarcoma in a 12-year-old girl. Sagittal reformatted contrast-enhanced CT image shows an infiltrative heterogeneously enhancing soft tissue tumor (asterisk) extending into the sacral spinal canal with destruction of the lumbar and sacral vertebrae



Currently, there is a lack of specific imaging biomarkers that can reliably indicate tumor response or recurrence in presacral tumors. Conventional imaging modalities, such as computed tomography (CT) and magnetic resonance imaging (MRI), primarily rely on changes in tumor size and morphology to assess treatment response. However, these changes may not always correlate with the actual tumor response, especially in cases where the tumor undergoes necrosis or fibrosis without significant reduction in size.

Following treatment, presacral tumors may exhibit various post-treatment changes, such as fibrosis, inflammation, or fluid collections. These changes can mimic or mask tumor recurrence, leading to false-positive or false-negative imaging results. Distinguishing between treatment-related changes and tumor recurrence can be challenging, requiring close collaboration between radiologists, oncologists, and surgeons. Functional imaging techniques, such as positron emission tomography (PET) and diffusion-weighted imaging (DWI), can provide valuable information about tumor metabolism and cellular activity. However, these techniques may not always be readily available in all healthcare settings or may have limited sensitivity and specificity in presacral tumors, further complicating the assessment of tumor response and recurrence.

Fig. 4 Clear cell sarcoma in a 10-year-old boy. Axial T2-weighted MR image shows a lobulated hyperintense tumor (asterisk) in the presacral space showing areas of necrosis and hemorrhage within and lytic destruction of the sacrum

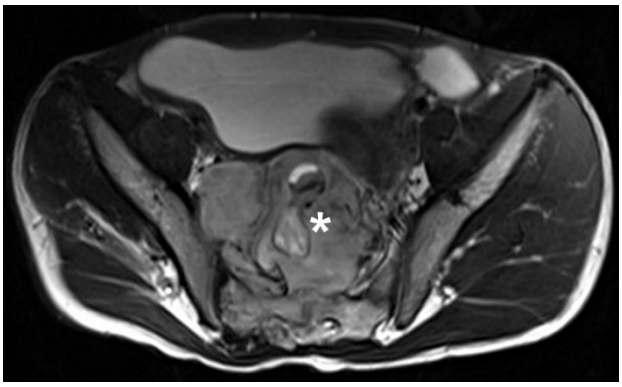
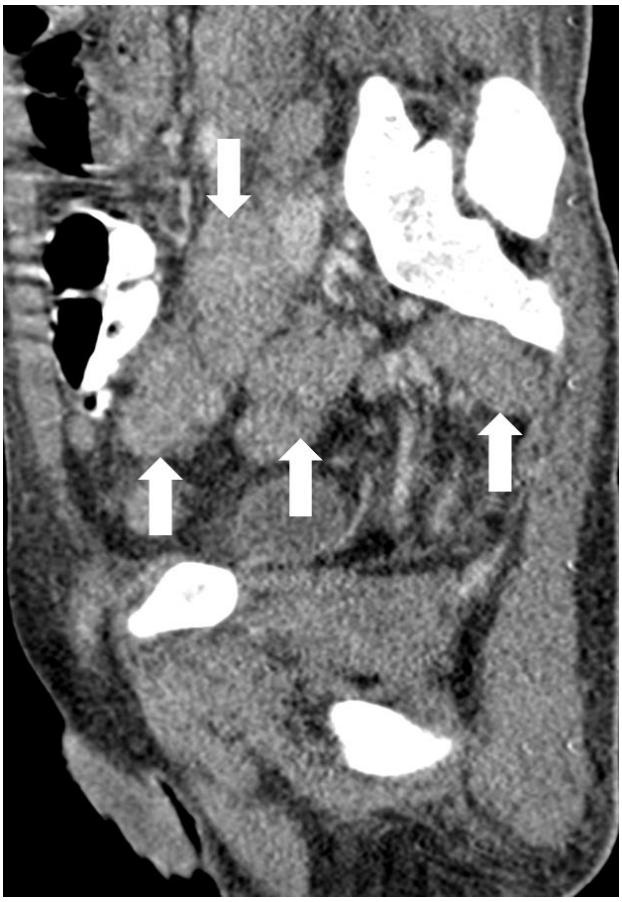


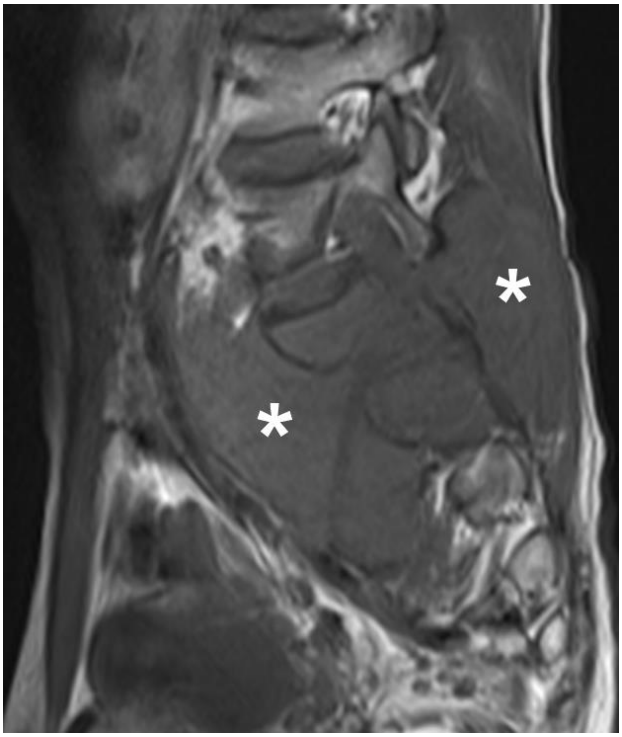
Fig. 5 Lymphoma in a 9-year-old child. Sagittal reformatted contrast-enhanced CT image shows multiple enlarged homogeneously enhancing lymph nodal masses (arrows) in the presacral region



Interdisciplinary approach and expertise:

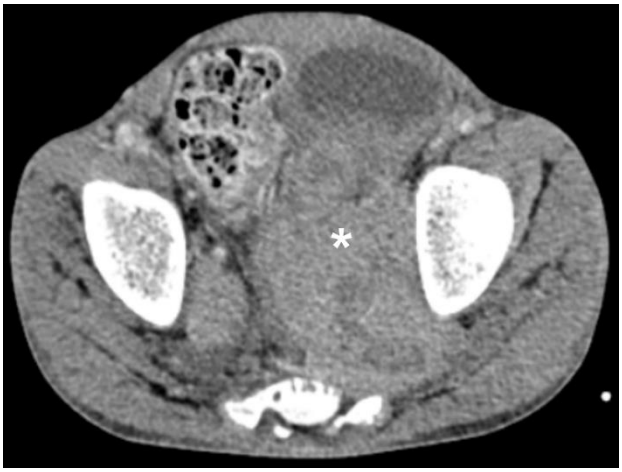
Due to the complex nature of pediatric presacral tumors and the challenges associated with their imaging, a multidisciplinary approach is essential. Radiologists, pediatric surgeons, pediatric oncologists, and pathologists must collaborate to ensure accurate diagnosis, appropriate staging, and optimal treatment planning [7]. Each discipline brings unique perspectives and expertise to ensure a comprehensive evaluation of the tumor. Interdisciplinary collaboration facilitates the development of an optimal treatment plan. The expertise of radiologists with a strong understanding of the imaging characteristics of pediatric presacral tumors is crucial for accurate interpretation and guiding clinical decision-making. Pediatric oncologists contribute their expertise in systemic therapies, such as chemotherapy or targeted therapies, while surgeons determine the feasibility and extent of surgical resection. The combined knowledge and perspectives ensure a tailored approach to maximize treatment effectiveness. Regular imaging evaluations are necessary, and an interdisciplinary team can collectively interpret imaging findings and correlate them with clinical outcomes.

Fig. 6 Ewing's sarcoma in an 11-year-old child. Sagittal T1-weighted MR image shows an expansile lesion involving the sacrum (asterisk) with associated soft tissue extending anteriorly into the presacral space and posteriorly into the sacral spinal canal and the paraspinal soft tissue



Effective communication and coordination among multiple specialists can be challenging, especially when managing complex cases. Different disciplines may use specialized terminology and have unique perspectives, necessitating clear and efficient communication to ensure a shared understanding of the case. Ensuring that all team members are up to date with the latest advancements and research in their respective fields is essential for optimal decision-making and patient care. Coordinating schedules and availability of team members can pose logistical challenges, particularly in time-sensitive cases. Achieving consensus in diagnosis and treatment planning is crucial to avoid conflicting recommendations and ensure consistent care for the patient. Addressing these challenges necessitates effective communication channels, regular team meetings, and a culture of collaboration and mutual respect. Continuous education and knowledge-sharing activities among team members can help mitigate differences in expertise. Emphasizing patient-centered care and shared decision-making further enhance the effectiveness of the interdisciplinary approach in managing pediatric presacral tumors.

Fig. 7 Neuroblastoma in a 5-year-old child. Axial contrast-enhanced CT image shows a heterogeneously enhancing infiltrative tumor in the presacral space (asterisk) towards the left of the midline with areas of necrosis within



CONCLUSION

Imaging pediatric presacral tumors presents several diagnostic challenges due to their rarity, diverse histopathology, and complex anatomy of the presacral space. An integrated imaging approach that combines various modalities such as ultrasound, MRI, and CT is crucial for accurate characterization, localization, and preoperative planning. Differentiating between benign and malignant tumors, assessing treatment response, and

detecting recurrence are additional challenges that require specialized imaging techniques. A multidisciplinary approach and the expertise of radiologists, along with other medical specialists, are vital for overcoming these diagnostic challenges and providing optimal management for pediatric patients with presacral tumors. Future research and advancements in imaging technology will further enhance our ability to accurately diagnose and manage these rare tumors, ultimately improving patient outcomes.

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