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Review Article

Controversies in presacral tumors management

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ABSTRACT

Presacral tumors are rare lesions of the retrorectal space that can present diagnostic and therapeutic difficulty because of their anatomic location and the different tissue types and etiology. Although the diagnosis and management of these tumors has evolved in recent years, several points still to be addressed in order to improve perioperative diagnosis and treatment. In the upcoming we will try to highlight some controversial points; the preoperative biopsies, neoadjuvant therapy, the necessity of surgery and the role of minimally invasive surgeries of presacral tumors.

Controvérsias no tratamento de tumores pré-sacrais

Palavras-chave:
Tumor pré-sacral
Biópsia pré-operatória
Terapia neoadjuvante

RESUMO

Tumores pré-sacrais são lesões raras do espaço retrorretal que podem trazer dificuldades diagnósticas e terapêuticas por causa de sua localização anatômica e também pelos diferentes tipos de tecidos e etiologia. Embora nos últimos anos o diagnóstico e tratamento desses tumores tenham evoluído, diversos pontos ainda devem ser estudados com vistas à melhora do diagnóstico e tratamento no perioperatório. Mais adiante, tentaremos esclarecer alguns pontos controversos; biópsias pré-operatórias, terapia neoadjuvante, a necessidade de cirurgia e o papel das cirurgias minimamente invasivas para os tumores pré-sacrais.

Introduction

Presacral tumors are heterogeneous and relatively rare. Given these characteristics, the clinical presentation, natural history and treatment of presacral tumors is not well understood.

The presacral space is bounded by the peritoneal reflection and the recto-sacral fascia. The embryologic development at this anatomical site is complex due to the presence of totipotential germ cells that differentiate into 3 germ cell layers. This in turn leads to the development of different types of tissues including connective tissue, nerves, fat, and blood vessels. The multitude of tissue types gives rise to numerous tumor pathologies. In addition, because the presacral space has diverse histopathological findings, physicians from

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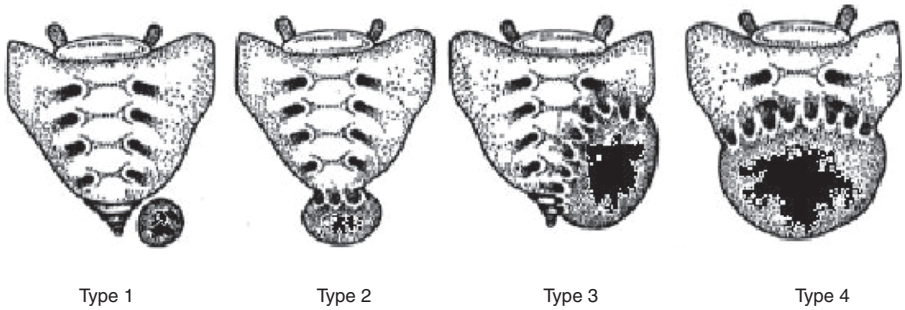

Fig. 1 – Classification based on tumor location.

diverse subspecialties including colorectal surgeons, obstetricians and gynecologists, urologists, neurosurgeons, and orthopedics are often involved in patient care.

Pre-sacral tumors were reported for the first time in the middle of the 19th century. Kiderlen et al. reported six cases in 1899 including a teratoma, which was the first reported case by Emmerich. The first pre-sacral tumor was resected by Dr. Middledorpf, and this tumor was named after him for many years.[1]

The true incidence of pre-sacral tumors in the general population is hard to assess because many reports come from tertiary centers and so do not represent the true incidence. The estimated incidence is about 1 case in every 40,000 hospital admissions. Most of the publications are individual case reports and small case series.[2–4]

As a result of the low incidence, there is no uniform classification system for these lesions. Four years ago, Uhlig and Johnson suggested a classification for these tumors according to its tissue of origin.[2] Physicians at Tel Aviv Medical Center proposed another classification system based on whether the lesion is congenital or acquired as well as whether it is benign or malignant.[5]

Yet another classification system is based on tumor location and its communication with the sacrum and/or coccyx (Fig. 1). Using tumor location as the basis for the classification facilitates the surgical approach.[6] According to this classification type 1: the lesion is located at the coccyx level (below S3) and separated from the bony trunk of sacrococcyx, type 2 as type 1 but has connection with the coccyx sacrum. Type 3 the lesion involves the sacrum at or above the S3 nerve root unilaterally, and type 4 when large communication with the sacrum at or above S3 bilaterally.

Further, the above classification system reflects postoperative complications: resection of type 1 and 2 usually does not have neurological sequela and types 3 and 4 usually result in temporary or permanent incontinence.[6]

In recent years, the classification system proposed by Dozois and Jacofsky from the Mayo clinic is most commonly used. According to this classification (Table 1), tumors are divided into 5 categories which are further grouped into benign and malignant.[7]

About two thirds of all the pre-sacral tumors are congenital. Most of those are cystic and benign. 10% are of neurogenic in origin, 5–10% are of bone origin and about 15% are from other origins, including metastasis.[8] In general, benign lesions are more common in females, while malignanttumorsare equal in both sexes. The most common lesions are cystic, benign, asymptomatic, and found incidentally. These lesions most often present with rectal fullness, painful defecation, or dysuria.[3]

Table 1 – Classification of presacral retrorectal tumors.

Source of origin	Histopathology
Congenital or developmental	
Benign	Developmental cysts
	Dermoid cysts
	Epidermoid cysts
	Tail gut cysts
	Enteric (rectal) duplication
	Anterior sacral meningocele
	Teratoma
	Adrenal rest tumors
Malignant	Chordoma
	Teratocarcinoma
Inflammatory	Granulomas (foreign body)
	Perineal/pelvirectal abscess or fistula
Neurogenic	
Benign	Neurofibroma
	Neurolemmoma (schwannoma)
	Ganglioneuroma
Malignant	Ependymoma
	Ganglioneuroblastoma
	Neurofibrosarcoma
Osseous	
Benign	Osteoma Sacral bone cyst
	Osteoblastoma
	Osteogenic sarcoma
	Giant cell tumor
Malignant	Ewing's tumor
	Chondromyxosarcoma
	Osteogenic sarcoma
	Myeloma
Miscellaneous	
Benign	Lipoma
	Fibroma
	Leiomyoma
	Hemangioma
	Endothelioma
	Desmoid tumor
	Lymphangioma
	Ectopic kidney
Malignant	Fibrosarcoma
	Liposarcoma
	Leiomyosarcoma
	Metastatic disease

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