Educational Forum

Pelvic retroperitoneal masses in children: what does the radiologist need to know?

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ABSTRACT

Pelvic retroperitoneal tumors pose a diagnostic and therapeutic challenge due to their rarity, late presentation, and complex anatomical location in the retroperitoneum. This article reviews the diagnosis and management of retroperitoneal tumors in the pelvis, and highlights the implications of imaging on management. This study was conducted in a tertiary care hospital of western Gujarat over a period of 2 years. We reviewed the various retroperitoneal masses in pelvis in pediatric age group and then discuss potential diagnostic and therapeutic challenges.

Keywords: pediatric pelvic retroperitoneal masses; malignancies; neurogenic tumors

INTRODUCTION

Retroperitoneal tumors in the pelvis pose a diagnostic dilemma for interpretation of surgical pathology, because of its rarity, high recurrence rate after surgical excision, and unpredictable response to adjuvant therapy.[1] Due to the inaccessible anatomical position and late clinical presentation, the diagnosis is often delayed, in the advanced stages of local or systemic spread. Therefore, many of these cases do not benefit from complete surgical removal, and this renders a real diagnostic and therapeutic predicament.[2] We review the diagnosis and management of retroperitoneal tumors in the pelvis, and highlight the potential diagnostic and therapeutic challenges.

AIMS AND OBJECTIVES:

1. To discuss anatomy of pelvic retroperitoneal structures.
2. To discuss etiology and pathogenesis of pelvic retroperitoneal masses in children.
3. To review imaging features with implications on management

MATERIAL AND METHODS

The present study was a prospective longitudinal study The study was conducted in the Radiology department of a tertiary care hospital of western Gujarat. The study was conducted over a period of 2 years.
To review this topic, the research articles were studied from PubMed and Google Scholar with the aims and objectives as mentioned above. The literature search was conducted by 2 independent authors in August 2021. All the patients of pediatric age group having pelvic retroperitoneal masses on imaging were included in the study and analyzed.

ANATOMY:

The prevesical space is a large potential space that lies between the infraumbilical lower anterior abdominal wall anteriorly and the umbilicovesical fascia posteriorly.[2] The perivesical space is located below the pelvic peritoneal reflection, is surrounded by the umbilicovesical fascia, and contains the urinary bladder, uterus (in females), urachus, and obliterated umbilical arteries. Perirectal space surrounds the rectum and is divided into the central

retrorectal space and the peripheral presacral space by the intervening presacral fascia.
The masses arising from the retroperitoneum in pelvis can be of varying nature of the tissue or varying etiology. Based on the etiology/nature of the tissue on imaging, we can further classify the lesions. The examples of each are given in the table 1

Table 1: Etiology/nature of tissue and its corresponding lesions

ETIOLOGY/NATURE OF TISSUE	LESION
Congenital and developmental masses	Sacrococcygeal teratoma, Tailgut cyst, Anterior sacral meningocele, Rectal duplication cyst, Hemato/hydrocolpos
Vascular masses	Lymphatic, Venous or Capillary, and Arteriovenous malformations
Neurogenic masses	Neuroblastoma, Ganglioneuroblastoma, Ganglioneurom, Neurofibroma, Schwannoma
Fat containing masses	Lipoblastoma
Inflammatory masses	Abscess
Mesenchymal masses	Rhabdomyosarcoma and undifferentiated sarcoma, Fibroma, Sarcoma, Ewing's sarcoma
Extension of bone tumors	Giant cell tumor, Aneurysmal bone cyst, Chordoma, other bone tumors

CONGENITAL AND DEVELOPMENTAL MASSES: SACROCOCCYGEAL TERATOMA (Fig. 1)

Sacrococcygeal teratoma is the most common presacral germ cell tumor in children and the most common solid tumor in neonates.[1] It is classified into 4 surgical subtypes as per Altman/AAP classification. Type I is primarily external. It seen in 47% of the cases. These patients have the best prognosis among all the types. Type II (34%) is dumbbell shaped, it has equal external and internal portions. Type III (9%) is primarily internal within abdomen/pelvis. Type IV (10%) is entirely internal and has no visible external component; it has the worst prognosis.

The tumor arises from totipotential cell rests at caudal spine/notochord (Hensen node). On imaging it appears as a large heterogeneous sacral mass which variably contains calcifications, mixed solid and cystic components, fat-debris levels, bone, hair, teeth, or cartilage. MR is used for surgical planning and CT is used to look for bone destruction and calcification. Surgery alone is curative if entire benign tumor and coccyx are removed. However, it recurs if coccyx not resected. It is often associated with Currarino triad which includes anorectal anomalies, caudal regression syndrome, epidermoid/dermoid tumor or other tethering lesion.

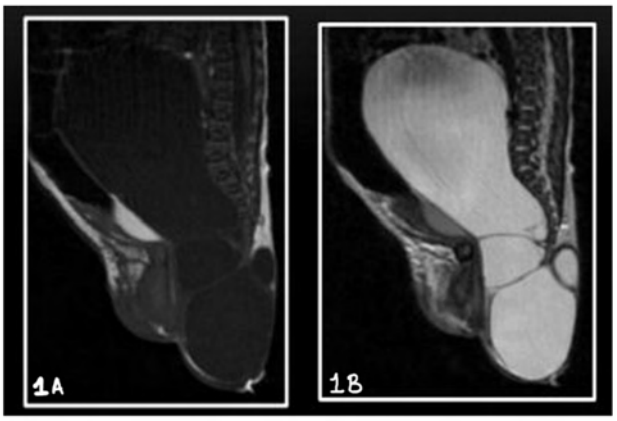

Fig 1a shows T1 hypointense and Fig 1b shows T2 hyperintense multiloculated mass with internal and external portions; Cystic sacrcoccygeal teratoma.

HEMATOCOLPOS/HEMATOMETROCOLPOS

Hematocolpos/hematohydrocolpos results from distension of uterus and vagina by accumulated blood; most common cause being imperforate hymen. In teen it presents with primary amenorrhoea and cyclic pelvic pain. Imaging with US shows mixed echogenicity material within uterine &/or vaginal cavities with no internal flow. Further MR is best to confirm blood products, absence of solid mass and to clarify anatomy. Transperineal view may aid imaging cause of obstruction. Treatment includes treating the underlying condition. Avoid aspiration (due to risk of infection).

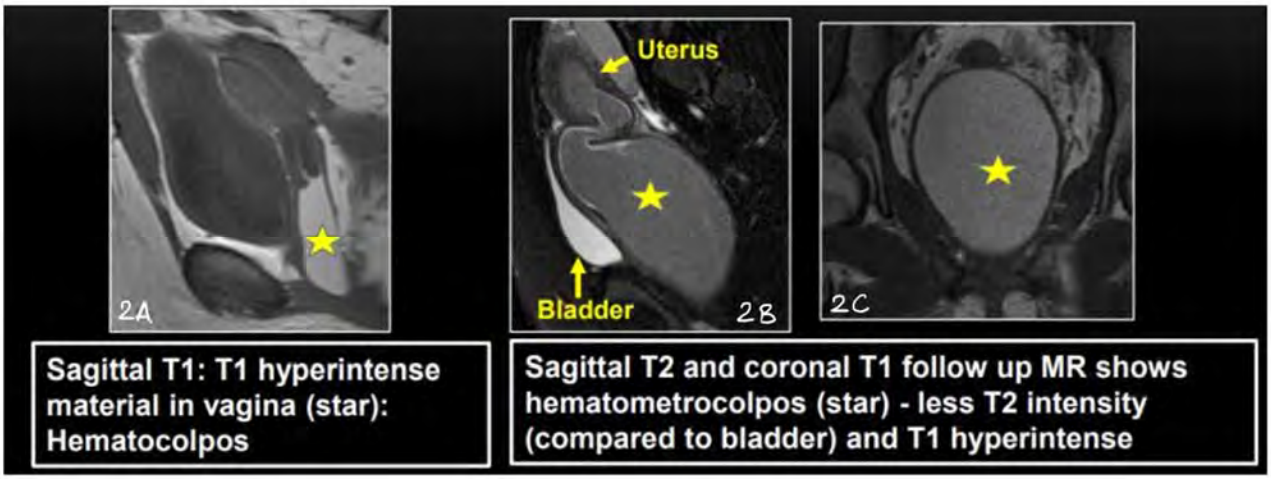

Fig. 2A showing T1W and Fig.2 B and 2C showing T2W MR image in a 15 year old girl showing hematometrocolpos.

ANTERIOR SACRAL MENINGOCELE

An anterior sacral meningocele (Fig. 3) is a congenital abnormality that arises from herniation of the cerebrospinal fluid–filled dura mater through a sacral foramen or a defect in the sacral bone. It may be accompanied by symptoms, but it is usually asymptomatic in older children.[1] It may be complicated by mass effect, neurologic compromise, meningitis, or rupture of the meningocele
Radiography and CT is useful to look for vertebral scalloping, hypoplasia, and aplasia. MR is the modality of choice for the assessment of sacral defects, neck of meningocele, hernia sac, nerve roots (which appear as areas of intermediate signal intensity on T2-weighted images), and dysraphism.

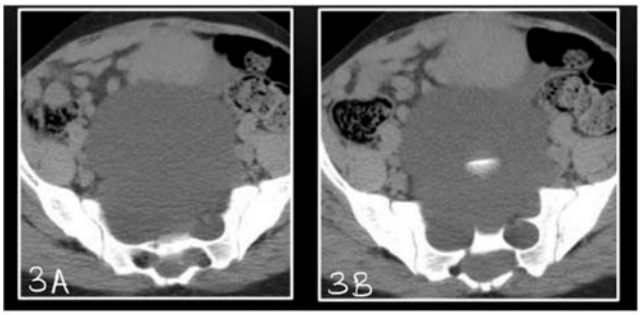

Fig. 3: Presacral water attenuation mass with multilobulated contour and extension to sacral foramen (Fig 3B) with scalloping; Anterior sacral meningocele.

TAILGUT CYST

Tailgut cysts (Fig. 4) are rare congenital abnormalities in the presacral space and may be manifested in childhood or adulthood.[2] The cysts may be uni- or multilocular and are lined with various epithelia, usually in combination. Mucin-secreting cells are responsible for the mucoid contents of the cysts. The cysts can be lined by a variety of epithelial types.
Pelvic ultrasound demonstrates cystic lesion with homogenous echoes due to presence of gelatinous material. CT would show a presacral cystic mass without osseous involvement; calcification may be seen in the cyst wall. On post contrast T2 fat sat images there is a rim enhancement with a T2 hyperintense mass. Surgical excision is the treatment of choice.

RECTAL DUPLICATION CYST

Rectal duplication cyst (Fig. 5) is almost always located posterior to the rectum.[2] Up to 45% of all rectal duplication cysts are associated with a posterior midline fistula to the anus or perianal region that can be seen on physical exam. If a fistula is noted, further evaluation with injected contrast may confirm the presence of a rectal duplication cyst.
Chronic constipation, rectal bleeding, rectal prolapse, hemorrhoids, and perirectal abscess formation are generally the presenting features. On CT there is a well-defined thin-walled uni- or multilocular presacral lesions with low attenuation and without contrast enhancement. Surgical excision is the treatment.

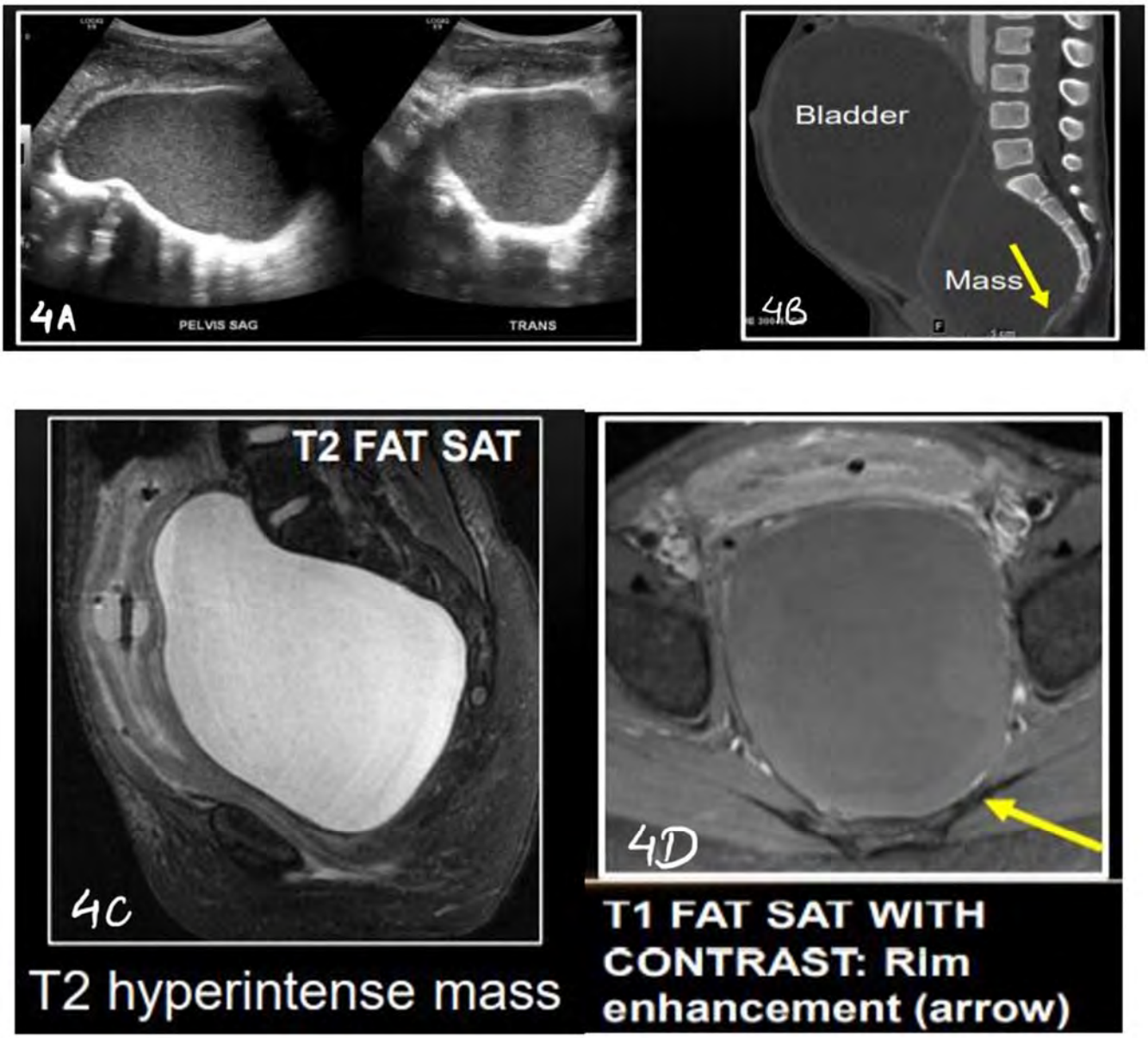

Fig. 4: Ultrasound(Fig. 4A), CT (Fig. 4B)and MR (Fig. 4C, 4D) images showing Tailgut cyst in a 12 year old with rectal fullness.

VASCULAR MASSES:

Vascular masses include lymphatic, venous (Fig. 6) or capillary, and arteriovenous malformations. MR is the modality of choice for diagnosis and to delineate the size and extent for preoperative evaluation and pre-embolization planning. Association include Klippel

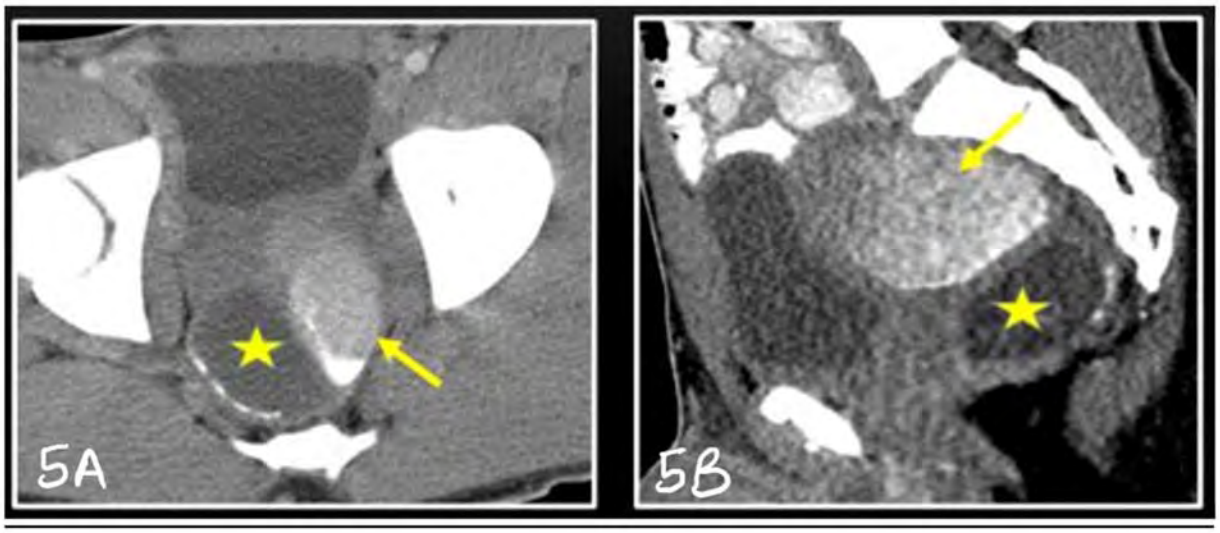

Fig. 5: Axial (Fig. 5A) and sagittal CT(Fig. 5B) demonstrates a nonenhancing fluid attenuation structure with rim calcification (yellow star) in close apposition to rectum (arrow): Rectal duplication cyst.

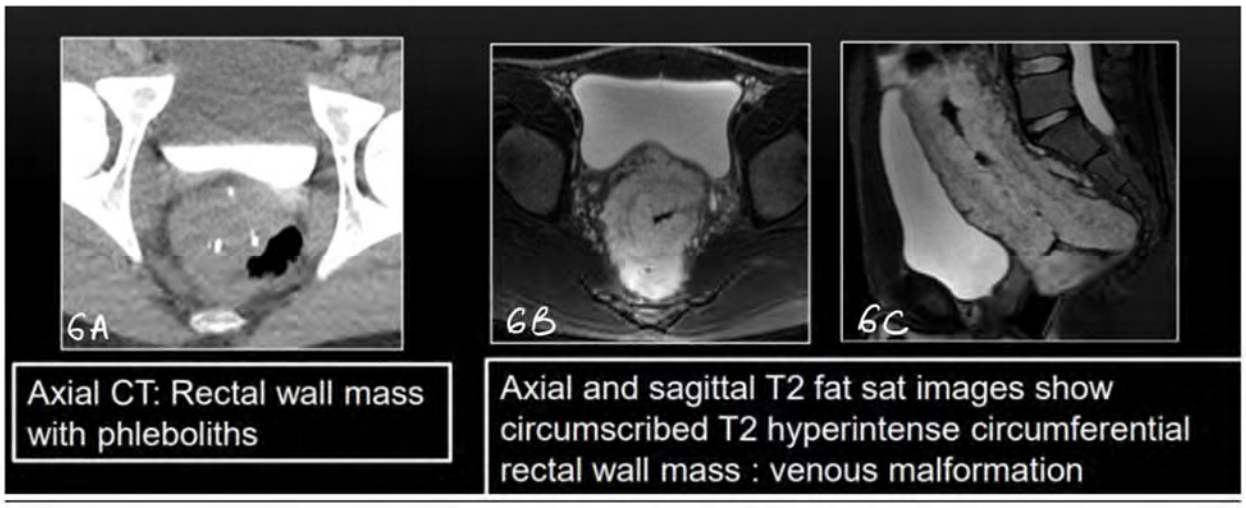

Fig. 6: CT(Fig. 6A) and MR (Fig. 6B) images showing venous malformation in a 12 year old male.

Trénaunay-Weber syndrome, triad of capillary malformations, soft tissue or bone hypertrophy, and venous varicosities or malformations.

LYMPHANGIOMA:

Lymphoma (Fig. 7) is a congenital benign malformation of lymphatic system due to failure of embryologic lymphatic development. Computed tomography shows circumscribed cystic multiloculated non-enhancing mass with water density; Soft lesions without mass effect: Easily indented by surrounding structures. MR shows hypointense lesion on T1WI MR and appears hyperintense on T2WI. Open or laparoscopic surgical resection if lesion is symptomatic is the treatment

NEUROGENIC MASSES: GANGLIONEUROMA (Fig. 8)

It is a rare benign neurogenic mass. On imaging, it tends to be homogenous except for calcifications (seen in 2/3rd of cases). MR is superior to CT in its depiction of involvement of ganglioneuroma when in the spine or neural foramina.

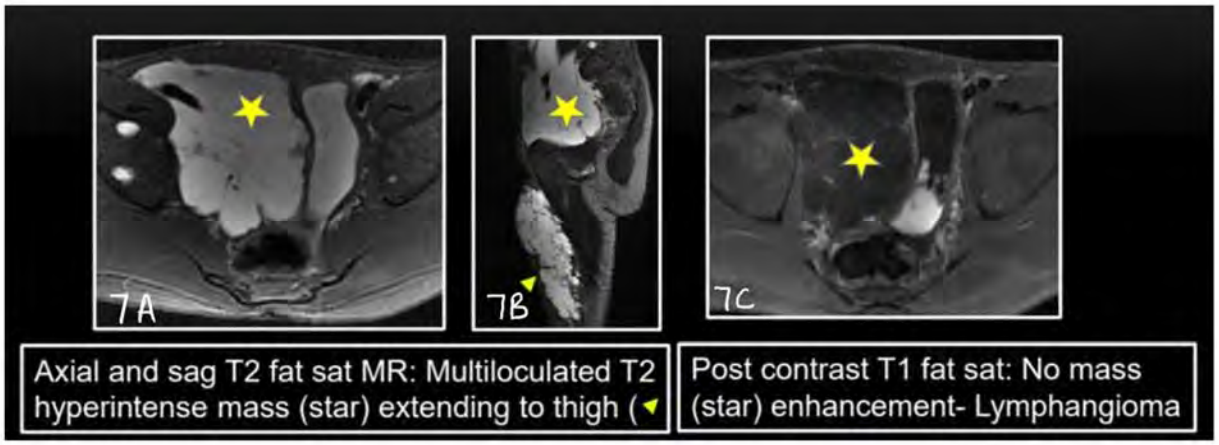

Fig. 7A,7B,7C: Multiplanar MR images show lymphangioma in a 10 year old female

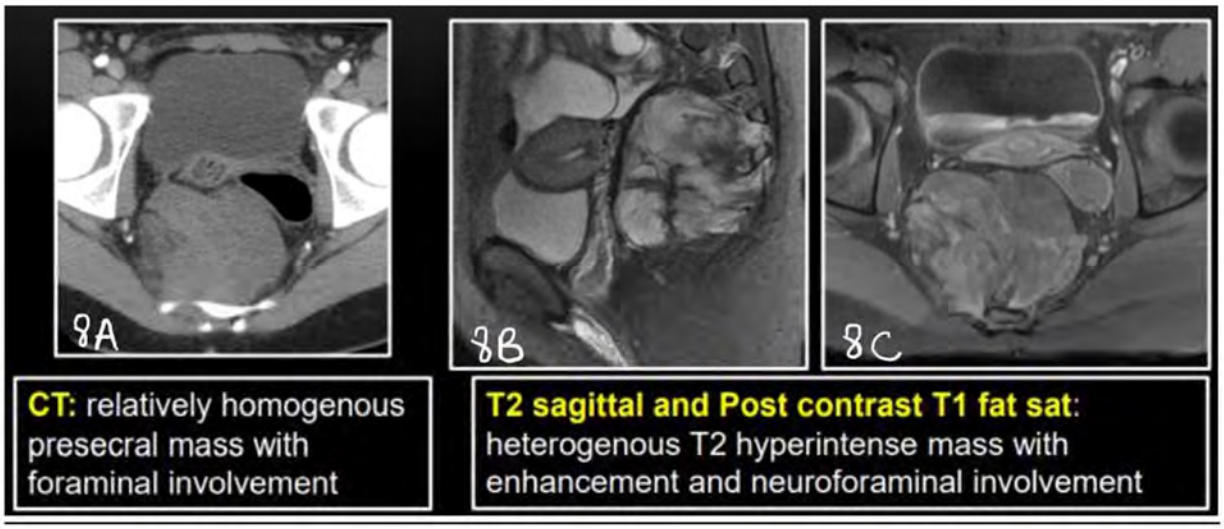

Fig. 8: CT(Fig. 8A) and MR (Fig. 8B, 8C) images show neurogenic mass in the pelvic retroperitoneum. Histopathology confirmed ganglioneuroma.

NEUROFIBROMA

Multiple neurofibromas are seen in in Type 1 Neurofibromatosis, a common autosomal dominant disease that affects one person in every 2000– 4000.[5] NF1 with abdominopelvic involvement tends to arise in the retroperitoneal, mesenteric, and paraspinal regions; it may be difficult to distinguish from adenopathy at CT. The multiplanar capabilities of MR imaging, particularly with T2 weighting, make this modality helpful in evaluating affected patients and making the diagnosis.[6] MR T2-weighted images shows the lesion has a target like appearance, with a hyperintense rim of myxoid material and a central zone of low signal intensity that indicates a fibrous core.

FAT CONTAINING MASSES:

Fat containing masses like lipoblastoma (Fig 10) are soft-tissue neoplasm that arises from fetal adipose tissue.[4] A well-circumscribed soft-tissue lesion that predominantly contains fat is a lipoblastoma is seen on CT. Lesions with an infiltrative growth pattern on imaging are identified as lipoblastomatosis. MR is the preferred tool for both diagnosis and preoperative evaluation. Absence of calcification differentiates it from teratoma. Complete surgical excision is the treatment of choice.

INFLAMMATORY MASSES:
ABSCESS (Fig. 11)

Unlike neoplastic processes, the lytic lesion associated with osteomyelitis is often ill defined and poorly marginated with presence of extensive soft-tissue inflammatory change, with or without abscess formation. Sclerotic foci or osseous sequestra may also be observed in chronic infection.

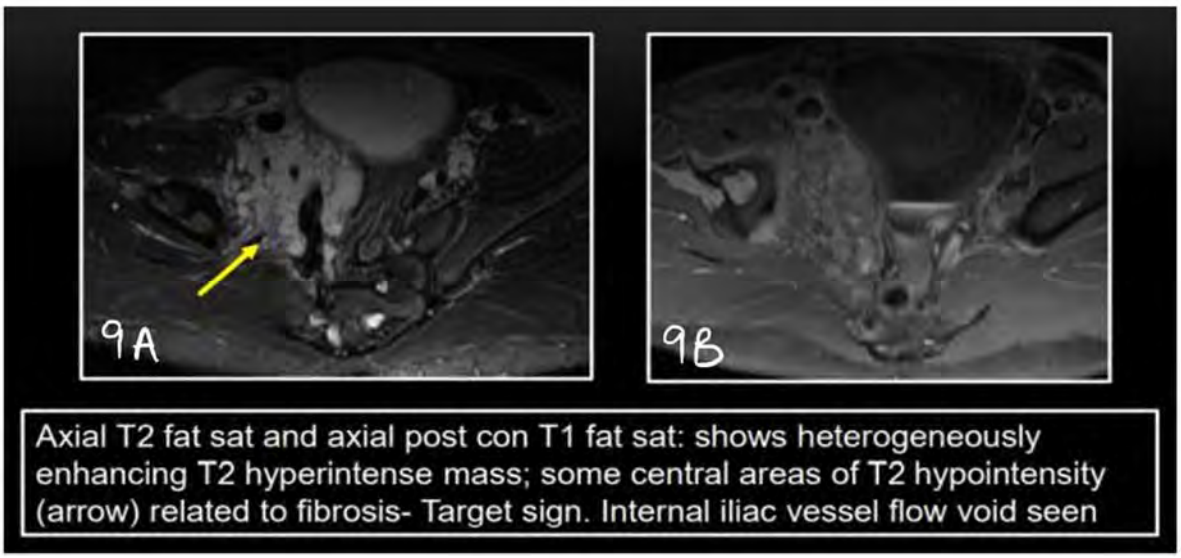

Fig. 9: Axial MR post contrast(Fig. 9B) images shows neurofibroma in a 9 year old child.

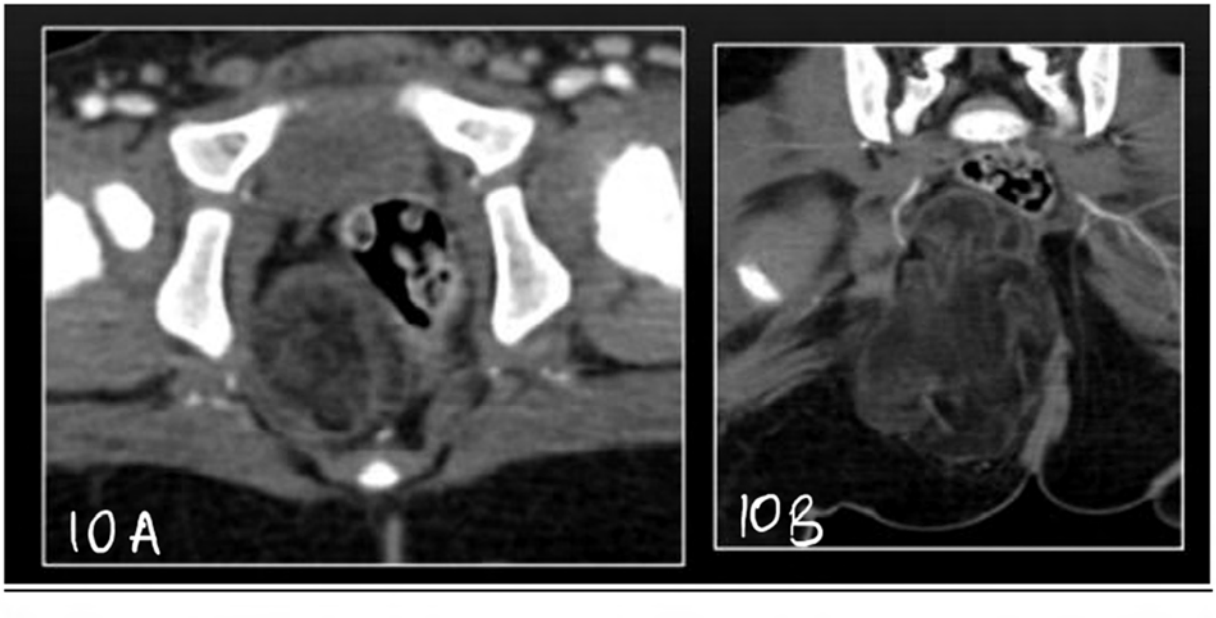

Fig 10: CT images shows a fat containing mass posterior to rectum with extension to ischiorectal fossa.

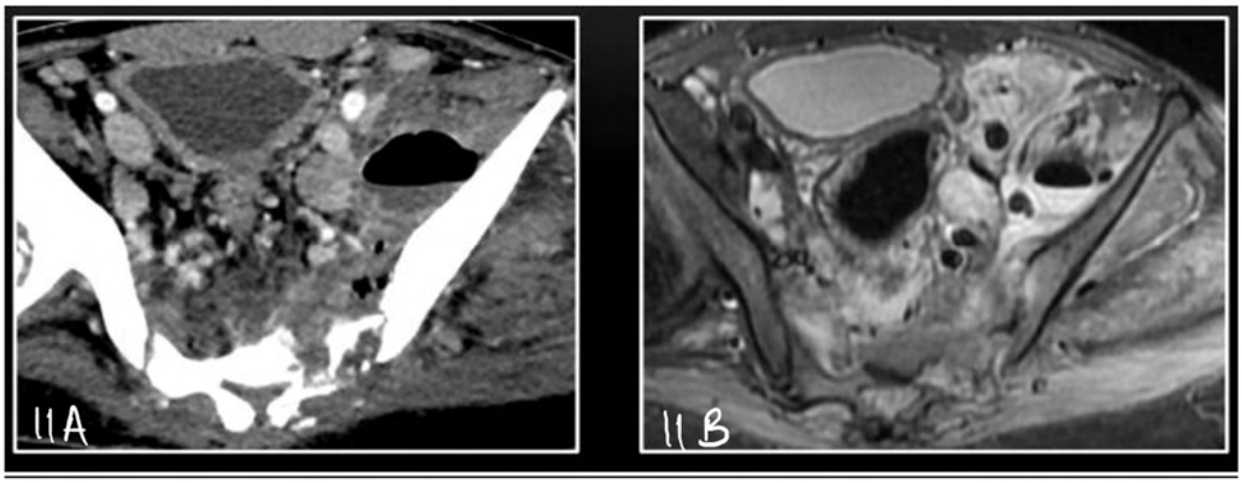

Fig. 11: Contrast enhanced CT(Fig. 11A) and MR(Fig. 11B) images show fluid collection with air fluid level in left iliacus muscle with inflammatory erosion and destruction of left sacral ala; Sacral osteomyelitis with abscess in left iliacus muscle.

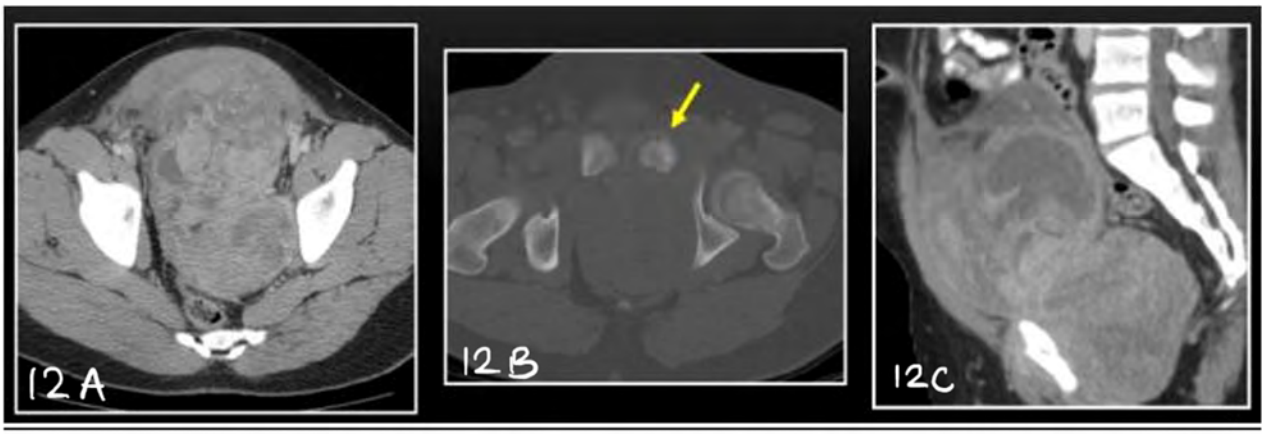

Fig. 12: Contrast enhanced CT images show a large heterogeneous pelvic extraperitoneal mass with involvement of left pubic bone(Fig. 12B); Soft tissue sarcoma.

MESENCHYMAL MASSES:
DESMOID FIBROMA

Desmoids are benign, non-inflammatory fibroblastic tumours with a tendency to invade locally and recur but they do not metastasize. There are associated with Gardner syndrome, Familial polyposis coli syndrome. On CT most desmoids are homogenous well circumscribed hyperattenuating and enhancing masses. MR shows low signal intensity on T1/T2 with variable enhancement. Watchful waiting in asymptomatic patients can be done

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SARCOMA

The retroperitoneum is a common location for soft-tissue sarcomas. Certain types of sarcomas may demonstrate calcification, including malignant fibrous histiocytoma (or undifferentiated pleomorphic sarcoma), synovial sarcoma (up to 30% of cases), dedifferentiated liposarcoma, extraskeletal osteosarcoma, and other spindle cell sarcomas. When a large heterogeneous pelvic retroperitoneal mass with local invasion is encountered, Sarcoma should be considered a likely diagnosis.

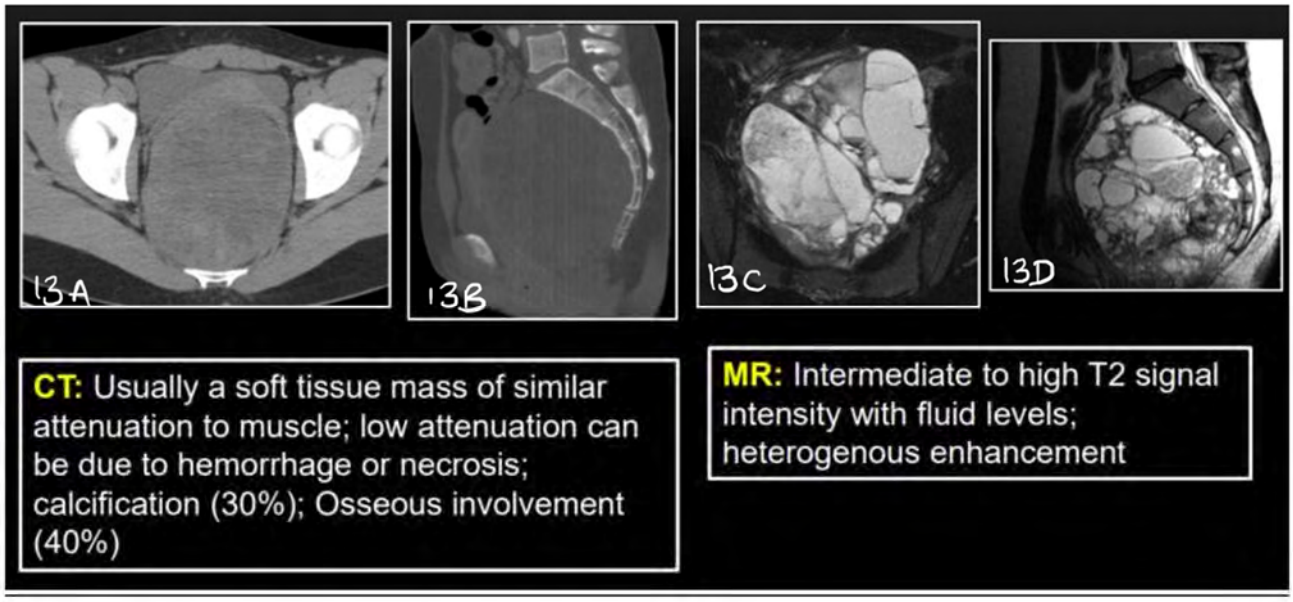

Fig 13: CT(Fig. 13A,13B) and MR (Fig.13C,13D) images show extraskeletal Ewing's sarcoma

EWING'S SARCOMA

Extraskeletal Ewing's sarcoma is included in the Ewing's sarcoma family of tumors and is 15 to 20% as prevalent as osseous Ewing's sarcoma.[7] It presents as a rapidly growing soft tissue mass which is usually 5-10 cm in size at presentation. Retroperitoneal involvement is seen in 10% of cases

RESULTS

The various pathological causes are encountered while reporting the retroperitoneal masses in children. The most common cause being congenital and developmental followed by neurogenic and vascular causes

CONCLUSIONS

The pelvic retroperitoneal space is a complex anatomic region that may be affected by a wide variety of masses in infants and children. CT and MR imaging play a key role in the evaluation of primary lesions that occur in this region. Congenital and developmental masses were the most commonly encountered pelvic retroperitoneal masses in children. Knowledge of the normal anatomy and familiarity with the imaging features and clinical manifestations of these lesions are important for determining the type of mass or narrowing the differential diagnosis, and also for defining the extent of the mass, an especially important surgical consideration

REFERENCES

1. Kocaoglu et al, Pediatric presacral masses; RadioGraphics 2006; 26:833–857

2. Tirkes et al, Peritoneal and Retroperitoneal Anatomy and Its Relevance for Cross-Sectional Imaging. RadioGraphics 2012; 32:437–451

3. Shanbhogue et al, Uncommon Primary Pelvic Retroperitoneal Masses in Adults: A Pattern-based Imaging Approach. RadioGraphics 2012; 32:795–817.

4. Al-Dasuqi K et al. Radiologic-Pathologic Correlation of Primary Retroperitoneal Neoplasms. RadioGraphics. 2020 Oct;40(6):1631-57.

5. Ros PR, Eshaghi N. Plexiform neurofibroma of the pelvis: CT and MRI findings. Magnetic resonance imaging. 1991 Jan 1;9(3):463-5.

6. Fortman BJ, Kuszyk BS, Urban BA, Fishman EK. Neurofibromatosis type 1: a diagnostic mimicker at CT. Radiographics. 2001 May;21(3):601-12.

7. Osman S et al. A comprehensive review of the retroperitoneal anatomy, neoplasms, and pattern of disease spread. Current problems in diagnostic radiology. 2013 Sep 1;42(5):191-208.

8. Nishino M, Hayakawa K, Minami M, Yamamoto A, Ueda H, Takasu K. Primary retroperitoneal neoplasms: CT and MR imaging findings with anatomic and pathologic diagnostic clues. Radiographics. 2003 Jan;23(1):45-57.

9. Yang DM, Jung DH, Kim H, Kang JH, Kim SH, Kim JH, Hwang HY. Retroperitoneal cystic masses: CT, clinical, and pathologic findings and literature review. Radiographics. 2004 Sep;24(5):1353-65.

10. Xu Y, Wang J, Peng Y, Zeng J. CT characteristics of primary retroperitoneal neoplasms in children. European journal of radiology. 2010 Sep 1;75(3):321-8.

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