

MATURE RETROVESICAL TERATOMA IN AN ADULT MALE: A CASE REPORT AND LITERATURE REVIEW

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ABSTRACT

Objective: We reported a case of mature (benign) cystic retrovesical teratoma. Retrovesical teratoma is a very rare extragonadal tumour, especially in adults, moreover in male patients. Grading and classification of teratoma is important for management and prognosis. **Material & method:** The patient in this study was a 49-year-old male who had chief complaints of abdominal mass and difficulty in urination. We performed Ultrasonography (USG) and Abdominal Multislice Computed Tomography (MSCT) scan on the patient. **Results:** USG showed a large mass in the pelvic region demonstrating a well-defined hypoechoic mass with septations at posterior of vesica urinaria. MSCT scan showed a large inhomogeneous hypodense mass with thin septations as well as multiple areas of fatty collections and coarse calcifications in pelvic region. We performed complete surgical resection per laparotomy. Grossly, the mass measured 12 x 10 x 5 cm and had rubbery consistency. Cut section of the mass revealed multilocular cystic spaces, whitish-gray walls, scattered yellowish adipose tissue collection, mucus secretions, and areas of calcifications. Pathological diagnosis of the resected tumour was a matured teratoma. The diagnosis was made because the tumour showed signs of a mature teratoma such as lined by stratified squamous and respiratory columnar epithelium, fat and muscle tissue, nerve tissue, and calcifications. **Conclusion:** There were no neuroepithelium appearance. To our knowledge, this is the first retrovesical teratoma case being reported in Indonesia.

Keywords: Teratoma, retrovesical, benign, cystic, adult.

ABSTRAK

Tujuan: Kami melaporkan sebuah kasus teratoma retrovesika matur (jinak). Teratoma retrovesika adalah suatu tumor ekstra gonad yang sangat langka, terutama pada dewasa, apalagi pada laki-laki. Klasifikasi dan penentuan derajat teratoma penting untuk manajemen dan prognosis. **Bahan & cara:** Pada penelitian ini pasien adalah laki-laki usia 49 tahun yang memiliki keluhan utama benjolan di perut dan kesulitan dalam berkemih. Pada pasien kami lakukan Ultrasonography (USG) dan Abdominal Multislice Computed Tomography (MSCT) scan. **Hasil:** USG menunjukkan adanya massa besar di daerah pelvis yang menggambarkan massa hipoekhoik dengan batas tegas dengan septa pada posterior vesika urinaria. CT scan abdomen menunjukkan adanya massa hipodens di homogeni dengan septa tipis disertai koleksi lemak dan kalsifikasi di daerah pelvis. Kami melakukan reseksi komplis per laparotomi. Secara kasar, massa berukuran 12 x 10 x 5 cm dengan konsistensi kenyal. Potongan massa menunjukkan daerah kistik multilokular, dengan dinding putih keabuan, jaringan lemak kuning, sekresi mucus, dan area kalsifikasi. Diagnosis patologi tumor yang diangkat menunjukkan tanda teratoma matur seperti lapisan skuamous bertingkat dan epitel kolumnar; jaringan lemak dan otot, jaringan saraf, dan kalsifikasi. **Simpulan:** Tidak ada penampakan neuroepithelium. Sepanjang pengetahuan kami, ini merupakan kasus teratoma retrovesika pertama yang dilaporkan di Indonesia.

Kata kunci: Teratoma, retrovesika, jinak, kistik, dewasa.

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INTRODUCTION

The Greek term teratoma literally translates to “monster tumor.” These tumors are germ cell in origin. Germ cell tumors originate from the cells of

an embryo's gonads.¹ They are rare tumours with a frequency of 1/40.000 birth.²

Teratomas are embryonal neoplasms that arise when totipotential germ cells escape the developmental control of primary organizers and

give rise to tumors containing tissue derived from all three blastodermic layers.² Primary mature teratomas are uncommon nonseminomatous germ cell tumors. They are made up of well differentiated parenchymal tissues that are derived from more than one of the three germ cell layers (ectoderm, mesoderm, and endoderm).³

Teratomas have been reported to occur in various sites and organs.² They usually occur in midline (paraxial) structures.³ Teratomas are located more often in the sacrococcygeal region and in the ovary, but they may be also found in many other anatomic regions.² The most common sites are gonads (testes and ovaries) followed by extragonadal sites such as intracranial, cervical, mediastinal, retroperitoneal, and sacrococcygeal regions.³⁻⁵

Teratomas are the second most common neoplasm in children following yolk sac tumour and occur with a relative frequency ranging from 13-19%.⁶ Mature cystic teratoma (MCT) is most common germ cell tumor both in gonads and at extra-gonadal sites (1-5% of all teratomas). They are seen in all age groups, predominantly in the third and fourth decades of life.⁷

The majority of cases are asymptomatic, present with nonspecific complaints, or identified incidentally on routine investigations. Surgical excision of mature (benign) teratoma is required for a definitive diagnosis (by histopathological examination) and remains the main stay of treatment. Prognosis is fortunately excellent after complete surgical excision with an overall five-year survival rate of nearly 100%.³

OBJECTIVE

We reported a case of mature (benign) cystic retrovesical teratoma. Retrovesical teratoma is a very rare extragonadal tumour, especially in adults, moreover in male patients. Grading and classification of teratoma is important for management and prognosis.

MATERIAL & METHOD

A 49 year old otherwise healthy male patient presented to Hasan Sadikin Hospital Bandung with a difficulty in urination for 2 years. Patient also complained about 4 years history of abdominal mass, growing bigger as time goes by. Physical examination was remarkable for a palpable, nontender, and limitedly mobile mass in pelvic region. Laboratory investigations were unremarkable, in which the Alpha Fetoprotein only accounts for 1.74, and human Chorionic Gonadotropin < 0.100.

We performed Ultrasonography (USG) and Abdominal Multislice Computed Tomography (MSCT) scan on the patient.

RESULTS

An Ultra Sonography (USG) was performed, showing a large mass in the pelvic region measuring $\pm 12.27 \times 9.11 \times 12.52$ cm and demonstrating a well defined hypoechoic mass with septations at posterior region of vesica urinaria, suggestive a teratoma.

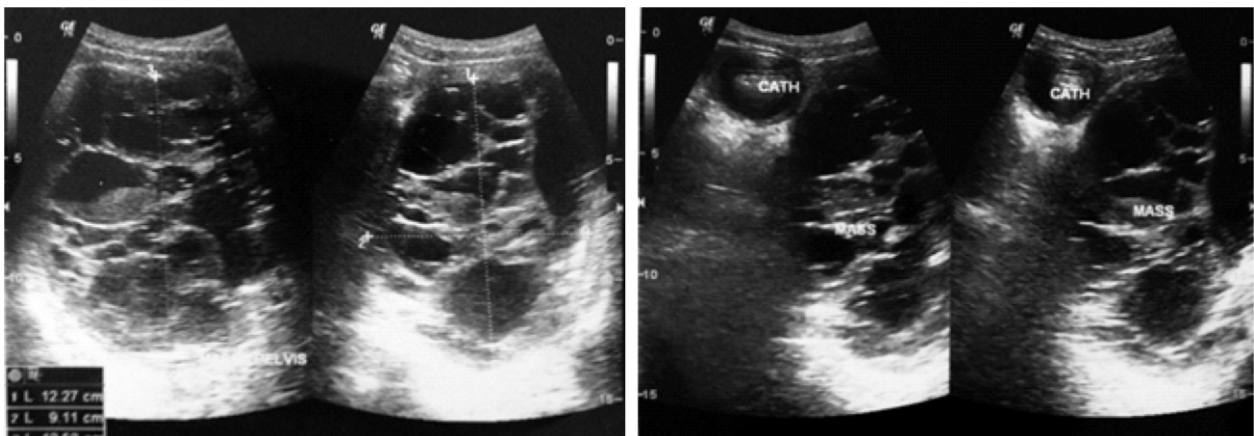


Figure 1. UltraSonography (USG). A large mass in the pelvic region measuring $\pm 12.27 \times 9.11 \times 12.52$ cm demonstrating a well defined hypoechoic mass with septations and at posterior region of vesica urinaria.

An abdominal contrast-enhanced computed tomography (CT) scan showed a large inhomogeneous hypodense lesion with multiple areas of fatty collections, well defined, irregular edge with thin septations and coarse calcifications measuring $\pm 13 \times 9 \times 9.6$ cm in pelvic region that displaced

the vesica urinaria anterolaterally, pushing the rectosigmoid colon and distal descendens colon posteriorly. Post contrast scanning demonstrating inhomogeneous enhancement especially within septations. From the Abdominal MSCT scan, the conclusion was a large inhomogeneous hypodense

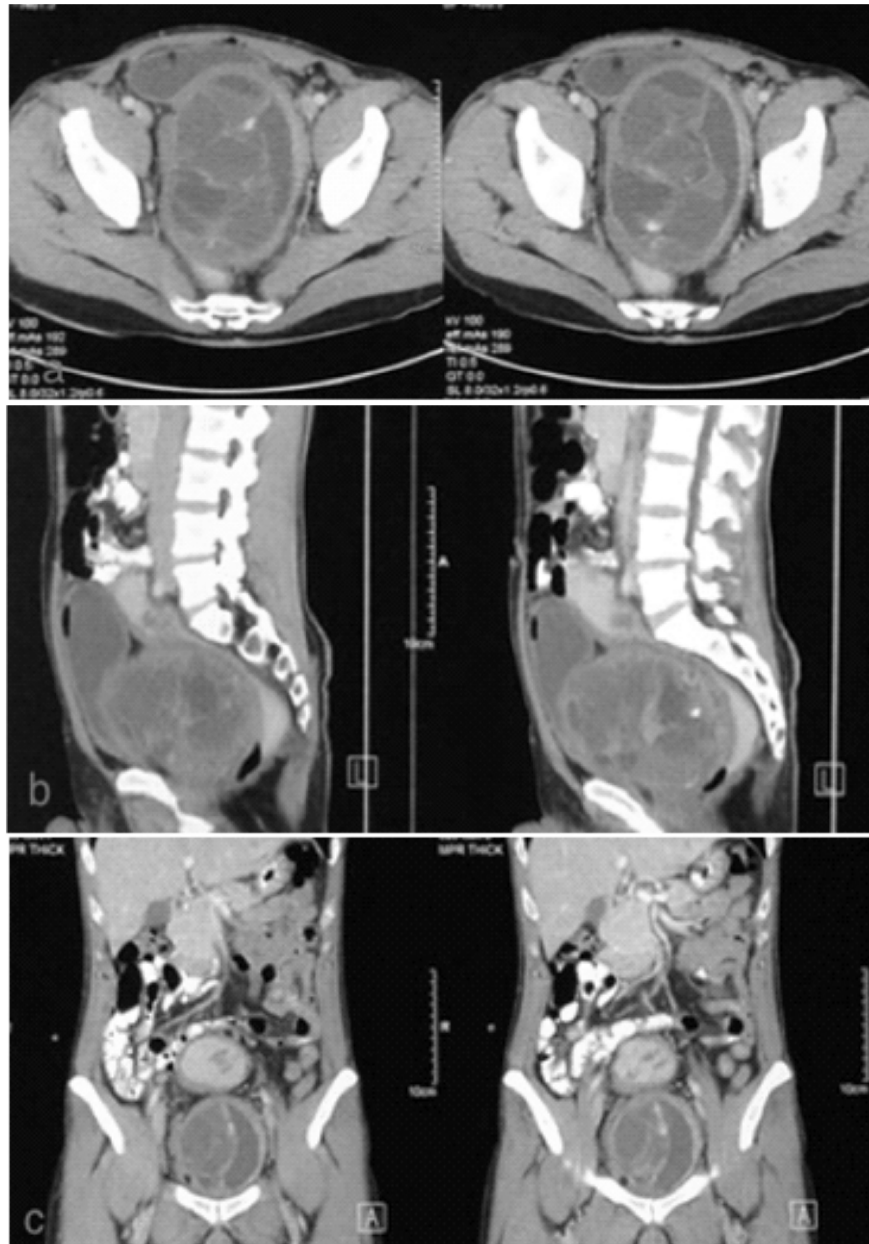


Figure 2. Abdominal contrast-enhanced computed tomography (CT) scan. (a) Axial (transverse) view: showing a large inhomogeneous hypodense lesion with multiple areas of fatty collections, well defined, irregular edge with thin septations and coarse calcifications measuring $\pm 13 \times 9 \times 9.6$ cm in pelvic region. (b) Sagittal view: showing the mass is located at posterior of vesica urinaria. (c) Coronal (frontal) view: showing displacement of vesica urinaria anterolaterally, pushing the rectosigmoid colon and distal descendens colon posteriorly.

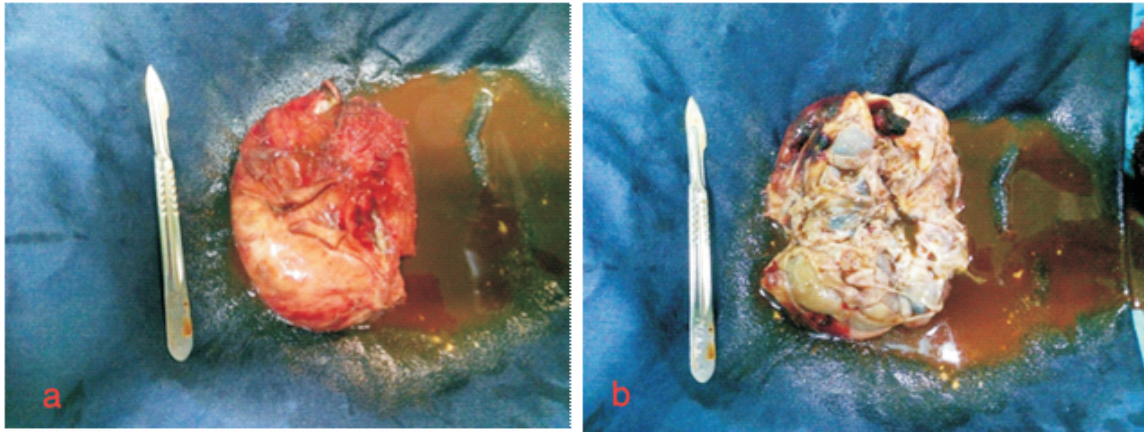


Figure 3. Mature cystic retrovesical teratoma. (a) Measured 12 x 10 x 5 cm with rubbery consistency. (b) Cut section of the mass revealed multilocular cystic spaces, whitish-gray walls, scattered yellowish adipose tissue collections, mucus secretions, and areas of calcifications.

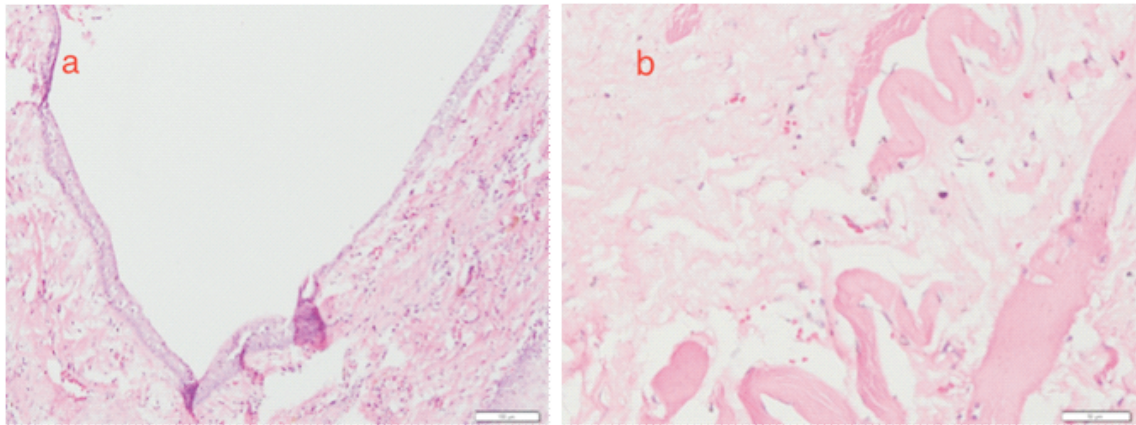


Figure 4. Histopathology examination. Hematoxylin & Eosin (H & E stain). (a) Scanning magnification view of the lesion. The lumen is lined by respiratory columnar epithelium. (b) Muscle and fat (adipose) tissues within the mass.

mass with thin septations as well as multiple areas of fatty collections and coarse calcifications in pelvic region, suggestive a teratoma.

The patient underwent complete surgical resection per laparotomy. Grossly, the mass measured 12 x 10 x 5 cm and had rubbery consistency (Figure 3a). Cut section of the mass revealed multilocular cystic spaces, whitish-gray walls, scattered yellowish adipose tissue collections, mucus secretions, and areas of calcifications (Figure 3b).

Microscopically, the walls were largely lined by respiratory columnar and squamous epithelium with various proportions of mature well-differentiated parenchymal tissue derived from the various three germ cell layers (Figure 4 (a) and 4 (b)). From the histopathology examination mature cystic

teratoma at regio pelvic conclusion was made. No evidence of malignancy was identified. A diagnosis of mature cystic retrovesical teratoma was made. There was no evidence of immature or malignant components. The patient was discharged in a stable condition. A post operative follow up showed no evidence of tumor recurrence.

DISCUSSION

Germ cell tumors (GCTs) can be broadly classified into two main categories, seminomatous and nonseminomatous GCTs. Teratomas belong to nonseminomatous GCTs and represent the most common form of all GCTs. Teratomas are encapsulated neoplasms composed of multiple parenchymal tissues (of varying degrees of

differentiation) that are derived from more than one germ cell layer (ectoderm, mesoderm, and endoderm).³ Ectodermal tissues most commonly include squamous epithelium and skin appendages. Endodermal tissues can be represented by respiratory or gastrointestinal epithelium, as well as "organoid" tissues such as pancreas, with mesoderm usually consisting of connective tissue and smooth muscle, and occasionally cartilage or bone.⁸

Teratomas are the second most common neoplasm in children following yolk sac tumour and occur with a relative frequency ranging from 13-19%.⁶ These tumors have been reported rarely in adults.⁸ Sacrococcygeal teratomas are the most frequent tumors in period neonatal, with an incidence of 1/25.000-40.000 and a female predominance (4 female/1 male).^{1,2,9} These tumors are 41-48% of teratomas in children (80% of these children have a congenital defect association). Discoveries in adult are rare.⁹ Incidence in the adult is probably unknown. Incidence in twins varies from approximately 10-50%.¹⁰ They are most commonly located in the sacrococcygeal region, followed by the ovaries, testis, anterior mediastinum, retroperitoneum, and finally the head and neck, which account for less than 5%.^{4,5}

Several theories of tumour pathogenesis have been raised in the literature, and Koen proposed that the pluripotent embryonic caudal mesenchyme gives rise to teratomas and other congenital tumours due to the dysfunction of several factors that probably involve gene function and cellular inductive interactions. The traditional view of tumour pathogenesis is that early in embryogenesis, primordial germ cells from the yolk sac become misplaced, most commonly into midline structures, after which they can give rise to germ cell tumours, including teratomas.¹¹ During embryogenesis, the endoderm layer produces the gastrointestinal tract, respiratory tract, and endocrine system; the mesoderm layer gives rise to the bones, cartilage, and tissues of the excretory system; the ectoderm layer produces structures such as skin, hair, nails, and the cells of the central nervous system.¹ In the 4th week of intrauterine fetal life, the germ cells migrate to the midline for development of gonads. During this migration, some of the cells may be retained in the tract and mature later in life to produce a teratoma. Thus, most of the extragonadal teratomas are usually seen in the midline, as in the anterior mediastinum, retroperitoneum and intracranial region.⁷ A widely acknowledged explanation

suggests that an SCT originates from the Henson node (also known as primitive knot), the embryonic precursor for the neural axis. Henson node cells reside within the embryonic distal spine, which is the same region where SCTs arise. An additional theory suggests that these tumors may actually be a result of abortive forms of twinning. Abortive forms of twinning include any twinning, either monozygotic or dizygotic, where 1 twin demises in utero. This assumption has been made because the incidence of twins appears higher than average in families of patients with teratomas.¹

Generally, teratomas arise from uncontrolled proliferation of pluripotent cells: germ cells and embryonal cells. The type of pluripotent cell greatly influences the presentation time and involved location of teratoma. Teratomas of germ cell sources can be congenital or acquired and are usually found in gonads (testes and ovaries). In contrast, teratomas of embryonic cell sources are always congenital and are usually found in extragonadal locations, such as intracranial, cervical, retroperitoneal, mediastinal, and sacrococcygeal sites.³

According to the location of tumor, teratomas can be classified into gonadal and extragonadal teratomas. Gonadal teratomas are more common, mostly primary neoplasms, mainly in adults, and usually take place in gonads (testes and ovaries). Conversely, extragonadal teratomas are less common, mostly secondary neoplasms, mainly in infants and young children, and usually take place in sacrococcygeal, mediastinal, retroperitoneal, and pineal gland sites (descending order of frequency).³ The majority of teratomas are located in the gonads and sacrococcygeal area.⁸ In a fetus or newborn, teratomas are most often located within the sacral region. This mass is referred to as a sacrococcygeal teratoma (SCT).¹

Grading and classification of both gonadal and extragonadal teratomas are important for the management and sometimes challenging for the anatomic pathologist. Teratomas are generally classified into four grades according to the Gonzalez-Crussi grading system. Grade (0): all component tissues appear well differentiated, grade (1): occasional microscopic foci contain incompletely differentiated tissues but not exceeding 10% of the sampled tissue, grade (2): Immature tissue composed of 10-50% of sampled tissue, and grade (3): more than 50% of sampled tissue are composed of undifferentiated tissue. Both grade (0)

and (1) are considered as mature teratoma and both grade (2) and (3) are considered immature teratoma (4).⁴

Furthermore, according to the content of tumor, teratomas can be classified into solid, cystic, or mixed teratomas. Solid teratomas lack organization and contain only parenchymal tissues. Cystic teratomas contain only sacs of fluid, semifluid, or fat, whereas mixed teratomas contain both solid and cystic components. Besides, according to the epithelial lining and dermal contents of tumor, teratomas can be classified into epidermoid, dermoid, and teratoid teratomas (cysts). Epidermoid teratomas are lined by stratified squamous epithelium and lack dermal contents. Dermoid teratomas are mostly lined by stratified squamous epithelium and contain various dermal contents such as hair, sweat, and sebaceous glands. Teratoid teratomas are mostly lined by respiratory columnar epithelium and contain sebum.³ In 1973 Miles and Stewart reported these tumors in adults. The following differences between the adult and infantile varieties were noted: 10 of the 11 tumors in this series were intrapelvic, whereas in the infant 90% are external. All adult tumors were cystic, while in infants they are bulky and contain solid as well as cystic components. In these adults, all tumors were benign and in the 24 collected cases only one was malignant. In infants the incidence of malignancy is much higher.¹⁰

In addition, according to the degree of tumor maturation, teratomas can be classified into mature and immature teratomas. Mature teratomas are generally benign, a symptomatic and more common, among females. They are highly variable on histology and can be solid, cystic, or mixed. They contain different types of parenchymal tissues that are well differentiated. Mature cystic teratomas (AKA dermoid cysts) may have partially to completely well-developed organ systems.³ Benign or mature teratomas are more likely to be cystic. Classically, a mature teratoma comprises tissue representing all three embryonic germ cell layers (endoderm, mesoderm, and ectoderm), with variation in the proportion of each type.⁸ On the contrary, immature teratomas are histologically solid teratomas and contain immature (undifferentiated/undeveloped) parenchymal tissues and can be possibly benign, possibly malignant, or frankly malignant. They are more common among males.³ The proportion of malignant teratomas ranges from approximately 7% in children to 26% in adults.⁵

Some mature (benign) and immature (possibly benign or possibly malignant) teratomas have an increased tendency to become frankly malignant teratomas, and frankly malignant teratomas have an increased propensity to metastasize. This group of exceptionally rare teratomas is known as teratomas with malignant transformation. The stratified squamous epithelial components of these teratomas are the ones at an increased risk of undergoing malignant transformations. In addition, teratomas with malignant transformation may produce components of somatic (non-germ cell) neoplasms such as carcinoma, sarcoma, and leukemia.³ Malignant transformation of benign cystic teratomas is thought to occur at a rate of less than 3%.⁵ Malignant transformation appears to increase in frequency with the time to diagnosis from birth. Only 7-10% of tumors diagnosed before age 2 months are malignant. However, after 2 months, the incidence of malignancy rises to 66% in boys and 50% in girls.¹² There is a sharp increase in the incidence of malignancy if the tumor is diagnosed after 2 months of age.⁸

Occasionally, a teratoma may contain various components of other germ cell tumor, and hence it is not a pure teratoma per se, but rather it is a mixed germ cell tumor and has malignant nature. In infants and young children, these components are frequently endodermal sinus tumor and chorioncarcinoma. A pure teratoma can be benign, however, highly aggressive in its clinical course as in a growing teratoma syndrome (GTS). GTS refers to a rapidly growing pure mature (benign) teratoma that appears during or following chemotherapeutic eradication of malignant components of a non seminomatous germ cell tumor, and it has normal serum tumor marker levels of alpha-fetoprotein and human chorionic gonadotropin.³

Teratomas can be diagnosed based on high index of clinical suspicion, routine laboratory, and radiographic investigations. Clinical presentations are variable and include nonspecific, abdominal/flank/back pain, obstructive gastro intestinal and genitourinary symptoms, as well as lower limb/genital swelling due to lymphatic obstruction. The majority of cases are asymptomatic, present with nonspecific complaints, or identified incidentally on routine investigations. They can rarely present with complications such as secondary infections (abscess formation), traumatic rupture leading to acute peritonitis, or malignant transformations. Midline (paraxial) teratoma masses, with

restricted mobility, can be easily detected on physical examination.³

With respect to laboratory investigations, retroperitoneal teratomas can express a diversity of serum tumor markers such as elevated alpha-fetoprotein (AFP), carcino embryonic antigen (CEA), and CA 19-9.³ Malignant teratomas may cause an increase in serum a-fetoprotein levels.⁵ These serum tumor markers are helpful in clinical practice and can be used to monitor successful treatment or detect relapse in patients with specific tumor marker-secreting teratomas.³

With respect to radiographic investigations, they play valuable roles in diagnosis of teratomas. Plain radiographs (X-ray) can identify calcified elements in 62% of cases whereas ultrasound (US) can greatly differentiate between cystic and solid elements.³ Although calcifications are present in 74% of benign teratomas, 15% of malignant cases also contain calcifications so their presence cannot be relied on to make the distinction either.⁵ Tooth-like calcifications are helpful in diagnosing teratomas but are rare. Sonography may show a predominantly cystic, solid, or complex mass with specular echoes and shadowing indicating calcifications, and occasionally a fat-fluid level. Ultrasound, however, does not provide reliable differentiation of fat from other soft tissues.⁵ Computed tomography (CT) scans can better distinguish between fat (adipose tissue) and bone (calcified) masses. On the contrary, magnetic resonance imaging (MRI) scans can offer better resolution of soft tissues, feasible identification of benign and malignant neoplastic features, and most importantly superior tumor staging assessment.³ Magnetic resonance imaging can distinguish fat fluid, calcium, and soft-tissue elements of teratomas and are superior to CT and ultrasound for showing anatomic relationships with adjacent structures but is less sensitive in detecting calcifications.⁵ MRI imaging characteristics of a teratoma are fairly specific and can assist in both timely and accurate diagnosis. In general, most teratomas will present as heterogeneously solid or cystic lesions with lipid components following fat signal intensity on all sequences. When present, other components such as calcifications, bone or teeth will show low-signal intensity on both T1 and T2 weighted images. CT imaging may also be of some utility in further demonstrating the presence of calcification or ossification, which may be present in up to 50% of malignant tumors.¹⁰

However, generally, a definitive diagnosis of teratoma demands a histopathological evaluation.³ Histopathologic examination of teratomas commonly reveals cystic and solid components containing hair, skin, cartilage, bone, sebum, fat calcifications, and nerve fibers, which have been found to correlate with both CT and magnetic resonance imaging findings.⁵

Teratomas are largely resistant to radio- and chemotherapy. Adjuvant radio- and chemotherapy are used only if malignant features of germ cell tumors are identified on histopathological examination. Surgical excision of benign (mature) teratoma is required for a definitive diagnosis (by histopathological examination) and remains the main stay of treatment. Prognosis is fortunately excellent after complete surgical excision with an overall five-year survival rate of nearly 100%.³ Although prognosis is excellent for surgical resection of benign teratomas, malignant teratomas recur and average survival is approximately 18 months.⁵

CONCLUSION

Extragenital germ cell tumors are a very interesting tumor entity with specific biological and clinical characteristics. In adults mixed GCTs with teratomatous elements are more common than pure teratoma. Mature cystic retrovesical teratoma is a rare entity in adults, especially in male. To our knowledge, only a few cases of retrovesical teratoma in adult have been reported. Although usually asymptomatic, large neoplasms can cause urinary disturbance. Preoperatively, the diagnosis can be established by its characteristic appearance on computed tomography. Distinction between mature and immature teratoma is made histologically. The definitive treatment for these neoplasms is surgical resection. Because of the risk of malignant change early removal of the tumor is indicated. The risk of recurrence is extremely low with radical resection. Post-operative complications that may be expected are bladder dysfunction, incontinence for faeces and dysesthesia, especially in the case of damage to pelvic splanchnic nerve.

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