Benign Neoplasms — Teratoma

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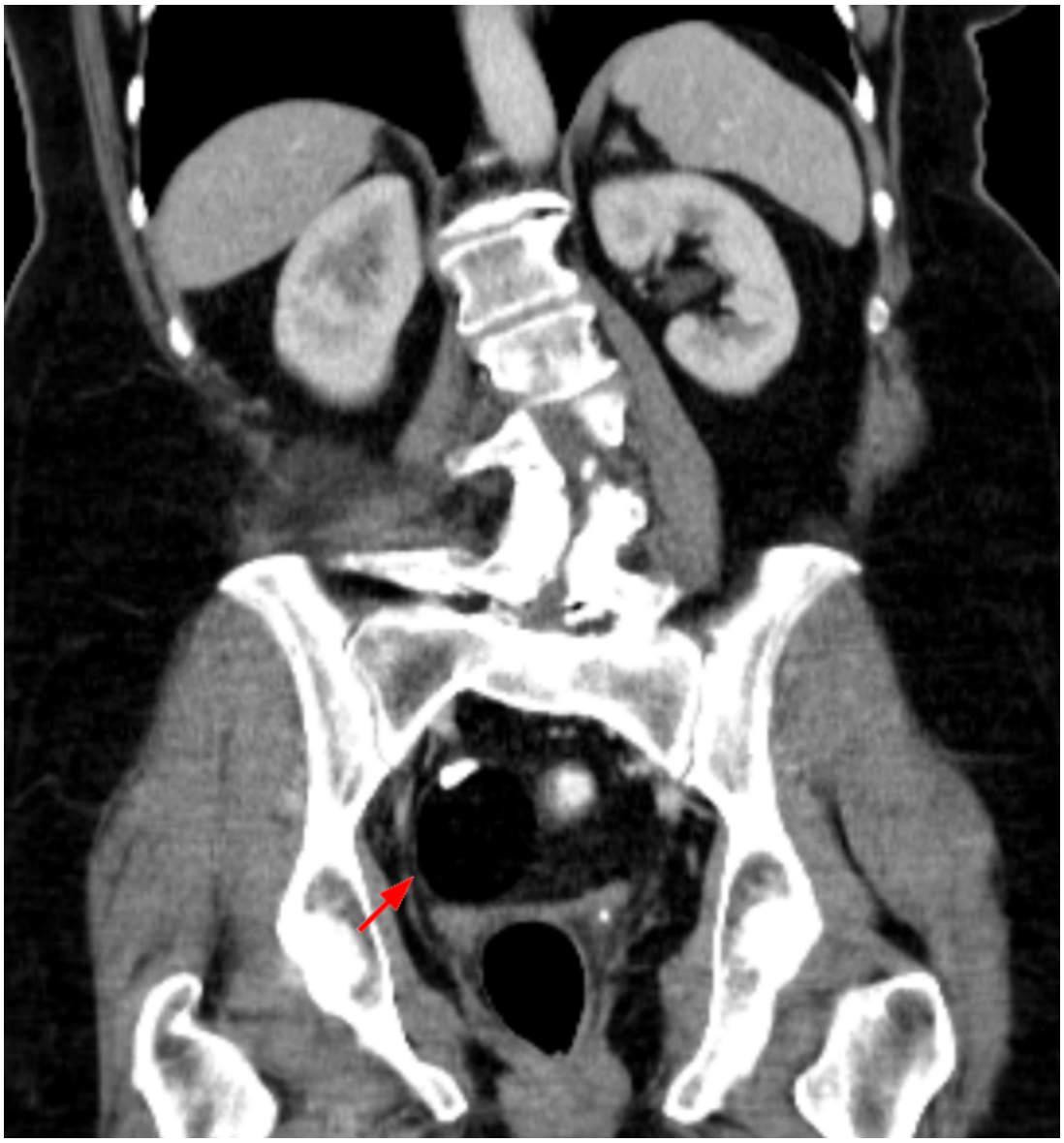
Teratomas are germ-cell tumors and are either mature (benign) or immature (malignant). They usually arise in the sacrococcygeal region, ovaries, testes, or mediastinum. Sacrococcygeal teratomas can be diagnosed antenatally, while teratomas in other locations are usually diagnosed postnatally. The treatment of teratomas is surgical. In most cases, complete excision of benign teratomas results in a cure, with minimal risk of recurrence.



Background

Teratomas are germ cell tumors that are composed of different cell types derived from one or more of the three germ cell lines. Cell types present may be ectodermal (e.g., skin, hair follicles), mesodermal (e.g., muscle, bone, teeth), or endodermal in origin (e.g., lung, gastrointestinal cells).

These tumors are broadly differentiated into benign, well-differentiated cystic lesions (mature) and malignant, poorly differentiated solid lesions (immature).

Epidemiology of Benign Cystic Teratoma

Sacrococcygeal teratomas are the most common neoplasm in newborns, with an estimated incidence of 1 per 20,000 births.

Up to 20% of ovarian tumors are mature cystic teratomas; however, pure testicular germ-cell teratomas are rare and account for only 5% of testicular tumors. Mediastinal benign teratomas are found in 8% of patients presenting with a mediastinal tumor.

The majority of teratomas occur in females (80%), and there is a clear gender discrepancy, especially for sacrococcygeal teratomas. While sacrococcygeal teratomas can be diagnosed in newborns, gonadal teratomas, such as ovarian cystic teratomas and testicular teratomas, are usually diagnosed in adolescents and adults.

Pathophysiology of Benign Cystic Teratoma

Teratomas are germ cell tumors arising from 1 or more of the 3 germ cell layers. These tumors arise from totipotent cells, which are capable of differentiating into a wide range of cell types and can form functional tissue.

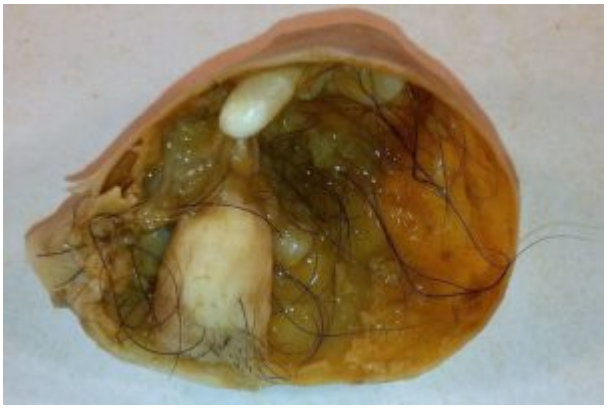


Image: “Incidental finding at the time of Caesarean section. The gynecologist was able to peel it off the ovary and leave the young woman with two presumably good ovaries.” by Ed Uthman from Houston, TX, USA – Mature Cystic Teratoma of the Ovary Uploaded by CFCF. License: CC BY 2.0

Totipotent cells are abundant in the gonads at the early stages of development, which is why teratomas are commonly diagnosed in this location. Totipotent cells are embryonic stem cells and can differentiate into any of the adult tissues.

Macroscopically, a benign cystic teratoma is often a cystic mass that contains hair, teeth, and skin mixed into thick, sticky, and often foul-smelling material, as seen in the figure.

The majority of these tumors are mature and well-differentiated into cystic teratomas. They are not expected to metastasize unless they occur in the testis.

Immature teratomas are usually diagnosed in postpubertal boys and are significantly more likely to metastasize and be malignant. Malignant transformation can also happen in benign cystic teratomas, especially ovarian ones.

Clinical Presentation of Benign Cystic Teratoma

The clinical presentation of teratomas depends on its location. Sacrococcygeal teratomas are usually diagnosed antenatally when the mother is diagnosed with excess amniotic fluid (polyhydramnios). If not diagnosed antenatally, they are usually diagnosed at the time of birth as a visible mass around the sacrum. Sometimes, these tumors are diagnosed later when the baby presents with asymmetric buttocks.

Ovarian teratomas are often asymptomatic and diagnosed incidentally. In most cases, the patient undergoes an imaging study or abdominal surgery for another indication, and an incidental teratoma is found in the ovary. Less commonly, ovarian teratomas may present with an abdominal mass. Rarely, ovarian teratomas can cause torsion or bleeding. These patients can present with abdominal pain, anemia, or an abdominal mass on physical examination.

Testicular teratomas present as diffuse enlargement of the testis that is painless. Discomfort can be a presenting symptom as well. When pain is present, testicular torsion should always be excluded.

Mediastinal teratomas are often asymptomatic or may present with shortness of breath and/or difficulty in swallowing due to compressive effects on the nearby structures.

Complications of Benign Cystic Teratoma

Sacrococcygeal teratomas carry a high morbidity and mortality risk due to antenatal and delivery-related complications. Patients with large teratomas diagnosed in utero can have an intrateratoma hemorrhage, which, if severe enough, can cause nonhemolytic hydrops fetalis.

Benign ovarian teratomas can become infected or rupture. Additionally, larger teratomas can cause ovarian torsion. Testicular teratomas, though rare, carry a high risk of metastasis even when they are mature.

Up to 2% of benign cystic teratomas can become malignant. The risk factors for malignant transformation include rapid growth, tumor diameter greater than 10 cm, and age over 45 years. Squamous cell carcinoma is the most common secondary neoplasm in these tumors, even though any component can undergo malignant transformation.

During surgery, a teratoma’s spillage can cause peritonitis or a foreign-body reaction and abdominal adhesions.

Diagnostic Work-up for Benign Cystic Teratoma

Laboratory investigations are not helpful in confirming the diagnosis of a teratoma but are beneficial in excluding associated complications such as malignancy transformation and anemia. Alpha-fetoprotein and beta-human chorionic gonadotropin are both reported to be higher in patients with malignant teratomas.

Imaging studies are the most important way to diagnose teratomas. In patients diagnosed antenatally, follow-up serial ultrasound studies are indicated to exclude associated complications, such as hydrops fetalis and polyhydramnios. Fetal magnetic resonance imaging (MRI) can be used to study the tumor's characteristics and invasiveness.

Patients who present with an abdominal mass benefit from ultrasonography and computerized tomography (CT) scanning. The ultrasonography demonstration of teratomas is the same regardless of their location and includes shadowing echoes of different densities, bright echoes, and fluid-fluid levels. Transvaginal ultrasound is the most sensitive and specific method to diagnose and differentiate ovarian cystic teratomas from other ovarian tumors.

Image: "CT of an Ovarian Teratoma" by Hellerhoff – Own Work.
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Abdominal CT scan is used to evaluate the liver and lymph nodes for possible metastasis. Abdominal CT scanning can be also valuable in the characterization of the mature teratoma. Several dividing septa, fat, and calcification can be seen on a CT scan of a teratoma.

Testicular teratomas should be evaluated by ultrasonography. The degree of maturity and invasiveness are important in deciding whether to spare the testis during surgery.

While mediastinal teratomas are rare, they can cause more serious complications due to their proximity to vital organs such as the heart, lungs, and major arteries. Chest CT and MRI scans are indicated to evaluate the tumor's extension and involvement of other vital organs, while echocardiography can be used to assess heart function in these patients.

If the imaging studies raise suspicion of malignancy, fine-needle aspiration or a core biopsy can be used to exclude possible malignant component. Histological examination of a benign cystic teratoma shows an inner lining of keratinized squamous epithelium, hair, bronchial and gastrointestinal cells, neuronal cells, retinal cells, or teeth.

Treatment of Benign Cystic Teratoma

In general, the treatment of benign cystic teratomas involves surgical excision of the tumor. Medical treatment is not indicated. Benign cystic teratomas do not recur if surgically resected.

Antenatally, sacrococcygeal teratomas can cause hydrops fetalis due to rich vascular flow

to the tumor. In these patients, fetoscopic laser ablation or vascular coiling are indicated to limit blood flow to the tumor. Additionally, if the antenatal evaluation reveals a large teratoma, cesarean delivery is indicated to avoid delivery complications.

If the sacrococcygeal teratoma does not cause any major complications antenatally, then tumor excision can be delayed until birth. Complete teratoma excision, along with excision of the coccyx, is indicated to avoid recurrence.

Benign cystic ovarian teratomas are removed by simple cystectomy rather than salpingo-oophorectomy, especially in young women who wish to avoid ovarian failure and infertility.

Testicular teratomas are usually excised along with the testis unless they are present in a prepubertal boy and are clearly benign.

Mediastinal teratomas should be excised completely along with any adherent structures to the tumor such as the lung or pericardium. Total excision of mediastinal teratomas is curative.

References

Cystic Teratoma via medscape.com

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Notes