Benign Neoplasms — Teratoma

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Teratomas are germ-cell tumors and can be 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. Complete excision of benign teratomas can virtually result in a cure with minimal risk of recurrence in the majority of the cases.
Background

Teratoma is a germ cell tumor, which is composed of different cell types derived from one or more of the three germ cell lines. The present cell types may be ectodermal (eg, skin, hair follicles), mesodermal (eg, muscle, bone, teeth), and endodermal origin (eg, 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 considered as 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 but pure testicular germ-cell teratomas are rare and account for only 5% of testicular tumors. Mediastinal benign teratomas are found in 8% of the 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 one or more of the three 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.

Totipotent cells are abundant in the gonads at the early stages of development, which explains why teratomas are commonly diagnosed in this location. Totipotent cells can be considered as embryonic stem cells and can differentiate into any of the adult tissue. Macroscopically, a benign cystic teratoma is often a cystic mass that contains hair, teeth, skin that is 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. Such mature teratomas are not expected to metastasize unless they occur in the testis. Immature teratomas are usually diagnosed in post-pubertal boys and are much more likely to metastasize and be malignant. Malignant transformation can happen in benign cystic teratomas, especially ovarian ones.

Clinical Presentation of Benign Cystic Teratoma

The clinical presentation of teratomas depends upon their location. Sacrococcygeal teratomas are usually diagnosed antenatally when mother presents with size greater than dates (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 is brought with the asymmetric buttocks.

Ovarian teratomas are often asymptomatic and are diagnosed incidentally. In the majority of the cases, the patient undergoes an imaging study or an abdominal surgery for another indication, only to find an incidental teratoma in the ovary. Less commonly, ovarian teratomas may present with an abdominal mass and rarely ovarian teratomas can cause torsion or bleeding and such patients can present with abdominal pain, anemia or an abdominal mass on physical examination.

Testicular teratomas present as a 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 intra-teratoma 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, even though rare, carry a high risk of metastasis even when they are mature.

Malignant transformation — 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. The 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 in the diagnosis of 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 to diagnose and differentiate ovarian cystic teratomas from other ovarian tumors.

Abdominal CT scan is used to evaluate the liver and lymph nodes status 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 appreciated on a CT scan of a teratoma.

Testicular teratomas should be evaluated by ultrasonography. The degree of maturity and invasiveness are important in making the decision of whether to spare the testis or not in the 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 are indicated to evaluate the tumor’s extension and involvement of other vital organs, while echocardiography can be used to assess the 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 is the surgical excision of the tumor. The medical treatment is not helpful. Benign cystic teratomas do not recur if surgically resected.

Antenatally, sacrococcygeal teratomas can cause hydrops fetalis due to a 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, caesarean 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 the coccyx, is indicated to avoid recurrence.

Bening cystic ovarian teratomas are removed by simple cystectomy rather than salpingo-oophorectomy especially in young women who have not completed their family to avoid ovarian failure and infertility in these women. 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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