

A Case of Childhood Ovarian Teratoma

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Abstract

Germ cell tumors are malignant (cancerous) or nonmalignant (benign, noncancerous) tumors that are comprised mostly of germ cells, the cells that develop in the embryo and become the cells that make up the reproductive system in male and female. Most ovarian tumors are of germ cell origin. A 5-year-old female child was taken to the Paediatrics department at Tawau General Hospital (TGH), Sabah, Malaysia on 10.4.2005 with gradual distension of abdomen which had been noticed by parents for the last 2 months. As bedside ultrasonography (USG) showed mixed echogenic mass in the lower abdomen, CT scan of abdomen was requested for further delineation. CT scan of abdomen done on 18.4.2005 showed a very large (about 12 × 9 cm) mixed density mass with homogenously enhancing well defined borders in the abdomen extending from the pelvic cavity to the upper abdomen causing significant pressure effect downwards towards urinary bladder and contrast material filled guts outwards and backwards mostly occupying the anterior part of abdominal cavity. The tumor was removed surgically and diagnosed as ovarian teratoma histopathologically.

Key words: Childhood, Ovarian, Teratoma

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Case Report

A 5-year-old female child was taken to the Paediatrics department at Tawau General Hospital (TGH), Sabah, Malaysia on 10.4.2005 with gradual distension of abdomen which had been noticed by parents for the last 2 months duration. As bedside ultrasonography (USG) showed mixed echogenic mass in the lower abdomen, CT scan of abdomen was requested for further delineation. CT scan of abdomen was done on 18.4.2005 which showed a very large (about 12 × 9 cm) mixed density mass, with well defined borders which extended from the pelvic cavity to the upper abdomen causing

significant pressure effect towards urinary bladder downwards and contrast material filled guts outwards and backwards mostly occupying the anterior part of abdominal cavity. No definite calcification could be depicted within the lesion. CT scan impression was huge solid mixed density mass in the abdominal cavity to rule out germ cell tumour or mesenteric sarcoma. However, laparotomy was done and the tumor was resected completely and sent for histopathology which confirmed the tumor as ovarian teratoma. The teratoma was benign in nature and there was no recurrence on one year follow-up.

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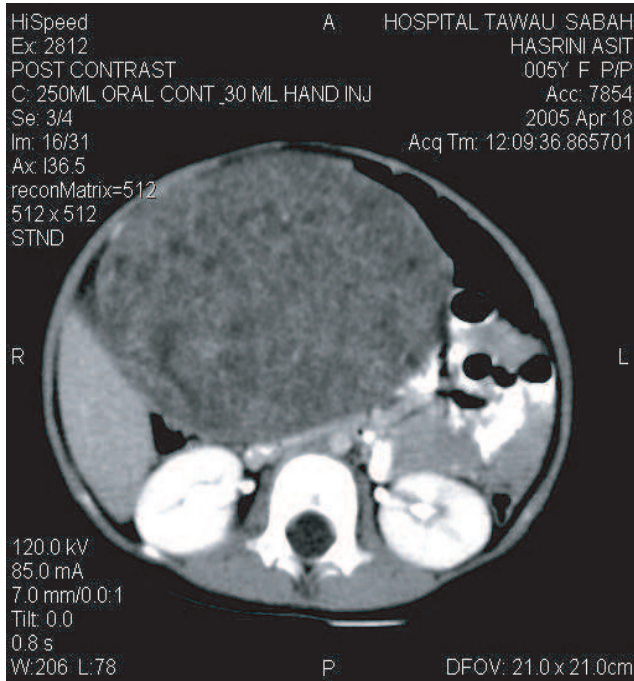


Fig 1. CT scan showing the ovarian teratoma

Discussion

Germ cell tumors are malignant (cancerous) or nonmalignant (benign, noncancerous) tumors that are comprised mostly of germ cells, the cells that develop in the embryo and become the cells that make up the reproductive system in male and female. These germ cells follow a midline path through the body after development and descend into the pelvis as ovarian cells or into the scrotal sac as testicular cells. Most ovarian and testicular tumors are of germ cell origin.¹⁻³

Tumors outside the gonad are called extragonadal tumors. These tumors also occur along the midline path and can be found in the head, chest, abdomen, pelvis and sacrococcygeal (lower back) area. Germ cell tumors represent about 3 percent of all childhood cancers. Germ cell tumors can spread (metastasize) to other parts of the body. The most common sites for metastasis are the lungs, liver, lymph nodes and the central nervous system. Rarely, germ cell tumors can spread to the bone, bone marrow and other organs.

Different types of germ cells

Diagnosis of germ cell tumors depends on the types of cells involved. The most common types of germ cell tumors include teratomas, germinomas, choriocarcinomas, embryonal carcinomas.

Teratomas

Teratomas contain cells from the three germ layers: ectoderm, mesoderm and endoderm. Teratomas can be malignant or benign, depending on the maturity and other types of cells involved. Sacrococcygeal (tailbone or distal end of spinal column) teratomas are the most common germ cell tumors found in childhood.

Teratomas derived from germ cells occur in the testes in males and ovaries in females. Teratomas derived from embryonal cells usually occur midline in the body: in the brain, elsewhere inside the skull, in the nose, in the tongue, under the tongue, and in the neck (cervical teratoma)⁴, mediastinum, retroperitoneum, and attached to the coccyx. However, teratomas may also occur elsewhere: very rarely in solid organs (most notably the heart and liver) and hollow organs (such as the stomach and bladder), and more commonly on the skull sutures. Embryonal teratomas most commonly occur in the sacrococcygeal region known as sacrococcygeal teratoma⁵⁻⁷ which is the single most common tumor found in newborn babies. Teratomas commonly are classified using the Gonzalez-Crussi⁸ grading system: 0 or mature (benign); 1 or immature, probably benign; 2 or immature, possibly malignant (cancerous); and 3 or frankly malignant. Teratomas are also classified by their content: a solid teratoma contains only tissues (perhaps including more complex structures); a cystic teratoma contains only pockets of fluid such as cerebrospinal fluid, sebum or fat; a mixed teratoma contains both solid and cystic parts. Cystic teratomas usually are grade 0 and, conversely, grade 0 teratomas usually are cystic.

Grade 0, 1 and 2 pure teratomas have the potential to become malignant (grade 3), and malignant pure teratomas have the potential to metastasize. These rare forms of teratomas with malignant transformation^{9,10} may contain elements of somatic (non germ cell) malignancy such as leukemia, carcinoma or sarcoma.

Initial diagnosis

Teratomas are thought to be present since birth, or even before birth, and therefore can be considered as congenital tumors. However, many teratomas are not diagnosed until childhood or adulthood. Large tumors are more likely to be diagnosed early. Sacrococcygeal

and cervical teratomas are often detected by prenatal ultrasound. Additional diagnostic methods may include prenatal MRI.

Beyond the newborn period, symptoms of a teratoma depend on its location and organ of origin. Ovarian teratomas often present with abdominal or pelvic pain, caused by torsion of the ovary or irritation of its ligaments.

Some teratomas contain yolk sac elements, which secrete alpha-fetoprotein (AFP). Detection of AFP may help to confirm the diagnosis and is often used as a marker for recurrence or treatment efficacy, but is rarely the method of initial diagnosis. (Maternal serum alpha-fetoprotein, or MSAFP, is a useful screening test for other fetal conditions, including Down syndrome, spina bifida and abdominal wall defects such as gastroschisis).

Time of presentation

Teratomas of germ cell origin usually are found in adult men and women, but they may also be found in children and infants. Teratomas of embryonal origin are most often found in babies at birth, in young children, and due to the advent of ultrasound imaging, in fetuses.

Treatment

Surgery

The treatment of choice is complete surgical removal (ie, complete resection).^{2,3} Teratomas are normally well encapsulated and non-invasive to surrounding tissues, so they can be relatively easily resected. Exceptions include teratomas in the brain, and very large, complex teratomas that have pushed into and become interlaced with adjacent muscles and other structures.

Chemotherapy

For malignant teratomas, usually, surgery is followed by chemotherapy. Teratomas that are in surgically inaccessible locations, or are very complex, or are likely to be malignant (due to late diagnosis and/or treatment) sometimes are treated first with chemotherapy.

Follow-up

Adequate follow-up includes close observation, involving repeated physical examination, scanning (ultrasound, MRI, or CT), and measurement of AFP and/or β hCG.^{11,12}

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