Scholars Journal of Applied Medical Sciences (SJAMS) **ISSN 2320-6691 (Online)**
Sch. J. App. Med. Sci., 2014; 2(2A):551-553 **ISSN 2347-954X (Print)**

(An International Publisher for Academic and Scientific Resources)
www.saspublisher.com

Review Article

Teratomas: the Unique masses: Embryological, Histopathological and Clinical Perspective

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Abstract: Teratomas are tumors that can be either benign or malignant. The clinical presentation of teratomas differs according to the site. This overview discusses on different types of teratomas like Mediastinal teratomas, Ovarian teratomas and on other sites.
Keywords: Teratoma, Ovary, Retro peritoneum, Germ disc

INTRODUCTION

Teratomas are unique types of tumors which can be either benign or malignant. They may contain components of germ layers either mesodermal or endodermal or ectodermal or mixed elements. They can be grossly confusing to a radiologist or a pathologist depending upon their constitution. The complexity of teratomas is a result of its components which can range from neural elements, connective tissue elements, epithelial elements, and rarer tissues like thyroid tissue. The location of teratomas is also wide and can present in any site like mediastinum, ovary, heart, liver, presacral region .They present unique challenges to embryologists, radiologists and clinicians.

DISCUSSION

The embryological basis of teratomas starts with the formation of germ layers like ectoderm, endoderm, and mesoderm. The first germ layer to be formed is the endoderm during the first week of followed by ectoderm and the last layer is the mesoderm. This process of gastrulation gets completed by three weeks when the germ disc is fully formed. The teratomas are embryological formed from any of the component or the mixed component. They usually contain components of two or three embryonic layers that may include teeth, skin, hair (ectodermal), cartilage and bone (mesodermal), or bronchial, intestinal, or pancreatic tissue (endodermal). The brain, respiratory and intestinal mucosa, cartilage, bone, skin, teeth, or hair may be seen in the neoplasm. The constituent tissues are not limited to those normally present in the area of origin and can be diverse. histologically they may range from well differentiated to poorly differentiated or malignant lesions [1, 2]. Arising from the totipotent cells they may be gonadal or extragonadal [3].

The clinical presentation of teratomas differs depending on the site.

The Thoracic teratomas usually present as an anterior mediastinal mass. They are not present in middle or posterior mediastinum. The Ovarian teratomas present as abdominopelvic masses, which can be silent or often with acute symptoms of torsion, bleeding, or rupture.

Retroperitoneal teratomas may present as a flank or abdominal mass.

Mediastinal teratomas

It has been observed that the Teratomas are the most common type of mediastinal germ cell tumors and accounting for 60 to 75% of such tumors. They are usually benign mature teratomas [4]. In the mediastinum they can cause compression of mediastinal structures and cause mediastinal syndrome in case of

rapid growth. Vital structures such as trachea, esophagus, thoracic duct can be compressed or eroded in case of invasion.

For mediastinal tumors especially the therapy for mature, benign teratomas is surgical resection, which confers an excellent prognosis.

Histologically any component of germ layers may be present [5]. In rare instances teratomas may undergo malignant transformation [6] and contain a focus of carcinoma. These malignant teratomas also known as teratocarcinomas are locally aggressive. Often diagnosed at anunresectable stage, they respond poorly to chemotherapy and in a limited manner to radiotherapy; the prognosis is uniformly poor.

Ovarian teratomas

The Teratoma is one of the most common ovarian tumor in children. They may be a completely silent condition or present as an emergency .The patient may simply present with a mass or abdominal pain due to torsion of the tumor. Benign teratomas of ovary present usually as cystic masses which can contain hair, thick sebaceous cheesy material.[7] Microscopically, usually skin elements dominate, including dermal appendages such as hair follicles and sebaceous glands. In most cases, however, structures of endodermal (respiratory and gastrointestinal epithelia) and mesodermal (muscle, fat, cartilage) origin are present. The rarer ovarian teratomas may also contain thyroid tissue and are then called as struma ovary. Grossly, immature teratomas are usually solid neoplasms with minimal cystic change. Microscopically, they are composed of immature (poorly differentiated) elements derived from all three germ layers. Primitive neuroectodermal (neuroblastic) elements are especially common. Mature teratomas usually contain well-differentiated tissues and are benign, whereas immature teratomas contain varying amounts of immature neuroepithelium or blastemal tissues. Immature teratomas can be graded from 1 to 3 based on the amount of immature neuroglial tissue present. Tumors of higher grade are more likely to have foci of yolk sac tumor. Malignant germ cell tumors usually contain frankly neoplastic tissues of germ cell [8].

Teratomas at other sites

Heart is an organ with lesser chances of malignancies. The Unusual benign lesions of the heart include fibromas, lipomas, angiomas, teratomas, and cysts. Lipomas have been rarelty reported in the heart. In addition Pericardial teratomas are also rare lesions [9] that can cause symptoms from compression of the right atrium with obstruction of venous return. Most of these occur in children and can be up to 10 cm in diameter.

Tumors occurring in the retrorectal space are rare. This specific anatomical region lies between the upper two thirds of the rectum and the sacrum above the rectosacral fascia. It is bound by the rectum anteriorly, the presacral fascia posteriorly, and the endopelvic fascia laterally. The retrorectal space contains multiple embryologic remnants derived from a variety of tissues (neuroectoderm, notochord, and hindgut). Tumors that develop in this space are often embryologically and developmentally heterogeneous. Congenital lesions are most common, comprising almost two thirds of retrorectal lesions. The remainders are classified as neurogenic, osseous, inflammatory, ormiscellaneous lesions. Malignancy is more common in the pediatric population than in adults, and solid lesions are more likely to be malignant than are cystic lesions.

Sacrococcygeal teratomas are the most common tumor in newborns **[**10] . Because cells from the primitive streak are pluripotent, the tumors contain various types of tissue derived from all three germ layers.They are three to four times more frequent in girls than in boys and usually presents as a large mass extending from the sacrum in the newborn period. The mass may be as small as a few centimeters in diameter or as massive as the size of the infant . The tumor has been classified based on the location and degree of intrapelvic extension. Lesions that grow predominantly into the presacral space often present later in childhood [11]. The differential diagnosis consists of neural tumors, lipoma, and myelomeningoceles. SCT are derived from all three germinal layers of germinal disc and contain neural elements, squamous and intestinal epithelium, skin appendages, teeth and, at times, calcium. Their inheritance may be sporadic but, occasionally, autosomal dominant. They are more common in girls (but are more often malignant in boys. About Fifteen percent have associated congenital anomalies like imperforate anus, sacral bone defects, duplication of uterus or vagina, spina bifida and meningomyelocele (scimitar sacrum, anorectal malformation and presacral mass forming Currarino's triad).

The radiological diagnosis is usually suspected when a calcified mass is noted in the pelvis on plain x-ray of the abdomen. CT scan can confirm this impression and may detect liver and retroperitoneal lymph node involvement in malignant cases. Operative excision, lymph node biopsy (both pelvic and retroperitoneal), peritoneal washings and biopsy, and omentectomy followed by combination chemotherapy result in a favorable outcome [12].

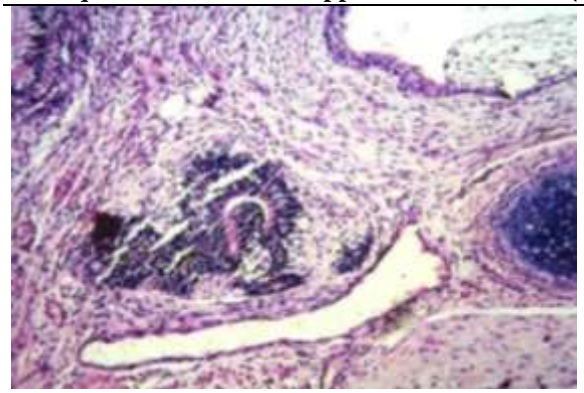

Fig. 1: A Teratoma containg immature elements in the form of Blood vessels, cartilage and glands with immature Neuroepithelial elements

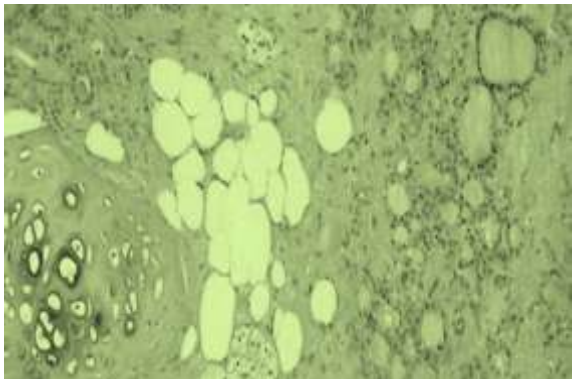

Fig. 2: A Teratoma containing fat, glands, thyroid follicles, cartilage cells in nests and sebaceous glands

CONCLUSION

Any clinician should suscpect the possibility of teratomas when a peculiar feature of a mass with different components is observed especially in mediastinum, ovary, presacral region or rarely heart, liver, pericardium, testis or brain.

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