Squamous Cell Carcinoma Arising from Primary Retroperitoneal Mature Teratoma

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Abstract
A 65 year old postmenopausal female presented with left sided abdominal pain. Sonogram revealed an intra-abdominal 7.4 X 5.7 cm heterogenous mass. On laparotomy, approximately 10 X 10 cm mesenteric mass was seen adherent to the descending colon. Multiple omental tumor deposits were also noted. Gross examination showed solid and cystic tumor with sebaceous material admixed with hair.

Histopathology showed mature cystic teratoma with a spectrum of well to poorly differentiated squamous cell carcinoma with omental metastasis.

INTRODUCTION
Teratomas are neoplasms with admixed elements from the three germ cell layers. They occur in the gonads and also in the extragonadal sites, particularly in the midline structures such as anterior mediastinum, retroperitoneum, sacrococcygeal region and pineal gland. Retroperitoneal teratomas represent only 1-11% of primary retroperitoneal tumors. Most of them are benign and only 1% of them undergo malignant transformation.

CLINICAL DETAILS
A 65 year old hypertensive, post menopausal woman presented with complaints of pain over the left side of the abdomen for the past 6 months that had increased over the last 2 weeks. She also complained of constipation, loss of appetite and loss of weight for the past 6 months. On examination, her general condition was found to be within normal limits. The abdomen was found to be distended with minimal free fluid but the flanks were free. On vaginal examination, the cervix was found to be healthy but a rigid mass was felt in the fornix. There was abdominal fullness of 14 weeks and the cervical movements were not transmitted to the mass.

Ultrasound revealed a heterogenous mass measuring 7.4 X 5.7 cm with solid and cystic areas on the left side (Fig. 1). The liver, bile duct, portal vein, gall bladder, pancreas, spleen, kidneys, urinary bladder, both the ovaries, fallopian tubes and uterus were within normal limits. The chest X ray and ECG were within normal limits. A guided fine needle aspiration cytology of the mass was performed and it showed clusters and scattered single malignant cells admixed with multinucleated osteoclast and foreign body type of giant cells. A cytological diagnosis of positive for malignant cells was rendered.

Operative Findings
Per operative findings was that of a large hard mass measuring 10 X 10 cm, arising from the mesentery and attached to the descending colon. Both the tubes and the ovaries were normal. A small fibroid was present on the posterior wall of the uterus. There was a deposit on the omentum, which was also resected and sent for histopathology.

Pathologic findings
The mass was irregular measuring 10 X 6 X 3 cm, grey white in color with focal areas of hemorrhage. The cut surface was predominantly cystic filled with pultaceous material with tufts of hair. There was an intra-cystic solid area, 3 X 1 cm. The histopathology of the mass showed a spectrum of well to poorly differentiated squamous cell carcinoma arising in a background of mature cystic teratoma with vascular invasion.

The omental mass was grossly solid in nature and microscopically showed a metastatic deposit of moderately differentiated squamous cell carcinoma.

DISCUSSION
Primary retroperitoneal teratomas account for 1-11% of retroperitoneal neoplasms and are most commonly found in neonates and young adults. They are the third most common retroperitoneal tumors in the pediatric population after neuroblastoma and Wilm’s tumor and account for 2-5% of all pediatric teratomas. However,
Retroperitoneal teratomas have been confused with ovarian and adrenal tumors such as adrenal myelolipomas as well as with a variety of renal and retroperitoneal masses including Wilm’s tumor, renal cysts, retroperitoneal fibromas, sarcomas, hemangiomas and enlarged lymph nodes. Macroscopically there are two variants, cystic and solid. Cystic teratomas are composed of fully mature elements and are benign. They contain sebaceous material and hair. Solid teratomas are most likely to be malignant and formed of immature embryonic tissue.

Malignant teratomas show either germ cell or non germ cell component. The former includes germinoma (seminoma, dysgerminoma), yolk sac tumor, embryonal carcinoma, choriocarcinoma or mixed germ cell tumors. The non-germinal component may include carcinoma, sarcoma, malignant embryonal tumors or again mixed tumors. Germ cell component worsens the prognosis but are radiosensitive (especially germinomas) and multimodality treatment often leads to effective control.

Tumors with extensive areas of immature somatic tissue require close patient follow up. Those with neural or rhabdomyosarcomatous differentiation have a poor prognosis. It has been reported that 1% of dermoids undergo malignant transformation, most commonly into squamous cell carcinoma. They are usually locally
advanced at presentation and have a more aggressive clinical course than teratomas as a whole. Malignant transformation of retroperitoneal teratoma is less common than in mediastinal teratomas. Mucinous adenocarcinomas have also been reported to arise in a primary retroperitoneal teratoma.\(^5\)\(^6\)

The most common chromosomal abnormality seen in teratoma is an isochromosome of the short arm of chromosome 12.\(^4\) An isochromosome is an abnormal chromosome with two identical arms due to loss of one arm and duplication of the other. For a diagnosis of primary retroperitoneal teratoma, plain antero-posterior and lateral radiographs, ultrasonography, computed tomography, MRI, arteriography and excretory urography are of use. The role of serum tumor makers is minimal. Retroperitoneal teratomas have been associated with occasionally elevated levels of CEA and Carbohydrate Antigen 19-9 (CA 19-9). This is not reproducible and hence clinically not very useful. The possibility of primary gonadal teratoma with retroperitoneal metastasis always should be ruled out before making the diagnosis of primary retroperitoneal teratoma. Surgical resection is paramount for both diagnosis and treatment. Prognosis depends on the tissue components within the teratoma and surgical resectability. Patients with complete resection of the benign teratomas have an excellent prognosis. Malignant teratomas do not fare as well. Malignant teratomas with germ cell elements as well as those with metastasis have a poor outcome. Chemotherapy and radiotherapy have relatively small roles in the management of these tumors.

We document this a case of metastatic squamous cell carcinoma arising from a retroperitoneal cystic teratoma for the rarity of its occurrence and the presentation in an elderly female. Teratomas with malignant transformation and metastasis have a very aggressive clinical course and poor prognosis.

**REFERENCES**