

Retroperitoneal Primary Yolk Sac Tumour in Adult Male

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ABSTRACT

Yolk sac tumour also called as endodermal sinus tumour is a malignant germ cell tumour. Extra gonadal germ cell tumours are very rare. The most common extra gonadal site is mediastinum. Retroperitoneum is the second most common site of occurrence. We report a case of a primary extragonadal yolk sac tumour in an adult male who presented with abdominal pain. Imaging showed a very large retroperitoneal mass. Histopathology of the biopsied specimen showed it as a Yolk sac tumour.

Keywords: Yolk sac tumour, retroperitoneum, malignant, germ cell.

Primary extragonadal germ cell tumours are very rare neoplasms. They account for approximately 1% to 2.5% of germ cell tumours [1]. They arise from aberrant primordial germ cell rests. There are no pathognomonic imaging findings however when there is a primary retroperitoneal mass with imaging features in favour of malignancy one should keep the possibility of extra-gonadal germ cell tumours in differentials.

CASE REPORT

A 49 year old male patient presented with dull aching type of diffuse abdominal pain and was referred to our department for ultrasound of the abdomen. USG abdomen showed a very large heteroechoic mass in the retroperitoneal region on right side extending into the posterior inferior segments of liver and into midline. The patient underwent contrast enhanced CT of the abdomen, which showed a large heterogeneously enhancing mass lesion in the retroperitoneum infiltrating the posterior segments of the liver and is extending into the midline and is displacing the right kidney inferiorly. IVC could not be delineated separately from the mass suggesting probable infiltration by the mass.

There were multiple round enhancing hypodense lesions in the liver suggestive of metastases. There was also mild intraperitoneal free fluid. USG of the scrotum showed normal sized testis with normal echotexture and there were no focal lesions. The patient underwent biopsy and the histopathology showed schiller duval bodies and immunohistochemistry was positive for AFP stain suggesting the diagnosis of yolk sac tumour.

DISCUSSION

Primary extragonadal germ cell tumours are rare neoplasms accounting for 1% to 2.5% of the germ cell tumours [1]. Extragonadal germ cell tumours arise from the aberrant primordial germ cell rests. Germ cells migrate through midline dorsal mesentery from fourth to sixth week during embryogenesis. Remnant tissue during migration can be the source of germ cell tumour [2]. On imaging non seminomatous germ cell tumours present as large heterogeneous masses containing hemorrhage and necrosis. Hemorrhage and necrosis are more common than in seminomas [3]. Germ cell tumours are divided into seminomatous and non seminomatous types which include embryonal carcinoma, yolk sac tumors, choriocarcinoma,

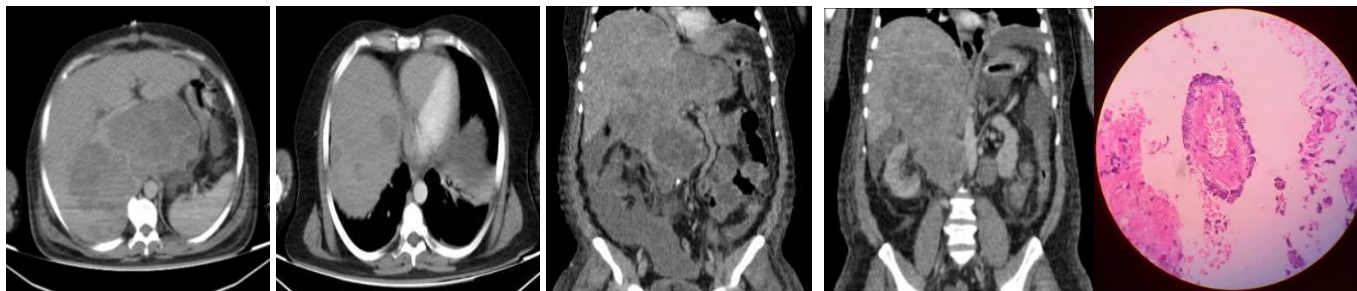


Figure 1-4: Axial and coronal contrast enhanced CT of abdomen showing Large heterogeneously enhancing retroperitoneal mass infiltrating the liver with multiple liver metastases. Figure 5: Histopathology slide showing Schiller Duval body

teratomas and mixed germ cell tumours [4]. The testicle is the most common site of germ cell tumours [5]. The prevalence of extra gonadal germ cell tumours is 1 to 2.5% of the germ cell tumours [3]. Extra gonadal germ cell tumours develop in or near the midline from primordial germ cell rests, that failed to migrate. Primordial germ cells travel from the yolk sac to the genital ridge. The genital ridge extends from the level of T6 to S2. The gonad stretches from diaphragm to the inguinal canal by the end of eighth week and then the gonad migrates to the pelvis or scrotum [5].

The failure of migration or aberrant migration of the primordial germ cells inhibits the apoptosis of ectopic primordial germ cells [3]. Germ cell tumours are also thought to arise from the physiologically distributed germ cells to the liver, brain and bone marrow [1]. As the overall incidence of germ cell tumours is higher in males, the prevalence of extragonadal germ cell tumours is also higher in males. The mean age for the discovery of extragonadal germ cell tumours in adults is 47 years. Mediastinum is the most common site of extragonadal germ cell tumours followed by retroperitoneum. The other common sites are pineal region, suprasellar region and sacrococcygeal region [4].

Gonadal germ cell tumours metastasizing to the retroperitoneum are much more common than the primary extragonadal germ cell tumours. So the diagnosis should only be made after excluding the primary tumour in the gonads. A midline mass favors the diagnosis of primary extra gonadal germ cell tumour over metastases [1]. Non seminomatous germ cell tumours appear as heterogeneous tumours with areas of hemorrhage, necrosis, and heterogeneous enhancement. And they commonly invade the adjacent structure [1]. Characteristic features of yolk sac tumour are elevated serum AFP. Primary

retroperitoneal Yolk sac tumours have poor prognosis. Chemotherapy with BEP (Bleomycin, etoposide, cisplatin) followed by surgical resection of the residual tumour is the treatment of choice [6].

CONCLUSION

Primary retroperitoneal Yolk sac tumours are extremely rare, if a large heterogeneous mass with necrosis and hemorrhage is seen in retroperitoneum one should include in differentials and testis should be screened for primary.

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