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Endodermal Sinus (Yolk Sac) Tumor of Broad Ligment Mesonephroma Adnexe Uteri

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Although inclusion cysts of the broad ligment are common, neoplasm arising at this site are rare. We have recently encountered a hitherto unreported variant of broad ligment neoplasm; called Yolk Sac Tumour. Even at other sites this highly malignant tumor of germ cell origin is rarely ever reported. A detailed review of literature is hereby presented with the case report.

CASE REPORT

A twelve year old girl was brought to the outpatient department by her worried mother when she noticed a mass in her abdomen. She was having pain of moderate intensity in the hypogastrium,radiating to her back. She also complained of constipation with passage of hard stools at times. The patient had lost about 2 kg weight within a week or so, and had fever for 2 weeks prior to her admission. She had a regular menstural history. Her LMP was 30 days prior to admission. On examination she looked pale. Her abdomen showed a bulge at left iliac fossa and hypogastrium. Palpitation reveald a well-defined huge mass arising from the pelvis filling the left iliac fossa extending upto the middle of left lumber region upto the midclavicular line. This mass was firm and slightly mobile. Pelvic examination was not carried out as girl the was unmarried and virgin. A provisional diagnosis of Retroperitoneal tumor (Fibrosarcoma), Ovarian/Utrine tumor was made. Pregnancy was ruled out by a negative pregnancy test. Ultrasound showed a very large mass, mostly solid, with septate cystic areas unrelated to abdominal viscera. Uterus appeared normal. Pre-operative impression was that of an ovarian tumor. Intravenous urography showed functioning kidneys. Both right and left ureters and the calyces were dilated. In addition the left ureter was pushed medially. Exploratory laparotomy showed a huge encapsulated mass arising from the left broad ligment. The mass weighed 1kg. Both ovaries and tubes were visibly normal (Fig-1). The tumor was attached to the omentum at mid transverse colon. A left salpingo-oophrectomy together with tumor was sent to the department of histopathology. As the tumor has ruptured a peritoneal lavage was also done. Histopathology revealed a normal left ovary and tube. The mass was reported as a yolk sac tumor (Fig-2). On the 6th post operative day, patient was hemodynamically stable, up and about and was on a regular diet. Chemotherapy was planned. The patient's father was explained about the malignancy of this tumor and was sent home with advice to followup in outpatient and to visit the onconlogist .She did not come for followup and or chemotherapy. Patient returned seven weeks later with complaints of intermittent attacks of high grade fever and pain in the legs especially the right leg with discomfort in epigastriaum and for the last two days a mass in the abdomen. She was pale & slightly pyrexic. A mass was palpable in right lumber and suprapubic regions which was fixed and firm in consistency. Patient had a low hemoglobin. Intravenous urogram showed dilation of both calyceal systems with kinking of right ureter and deviation of the left ureter. Ultrasound abdomen showed a large echogenic mass in the pelvis and abdomen. No liver metastases or enlarged glands were seen. CT. scan showed a large non-homogenous mass in the abdomen & pelvis, occupying almost the whole abdomen.Some elevation of right kidney was seen due to pressure of the mass. Massive ascites was present but no pelvic or paraaortic glands were visualized. Alpha feto proteins were raised five folds its normal value. Beta H.C.G. was not detectable. Second look surgery was done which showed the tumor invading the abdominal scar, filling the lower half of the abdomen, invading the small bowel, ascending colon & pushing descending colon forwards. It was very friable and hemorrhagic. About half a kilogram of tumor was debulked. Post-operative recovery was smooth and uneventful. Histopathology report was again of YST. Patient was given first dose of vincristine, actinomycin D

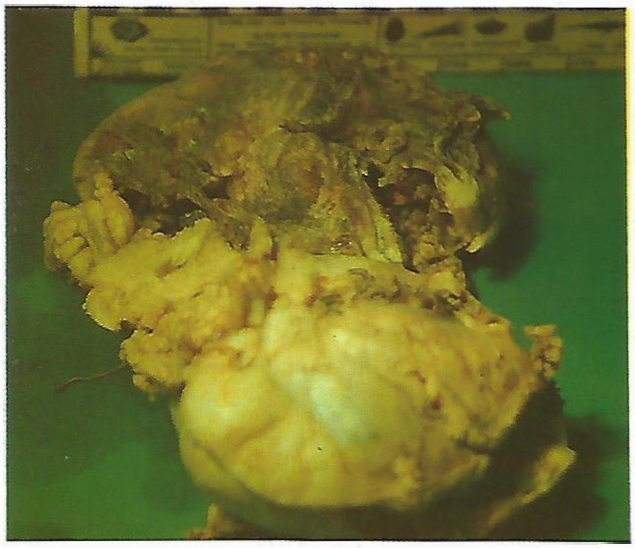

Fig.1: Macroscopic appearance of the tumor cut surface is mucoid, lobulated and cystic.

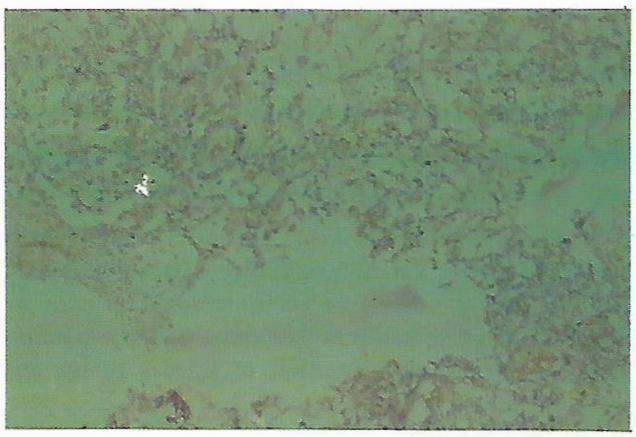

Fig.2: Microscopic appearance of endodermal sinus tumor: glandular spaces in a microcystic background. A schiller duvel body is also present.

and cyclophosphamide (VAC) combination chemotherapy on 6th post-operative day. Patient was discharged three days later. Second dose of chemotherapy was given a week later. Ascites had reduced by that time but patient was cyanosed. She died two days later.

DISCUSSION

This case is of a Yolk sac tumor arising from the broad ligament. It has never been reported at this site previously. Of the few tumors described in the broad ligment, most resemble epithelial neoplasms of the ovary including serous cystadenomata and carcinomata, Brenner cell tumors and endometroid type cyst adenocarcinoma. Five cases of leiomyosarcoma have also been reported at this site1[2]. German and French authors described several cases of sarcoma at this site. However, most of the authors did not adhere to the definition given by Gardener et al (1957), that "Tumor of the broad ligment may be defined on the basis of location as those which occur in or on the broad ligment but are not connected with either the uterus or the ovary[3]. The structure within the broad ligment most likely to give rise to neoplasms are: firstly remnants of the mesonephric (wolffian) system; secondly Ectopic paramesonephric (Mullerian) tissue trapped during formation of the fimbria of the fallopian tube; thirdly mesothelial inclusion cysts[4]. During the development of the embryo, primordial germ cells migrate from the yolk sac endoderm round the hind gut to the genital ridge in the retroperitoneum. The gonads develop in this position and subsequently descend to the scrotum or pelvis. Aberrant patterns of migration may lead to persistence of germ cells in sites such as the pineal body in the brain, mediastinum, bladder, liver or nasopharynx. Tumors of germ cell origin can therefore arise in any of these sites, as well as from ovary or testis. Neoplastic changes occurring in totipotent germ cell which have not under gone any differentiation will give rise to germinomas (commonly called dysgerminoma in ovary or seminoma in testis). Malignant extraembryonic embryonic differentiation to trophoblastic cells gives rise to chroicarcinoma and that to the yolk sac to yolk sac tumor[5].

First described by schiller in 1939 as tumors of Mesonephric origin yalk sac tumors were subsequently recognized by Telium (1971) as extraembryonic Membrane or endodermal sinus tumors (EST). A multiplicity of names have been applied to this neoplasm bearing testimony to the difficulty in ascribing a definitive origin of this tumor. It has been called a mesonephroma, angioreticuloma, mesoblastoma and "Embryonal carcinoma". In 1976 Kurman & Norris clarified this problem by describing 15 cases of embroynal carcinoma separating them histologically & clinically from EST. Telium coined, the term endodermal sinus tumor because of the morphological similarity of the neoplasm to the endodermal sinuses of the rodent placenta. These structures however do not occur in the human placenta. Thus the term, yolk sac tumor is more appropriate. Evidence exists to support the contention that the tumor is of yolk sac origin, for example alphafetoprotein (AFP) a product of normal human yolk sac is to be found in the serum of patients with yolk sac tumor and is also demonstrable in the

tumor itself. Embryonal and comparative anatomical evidence also lends weight to this argument[6,7,8].

Because description of YST is comparatively recent accurate incidence figures are difficult to obtain. Some writers now suggests that it occurs as frequently as the dysgerminomas. In all over 200 cases have been reported in the literature and accounted for 10% of all ovarian tumors. It is now regarded as the commonest highly malignant ovarian tumor to be seen in young girls; only rarely it may occur in women over 30 years of age. Kurman and Norris reported 71 patients with this tumor ranging in age from 14 months to 45 years. 60% were under 20 and the median age was 19 years. These figures are comparable with those of smaller series from M.D. Anderson hospital and the Emil Novak tumor registry[7]. 85% of all yolk sac tumor occur in ovaries or testis, 15% occur in extra-gonadal sites, most frequently in the anterior mediastinum and the retroperitoneal and sacrococcygeal regions. Less frequently yolk sac tumors are seen in the cerebrum cerebellum, pineal body of brain, eye, thymus, stomach, liver, cervix of uterus, vagina, clitoris, vulva, endometrium, urinary bladder and prostate[9].

The short duration of symptoms is the most striking feature of these tumors, Kurman and Norris found that symptoms had been present for less than 2 weeks in 66% cases and less than one week in 45% In 10% of patients the symptoms had been present for less than 24 hours. Abdominal pain is reported by 80% of patients and about 75% complain of abdominal mass. Non-specific abdominal distension occurs in about 30%. Many patients are febrile at diagnosis. Patient with acute abdomen are due to torsion or rupture of the tumor. Macroscopically yolk sac tumor are generally encapsulated, round or globular, grey in color, generally solid, although gelatinous cysts or areas of hemorrhage are not uncommon. Summarizing the findings of two major series, median diameter was 15-17 cm, weight was from 2-10 pounds. Localized disease was found in 50-70% of patients. Spread to contralateral ovary was found only if spread to other pelvic or abdominal viscera was present. In the AFIP series 135 ruptured before and 14% during surgery while in M.D. Anderson series the figures were 7 and 29 respectively 7. Yolk sac tumor may occur in various morphological guises or as a composite part of mixed germ cell tumor. In addition to characteristic schiller-Duval bodies and perivascular mantling, yolk sac tumor can show a variety of histological features. Morphological review of 50 cases has shown a reticular pattern in 98%, a solid pattern in 86%, the presence of hyaline globules in 84%, a festoon pattern in 54%, the presence of grannulomatous tissues reaction in 14% and a polyvesicular vitteline pattern in 8%[7,9]. Each suspected case of ovarian germ cell tumor should have alpha fetoprotein and beta fraction of human chronic gonadotrophin measured in the blood. Values of less than 15 units for alpha fetoprtein and 5 units for beta human chronic gonadotrophins are usually considered normal. Renal functions can be assessed by creatinin estimation. Hepatic function should be assessed. Ultrasound provides a rapid and easy assessment of response to treatment and may also diagnose liver metastases. C.T. Scan can help in staging and diagnosis of paraortic lymph nodes involvement and small liver metastases[10].

Studies have indicated no advantage for extensive radical surgery without chemotherapy to purely local excision with unilateral salpingo-oophrectomy. Biopsy of an apparently normal contralateral ovary is not justified for endodermal sinus tumors. Tumors masses should be debulked to the maximum possible extent. Radiotherapy is of no value. Probably the most successful chemotherapy regimens are PVB (cisplatin, vinblastin and bleomycin)[6]. Until recently the most widely used drugs were vincristine, actinomycin-D and Cyclophosphamide (VAC). As used originally, vincristine was given weekly, and severe side effects were not infrequent. But more recently studies show that these drugs should be given every 21 days or on monthly basis[11]. Without chemotherapy 30% patients survive after complete excision of stage I tumor and most die within a year of surgery. The addition of post-operative chemotherapy has increased the survival rate to 68% in stage I and 58% in more advanced disease. Results are less good when there is residual disease after surgery[6,7,10,12]. However, young age, limited surgery and curative chemotherapy have resulted in pregnancies and normal births in these patients[13,14,15]. Sessa et al have been shown eight long term survivors among 11 stage III patients, treated with PVB combination[11]. It is to be considered that great awareness of this neoplasm, together with earlier recognition, and the use of more efficacious combination chemotherapy will lead to better therapeutic results and improved prognosis in patients with yolk sac tumor.

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