ENDODERMAL SINUS TUMOUR OF THE OVARY

(Report of A Case)

by

NIRMAL GULATI,* M.S.

USHA VERMA,** M.D.

and

SUSHMA MADAN,*** M.D.

Endodermal sinus tumour of the ovary is a rare but highly malignant tumour seen in young women. Telium (1944) was the first one to recognise it as germ cell tumour and named it as endodermal sinus tumour. There is a multiplicity of names for this tumour: immature mesonephroma, yolk sac tumour, extraembryonic terratoma, embryonal carcinoma, extraembryonic mesoblastoma, mesoblastoma vitellinum and Telium's tumor. The microscopic characteristics diagnostic of this tumour are fairly straightforward, but the name and categorization are in dispute (Danforth *et al* 1978). It is a germ cell tumour with selective overgrowth of extraembryonic mesoderm and the yolk sac endodermal lining the sinusoid speces, thus mimicking endodermal sinuses of the rat placenta (Telium, 1965). Most American authors have recently agreed on the term endodermal sinus tumour. Two large series have been reported in the last two years. In 1976, Kurman and Norris reviewed 71 cases and in 1977, Jimerson and Woodruff reported an analysis of 41 cases from Novak Tumour Registry. In Kurman's series, 9 patients survived more than 2.2 years out of 65 followed up, whereas in Jimerson's review only 3 of 34 women followed up were alive for more than 3 years and 2 of these survivals had been treated with conservative surgery only, whereas the third one had chemotherapy in addition. This hopeless outlook calls for need for reporting individual experience with this rare and lethal tumour (Danforth, 1978). Following is a case report of a young nulliparous woman with endodermal sinus tumour seen at Medical College Hospital, Rohtak, (Haryana) who developed extensive metastases within 8 months of panhysterectomy and thio TEPA therapy.

*Reader in Obstetric and Gynaecology.

**Professor and Head, Department of Obstetrics & Gynaecology.

***Reader in Pathology.

Medical College Hospital, Rohtak (Haryana).

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CASE REPORT

Patient A. D., 20 years, married for 5 years, PO + O, was hospitalised for the first time on 2nd May, 1977 with history of mass in the lower abdomen for 7 months and amenorrhoea for 6 weeks. There were no other complaints in the form of pain, vomiting, loss of appetite or weight or urinary symptoms, neither had she noted any rapid increase in the size of the mass. She had menarche at 14, with regular menstrual cycle of 3-4/30 days.

On examination, she was in good condition but for slight pallor. Systemic examination: No abnormality. Abdominal examination revealed a firm mass 6 x 5½" arising from the

pelvis. Mass was mobile, non-tender and surface was smooth. There was no free fluid in peritoneal cavity. On pelvic examination: Cervix forwards, uterus retroverted nulliparous in size. Mass felt per abdomen was felt through anterior fornix. Per rectum examination: No nodules in the pouch of Donglas. Speculus examination revealed no abnormality.

Patient was given injection secrodyl and she had withdrawal bleeding and pregnancy was excluded. Provisional diagnosis of dermoid cyst was made and laparotomy was decided.

Investigations: Hb. 9.0 G%, TLC 8000/cumm. $P_{76}L_{20}E_2M_2$ ESR 3 mm in 1st hour. Stool: Cysts of E.histolytica present. Urine NAD. Xray chest—NAD. Blood sugar and urea were within normal limits. Blood group was 'A' Rh + ve.

At laparotomy, ovarian tumour 6" x 5½" was found to be arising from the left ovary and it was adherent to the dome of the bladder. Uterus, fallopian tubes and right ovary were normal. Ovariotomy was done after separating adhesions between tumour and the bladder serosa, which was subsequently repaired. Panhysterectomy was done and 400 mgms endoxan instilled in the peritoneal cavity. Postoperatively patient had urinary tract infection due to pseudomonas aerogenosa sensitive to gentamycin.

Patient was given injection Thio Tepa 30 mgms intravenously bi-weekly x 1 week and a total of 90 mgms was given in 3 weeks. Patient was discharged on 11th June 1977 with no evidence of secondaries in the pelvis and chest. She was advised to come for repeat cytotoxic therapy after 3 weeks.

Patient was lost to follow up for nearly 8 months, when she reported on 4th Feb., 1978 with secondaries in abdomen. She had a mass 8" x 7" in size, nodular firm and fixed which was arising from pelvis extending upto 3 fingers above umblicus. There was no ascites. No nodules in pouch of Douglas.

Investigations: Blood Hb. 7.0 G% TLC 5000/cum. DLC: P70, L22, M7, E1. X-ray chest: No secondaries. Blood urea 18 mgm and sugar 90 mg%. Urine NAD.

She was put on hematinics, analgesics and endoxan 500 mgm I/V daily after check on TLC. After 4 doses, patient became extremely sick. She had persistent severe pain not relieved by analgesics including injection pethidine. She developed jaundice, pernicious vomiting and fever and relatives took her home in moribund condition against advice on 13th Feb., 1978.

PATHOLOGY

Specimen: The ovarian tumour measured 14 x 10 x 10 cms. It was lobulated and well encapulated, except in one area where a capsular tear was identified. The cut surface had a variegated appearance, with grey white to pale yellow areas and large areas of haemorrhages and necrosis. Tiny cystic spaces filled with gelatinous material were also identified.

The hysterectomy specimen, along with one tube and ovary, showed no significant pathology.

Biopsy from the bladder wall comprised multiple pieces of grey tan friable tissue measuring together 2 x 1 x 0.6 cms.

Microscopic: The neoplasm revealed varying appearance in different areas of the tumour.

Some areas showed polyvesicular vitteline pattern with dense spindle cell stroma containing cysts of variable sizes which were lined by cuboidal or flattened cells. Other areas displayed reticular pattern consisting of a loose meshwork of spaces and channels lined by flattenor cuboidal cells with prominent nuclei (Fig. 1). Many areas of pseudopapillary or festoon pattern with characteristic Schiller-Duval bidies were seen. These bodies were identified as mantles of cells surrounding blood vessel. The cells were flattened or cuboidal, had well defined borders, light staining cytoplasm and prominent vesicular nuclei (Fig. 2). Intra and extracellular hyaline droplets were characteristically displayed in some of the areas particularly in relation to Schiller Duval bodies (Fig. 3). A few glands lined by mucuous columnar epithelium were also seen. Extensive areas of haemorrhages and necrosis were present. Calcification was not present. Uterus and contralateral ovary showed no significant abnormality.

Tissue from the bladder wall showed metastatic deposits of the same tumour.

Comment

Endodermal sinus tumour is an aggressive germ cell tumour seen in young women. Because of the multiplicity of the names of this tumour collection of information, comparison of results and ex-

perience of different authors is difficult. Endodermal sinus tumour is usually unilateral and treatment is salpingo-oophorectomy of the affected ovary, provided the tumour is encapsulated localised to one ovary and there are no metastases and the other ovary on bisection has no tumour. Uterus and the contralateral ovary are preserved since these individuals are young and results do not improve by radical procedure (Kurman and Norris 1976; Jimerson *et al*, 1977). In a series of 18 patients reported by Huntington and Bullock (1970) the only 2 survivors were the ones in whom conservative surgery had been done. Earlier-Santesson and Marrubini (1957) had encountered fatal results in 16 out of 17 patients treated by radical surgery and postoperative irradiation. In the present case, patient was young and nulliparous but panhysterectomy was done as initial frozen section report was cystadenocarcinoma and the tumour had spread to bladder serosa.

Most of the evidence suggests that endodermal sinus tumour is radioresistant (Smith and Rutledge 1975; Wharton, 1976; Jimerson, 1977) and radiotherapy does not improve salvage. Chemotherapy has been found to be useful in improving survival time in many of the reported cases (Aure *et al*, 1971; Smith *et al*, 1975; Forney *et al*, 1975; Kurman *et al*, 1976; Ettinger *et al*, 1977, Jimerson *et al*, 1977; Danforth, 1978). Smith and Rutledge (1975) reported on therapy of 48 patients seen between 1947-1974. Out of 20 patients given vincristine, Actinomycin-D, cyclophosphamide therapy (VAC), 15 survived from 3 to 78 months. Only one out of 8 patients who received actinomycin-D, 5-Fluorouracil and cyclophosharmide combination was well for 57 months after start of therapy.

In Kurman's (1976) series of 9 survivals out of 65 patients, 5 had surgery only and 3 had received VAC therapy after surgery, whereas 9th survival had spread of disease to omentum and panhysterectomy along with omentectomy was done and this was followed by intraperitoneal as well as I/V Thio TEPA therapy. Our patient had Thio TEPA therapy following panhysterectomy but developed extensive metastases within 9 months of diagnosis.

Conservative surgery followed by Vincristine, Actinomycin-D, Endoxan combination seems to be presently more effective for encapsulated unilateral endodermal sinus tumour as compared to radical surgery, radiotherapy or other chemotherapeutic agents (Smith *et al*, 1975; Kurman *et al*, 1976; Danforth, 1978).

Summay

A case of endodermal sinus tumour is reported in a young nulliparous woman of 20 in whom panhysterectomy was done followed by Thio TEPA therapy. Patient developed extensive metastases within 9 months of diagnosis. From literature, Vincristine, Actinomycin-D and Endoxan combination preceded by conservative surgery seems to give maximum survival time for this rare fatal tumour.

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See Figs. on Art Paper XIV-XV

Errata

August 1979 issue page 923

Article: **Clear Cell Carcinoma of the Endometrium.**

Name of 4th author

DR. (MRS) V. L. LAHIRI, M.D., M.A.M.S., M.I.A.C. has been inadvertently not printed as it should be.

Dr. Lahiri as fourth author of the above article.

Error is regretted.