ENDODERMAL SINUS TUMOUR

(A Review of 20 Cases)

by

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SUMMARY

There were 20 cases (2.01%) of Endodermal Sinus Tumours among 991 ovarian tumours or 8.9% of 224 germ cell tumours. The average age was 12.7 years. Lower abdominal mass (80%), acute abdominal pain (50%) and fever (30%) were the usual presenting features and rarely precocious puberty changes (10%). 85% of the tumours were unilateral, 35% had ruptured spontaneously and 10% had undergone torsion. 15% were of Stage I and the rest Stage III or IV. 60% were pure and the rest mixed with dysgerminoma, choriocarcinoma and/or teratoma. Surgery followed by chemotherapy was the treatment of choice. The prognosis of these tumours is bad in spite of advances in chemotherapy. The mortality rate was 90% and most of the patients were dead within a period of six months after the diagnosis and irrespective of the method of treatment.

Introduction

Endodermal sinus tumour is a highly malignant germ cell tumour usually occurring in young women and children. Pathologically it is characterised by a loose vacuolated network of mesoderm, peculiar perivascular structures similar to endodermal sinuses of rat placenta and intra and extracellular PAS positive hyaline globules of alphafetoprotein (Teilum, 1944, 1965; Czernobilsky, 1977. Following is a review of cases collected in the last 6 years from the Institute of Maternal & Child Health, Medical College, Calicut.

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Material and Methods

Patients were admitted and managed in the Departments of Surgical Paediatrics, Gynaecology and Radiotherapy, Medical College, Calicut, during a 6 year period, 1980 through 1985. The histopathological examination was done in the Department of Pathology.

Results and Discussion

Incidence

There were 20 endodermal sinus tumours among 991 ovarian tumours giving an incidence of 2.01%.

Clinical features

Age

The age of patients ranged from 6 months to 30 years with an average of 12.7 years. 85% were less than 20 years old and the rest between 20 to 30 years. This was similar to the reports of other authors. Neubecker and Breen (1962) reported an average of 20.3 years; Huntington and Bullock (1970) 15.9 years; Smith and Rutledge (1975) 19.2 years; Kurman and Norris (1976) 18 years.

Menstrual and Obstetric History

Fourteen, 70% of patients were premenarchal, 2 of them showed signs of precocious puberty such as thelarche and pubarche, indicating possible functional nature of these tumours. The rest were postmenarchal with regular menstruation. Two of them had 3 full term normal deliveries each, the last child birth being 55 days and 2½ years ago. This shows that in the adult the tumour may not interfere with normal function of ovary.

Presentation

The commonest presenting features were mass per abdomen in 16 (80%), followed by acute abdominal pain in 10 (50%), fever in 6 (30%), vaginal bleeding in 3 (15%), emaciation in 2 (10%) and recurrent urinary retention in 1 (5%-. The duration ranged from one day to one month with an average of two weeks.

In 17, 85% the tumour was unilateral in 2, 10% bilateral. In 1 case the tumour was protruding through the vagina, only biopsy could be taken; and presumed to be ovarian but could be extragonadal. The tumour was ruptured in 7 (35%) and had undergone torsion in 2 (10%). The unilaterility of the tumour has been stressed by other authors (Smith and Rutledge, 1975; Huntington and Bullock, 1970; Danforth, 1978). The unilaterility is important from the management point of view in young girls for whom conservative surgery may be undertaken. Duncan and Young (1980) have reported a normal pregnancy 2 years following unilateral ovariotomy. But in 80%, the tumour was of advanced stage of Stage III (75%) or IV (5%) and only 3 (15%) it was of Stage I. This indicates its extreme nature of malignancy.

Histopathology

Endodermal sinus tumours accounted for 2.01% of ovarian tumours or 8.9% of germ cell tumours (Table I) and 60% were pure in type and the rest were mixed type; dysgerminoma was associated with 6 cases, teratoma with 3 and choriocarcinoma with 2 cases. Two patients who presented with precocious puberty changes had pure Endodermal Sinus Tumour.

Treatment

Surgery following by chemotherapy was the treatment of choice (65%) (Table II). The rest had surgery alone (15%), surgery and chemotherapy followed by deep X-ray therapy (DXT) (10%), biopsy followed by DXT (5%) or biopsy followed by chemotherapy and DXT (5%).

Unilateral or bilateral ovariotomy (45%), total hysterectomy with bilateral salpingo oopherectomy (25%) and debulking (20%) were the mode of surgery. Only biopsy could be done in 10% (Table III).

Cyclophosphamide alone (20%) or in combination with vincristine (35%), actinomycin-D and vincristine (10%), 5

TABLE I
Histopathology

Histopathology		No.	% of germ cell tumours (N = 224)	% of total tumours (N = 991)
Endodermal Sinus Tumour (EST)	12			
EST with dysgerminoma	4			
EST with dysgerminoma and teratoma	2	20	8.9	2.01
EST with teratoma, choriocarcinoma	1			
EST with choriocarcinoma	1			
Dysgerminoma		28	12.5	2.8
Dermoid cyst		163	72.7	16.4
Malignant teratoma	3			
Teratocarcinoma	1			
Malignant teratoma with dysgerminoma	2	7	3.1	0.7
Teratoma with struma ovarii	1			
Choriocarcinoma		2	0.9	0.2
Struma ovarii		3	1.3	0.3
Mixed germ cell tumour		1	0.4	0.1

TABLE II
Mode of Treatment

	Number	Percentage
Surgery	3	15
Surgery and Chemotherapy	13	65
Surgery and Chemotherapy and DXT	2	10
Biopsy and DXT	1	5
Biopsy, Chemotherapy and DXT	1	5

TABLE III
Mode of Surgery

	Number	Percentage
Unilateral ovariotomy	8	40
Bilateral ovariotomy	1	5
Total hysterectomy with bilateral salpingo oopherectomy	5	25
Debulking	4	20
Biopsy	2	10

fluorouracil (5 FU) (10%) or methotrexate and 5 FU (5%) were the chemotherapeutic agents (Aable IV) used.

TABLE IV
Chemotherapy

	Number	Percentage
Cyclophosphamide and vincristine	7	35
Cyclophosphamide	4	20
Vincristine, actinomycin-D and cyclophosphamide	2	10
5 FU and Cyclophosphamide	2	10
Methotrexate 5 UF and cyclophosphamide	1	5

For children vincristine was given 0.2 mg- 1 mg I.V. twice weekly, cyclophosphamide 150-175 mg daily I.V. for 5 days, actinomycin-D 0.2 mg daily I.V. for 5 days, repeated every 3 months for 2 years. For adults combination treatment, with cyclophosphamide 500 mg and 5 FU 500 mg I.V. weekly for 3 courses, was given.

Period of Survival

Endodermal Sinus Tumour is a very malignant one associated with very high

mortality. There are only 2 survivers among the 20 patients at the time of reporting, rest were dead within 2-6 months after diagnosis (Table V).

TABLE V

Period of Survival

Survival period	Number	Percentage
2-3 months	14	70
4 months	3	15
9 months (alive)	1	5
3 years and 11 months	1	5
(alive)	1	5

Among the survivers one is a girl who had undergone unilateral ovariotomy at the age of 8 years for pure EST and followed with chemotherapy using vincristine and cyclophosphamide for 2 years. Now she has completed 3 years and 11 months and is disease free. The other is a woman of 30 years who had undergone total hysterectomy with BSO for unilateral pure EST which had undergone torsion on its pedicle. She was followed with 5 FU and cyclophosphamide 500 mg weekly I.V. for 3 weeks. She has completed 9 months and is disease free.

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