

Endodermal sinus tumor of the ovary

by

V. NINFO * and M. CARLI **

INTRODUCTION

Embryonal cell carcinoma of the ovary accounts for 10% of ovarian tumors in adults, and for about 60% in females under 20 years of age (^{1,4,6}). The term « endodermal sinus tumor » was first applied by G. Teilum (⁸) to certain embryonal carcinomas which exhibited a histological pattern resembling the structures of extra embryonic membranes. These neoplasms are extremely rare, and only 31 cases of pure endodermal sinus tumor of the ovary have been reported, to this date (²).

CASE REPORT

A 5 and 1/2 year old female child was admitted to the Hospital of the University of Padua on July 20, 1973, presenting with low fever, anorexia, pollakiuria, dysuria and recurrent abdominal pain. She had lost 1,5 kg in the previous month. Physical examination revealed an endoabdominal mass. Descending pyelogram disclosed that the mass occupied the entire pelvis and pressed down on the bladder. The right ureter was enlarged while the left one was compressed. During explorative laparotomy a mass, 15 x 10 cm, was seen which seemed to originate from the right ovary, and involved the Douglas cavity, uterus, left ovary, rectum, sigma and left ureter. The mass was removed together with the uterus, ovaries and tubes, and since a radical excision was not possible, the rectum was left intact.

Histological examination of the neoplasm revealed a vacuolated network together with microcysts lined with mesothelial cells usually within a loose myxoid stroma (Fig. 1). Intermixed with these structures were many perivascular formations made up of a central capillary within a mesodermic agglomerate which appeared surrounded by a row of epithelial-like cylindrical cells (Fig. 2). In addition, there were many small intercommunicating cavities furnished with papillary processes (Fig. 3), and cysts lined by a single layer of flat cells with protruding nuclei (Fig. 4). Occasionally, agglomerates of non-differentiated embryonal cells were seen along the edges (Fig. 5). Many intra and extracellular hyaline, PAS-positive globules were seen (Fig. 6).

The diagnosis was pure endodermal sinus tumor of the ovary.

Twelve days after surgery, the patient underwent Co⁶⁰ radiation of the abdomen for a total of 4000 rads, plus 1000 more rads directed on a residual palpable mass (5 x 5 cm) 4 cm below the umbilicus. Upon termination of radiation, she received adriamycin (60mg/m²/24 hr) by i.v. injection which was repeated after an interval of 15 days. The child tolerated the adriamycin well, and was discharged after the 2nd cycle in good condition.

At the beginning of the third adriamycin cycle, however, on abdominal pal-

* From the Institute of Pathological Anatomy University of Padova.

** From the Pediatric Clinic University of Padova.

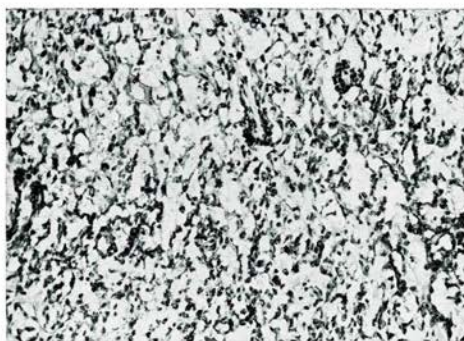


FIG. 1 - Vacuolated network made up of microcysts lined by «mesothelial» cells. H. E. $\times 120$.

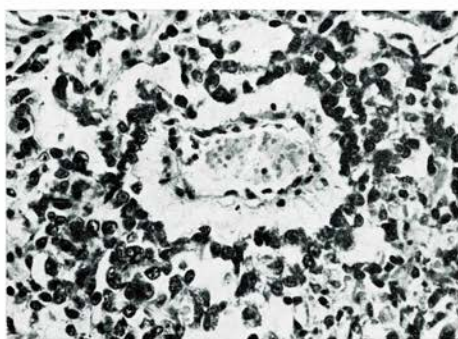


FIG. 2 - Perivascular formations with central capillary in a mesodermic agglomerate surrounded by epithelial-like cells. H. E. $\times 300$.

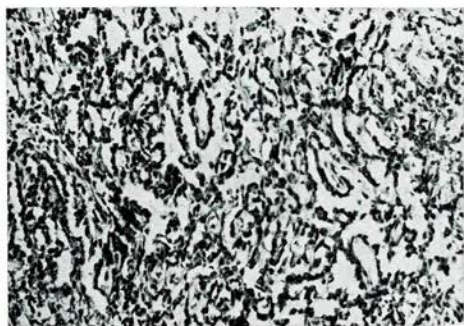


FIG. 3 - Papillary processes with small communicating cavities. H. E. $\times 120$.

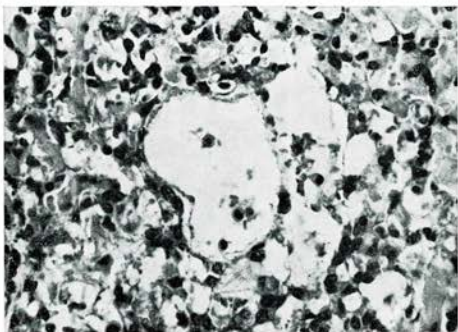


FIG. 4 - Cystic cavity lined by a single file of flat cells. H. E. $\times 300$.

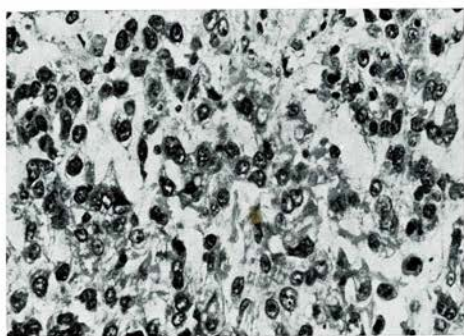


FIG. 5 - Agglomerate of undifferentiated embryonal cells. H. E. $\times 300$.

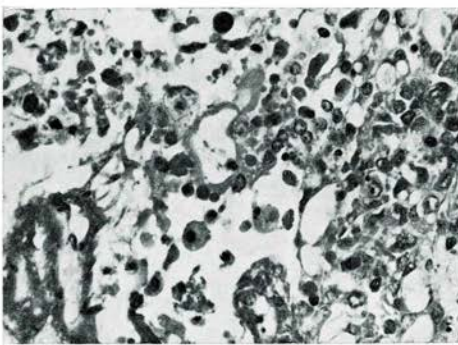


FIG. 6 - Intra- and extra-cellular, PAS-positive, hyalin globules. PAS $\times 300$.

pation a hard, cord-like mass was felt near the right iliac fossa, in addition to the paraumbelical mass which had remained unchanged. During laparotomy, the presence of hemorrhagic fluid in the peritoneum, and peritoneal and omental metastase were observed. The paraombelical mass was a purulent organization

whose excision required a small resection of the intestine. Numerous peritoneal nodules were removed. Histological examination disclosed that these were similar to the original tumor, but in some circumscribed zones, voluminous epithelial cells were observed assembled in syncytia, reminiscent of chorio-carcinomatosis. On Nov. 16, 1973, polychemotherapy was initiated, consisting of Actinomycin D, $350 \mu\text{g}/\text{m}^2 \times 5 \text{ d}$, iv; methotrexate, $7 \text{ mg}/\text{m}^2 \times 5 \text{ d}$, iv; endoxan, $150 \text{ mg}/\text{m}^2 \times 5 \text{ d}$, iv, which was repeated once a month.

The general condition of the child remained stationary until Jan 15, 1974 when she complained of abdominal pain and tenesmus, which presented with low fever. These symptoms subsided following administration of the polychemotherapy cycle. A month later, a voluminous mass was felt at the right iliac fossa; the abdomen was increased in volume, contracted and presented marked hypophonensis on percussion. Rectal exploration revealed that the Douglas cavity was occupied by a hard mass which was pushing the rectum posteriorly producing stenosis. X-ray showed a uniformly opaque abdomen, almost completely lacking in gas. A synchronous chemotherapeutic cycle was attempted employing Vincristine ($1 \text{ mg}/\text{m}^2$ iv) on day 1 and 2, and adriamycin ($10 \text{ mg}/\text{m}^2$ iv) on days 3, 4, 5, 6, and this brought about a subjective improvement. The child expired on March 17, 1974, with generalised carcinomatosis.

DISCUSSION

Despite the terminology adopted by Teilum, there is still some confusion regarding the classification of endodermal sinus tumors. This confusion is in part due to the neoplasm described by Schiller in 1939 as a mesonephroma of the ovary. Novak and Woodruff⁽⁵⁾ sustain that endodermal sinus tumors are a rare but specific entity, quite distinct from other embryonal carcinomas and Schiller's mesonephroma. Their judgement is based on the morphological features of the neoplasms, the poor prognosis, and the fact that it is only seen in subjects younger than 20 years of age. More recently, Huntington and Bullock⁽³⁾ confirmed these concepts and underlined the highly malignant behavior of this neoplasm.

The peculiar histological features of the endodermal sinus tumor are non-differentiation of the cells, and presence of structures analogous to those found in the earlier stages of embryonal development, representing the extracelomatic mesoblast and the endoderm of the vitelline sac. In agreement with Teilum⁽⁸⁾ we observed all the characteristic features of the endodermal sinus tumor which can be summarized in four basic groups:

a) cystic cavities lined externally by flat epithelia, and internally by cubical or cylindrical cells which surround the mesodermic core in which the allantoid vessels are contained

b) a mesh-like network of small cavities that continue with the outer layer of the endodermal sinus and which are lined by epithelial cells

c) aggregates of highly non-differentiated neoplastic cells

d) presence of hyaline, PAS-positive globules located within and without the cell.

The presence of areas resembling choriocarcinoma during remission are not surprising since they have been described, albeit rarely, in pure endodermal sinus tumor.

Despite the combination therapy employed, the child died 8 months following tumor appearance, thus confirming that, chemotherapy currently available and/or radiation therapy is not efficacious with this neoplasm. Radical surgical inter-

vention is not always possible due to the rapid spread of the neoplasms which tends to involve various endoabdominal structures, thus rendering complete exeresis impossible. The average survival time after tumor appearance is approximately one year.

SUMMARY

A case of pure endodermal sinus tumor of the ovary is described in a 5 year old girl. Histological features of this rare neoplasm are described. Despite various therapeutic attempts, the child died within one year.

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Description of a simple method for the dosage of glycerophospholipids in human amniotic fluid

by

A. CASU *, R. MONACELLI * and D. PECORARI **

Amniotic fluid phospholipids have been the object of increasing research activity in recent years owing to their relationship with the respiratory distress syndrome of the newborn. In most mammals toward the term of gestation there is a marked rise of concentration of dipalmitoil-phosphatidyl-choline in amniotic fluid, which is coincident with the acquisition of surface tension lowering properties by fetal lung extracts⁽¹⁰⁾. Among the many papers on this argument, however, only a minority report comparable and reproducible data; this is mainly due to the fact that:

- a) amniotic fluid often is not centrifuged, so that the phospholipids measured are the sum of phospholipids actually present in amniotic fluid and of those present in much greater amount in cells and subcellular membranes suspended in amniotic fluid.
- b) Amniotic fluid is centrifuged, but not immediately after it is obtained; hence, there is a diffusion of phospholipids into the liquid phase, owing to decomposition of subcellular particles. An important review of the main errors that

* From the Institute of General Pathology and Chair of Molecular Pathology University of Genova.

** From the Department of Obstetrics and Gynaecology I., University of Genova.