Endodermal sinus tumour of Teilum

by H.S. Ahluwalia and *D.C. Sharma*

Institute for Medical Research, Kuala Lumpur

General Hospital, Penang

WHILE ENDODERMAL SINUS TUMOUR of Teilum generally originates from the germ cells in the gonads or in certain extragonadal sites, we have no knowledge of any case originating in the umbilical region and hence consider it worthwhile placing the following report of a case on record.

Case Report

The patient, a three-year-old Chinese male child was asymptomatic until two months before his admission to the General Hospital, Penang. The mother had noticed a gradually enlarging abdominal lump with an accompanying loss in weight and decrease in appetite. Examination of the abdomen revealed a firm, mobile lump over the umbilical area with distension of the overlying superficial cutaneous veins.

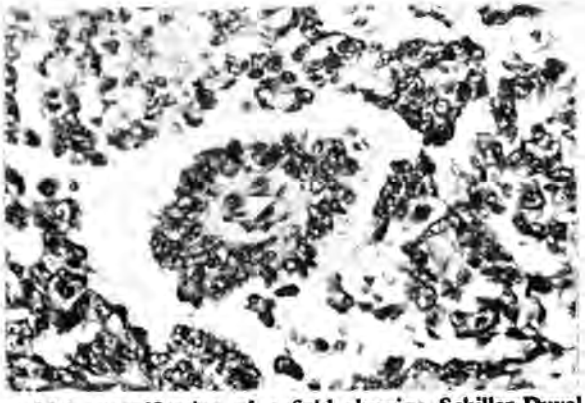
An abdominal X-ray taken revealed a soft tissue mass in the centre part of the abdomen which on lateral view, appeared to be situated anteriorly.

At laparotomy, a fleshy, friable, grey growth of about 7.5 cms size with central areas of necrosis and haemorrhage was found. It was seen arising from the umbilical region and was adherent to the adjoining small intestine, omentum and peritoneum. The testes were free from the tumour as were the lungs, kidney, liver and adrenals. The tumour was excised from the umbilicus after separating it from the surrounding structures.

The patient was lost to follow-up and presumably died shortly after being seen.

Histopathology

The sections from the tumour showed a variable appearance with solid and cystic areas. The epithelial cells were undifferentiated and arranged in solid aggregates in some areas: in other areas, there were papillary and glandular alveolar patterns. The tumour stroma showed fibromyxoid change. In the more vascular parts, the appearance was angioblastic. Epithelial vascular mantling was common. Also conspicuous were peculiar structural units with glomerular-like invaginations (Schiller-Duval bodies). In addition, there were areas of haemorrhage, necrosis and cystic degeneration.



A high magnification of a field showing Schiller-Duval body.

All these findings corresponded well to the description of endodermal sinus tumour of Teilum (1959) or Yolk sac tumour (Huntington 1970).

The endodermal sinus tumour of Teilum has been given a variety of names. In the ovary, Schiller (1939) called it a mesonephroma, supposing that it arose from misplaced foetal remnants of the mesonephros and he described a glomerular-like unit as its distinguishing feature. Kazancigil et al (1940) failed to find evidence of its derivation from remnants of the primitive mesonephros and regarded it as papillo-endothelioma. Other names suggested were mesoblastoma and embryonal cell carcinoma. In the testis, it has been called embryonal carcinoma, clear cell carcinoma and distinctive carcinoma. At extragonadal sites it has been reported as teratoma, ependymoma and choroidal teratoma.

Thus, it would appear that the microscopic pattern is easily recognised, although the histogenesis has been controversial. There are many arguments as to the origin of this tumour but Teilum's concept seems most acceptable.

In 1959, Teilum presented an excellent study of the morphogenesis of the mesonephroma ovarii and compared it to extraembryonic structures of the rat's placenta. He presented convincing evidence that the pattern of this tumour reproduces characteristic stages in the phylogenetic development of such extraembryonic structures as the allantois and yolk sac. He identified the so-called glomerular-like units of Schiller's "mesonephroma" and found these to compare in every detail with the specific intraplacental, perivascular structures in the rat's placenta, i.e. endodermal sinuses of Duval. He also suggested that the irregular communicating vascular channels reflected the supporting vascular mesoderm of the labyrinthine placenta. He named this entity an endodermal sinus tumour.

The tumour is rare and generally occurs in the ovaries and testes and has been reported also at extragonadal sites such as the anterior mediastinum, the region of the pineal gland, the sacrococcygeal region and the vagina in infants. It may occur in either sex and is seen most commonly in the first two decades of life. One case has been described by Huntington, on the first day of life. The patients most commonly present with a history of pain or an enlarged abdominal mass. Generally the tumours vary in size, and are friable, haemorrhagic and necrotic. The salient histological features in the recognition of this tumour are:

(1) Schiller-Duval body, i.e. the glomerular-like unit which is a projection containing a blood vessel surrounded by loose connective tissue and covered by undifferentiated cuboidal or columnar malignant cells.

(2) A cavity lined by endothelial cells.

(3) In other areas, the tissue may appear angiomatoid, fibrous, or myxoid.

The prognosis of these cases is poorly treated or untreated; and the longevity is a few months. Radiation and chemotherapy have produced discouraging results.

Acknowledgement

I would like to thank Prof. G. Teilum of the Copenhagen University, Institute of Pathology for having confirmed the histological diagnosis, and Prof. G. Montogomery of the University of Malaya, for his encouragement. I am also grateful to Dr. R. Bhagwan Singh, Director, Institute for Medical Research, for approval to publish this case.

References

Huntington, R.W., & Bullock, W.K.: Yolk Sac tumours of the ovary. *Cancer 25*: 1357-1367, 1970.

Huntington, R.W., & Bullock, W.K.: Yolk Sac tumours of extragonadal origin. *Cancer 25*: 1368-1375, 1970.

Huntington, R.W., Morgenstern, N.L., Sargent, J.A., Giem, N.R., Richards A. & Hanford, K.C.: Germinal tumour exhibiting the endodermal sinus pattern of Teilum in young children. *Cancer 16*: 34-47, 1963.

Pierce, G.N., Bullock, W.K. & Huntington, R.W.: Yolk Sac tumour of testes. *Cancer 25*: 644-658, 1970.

Teilum, G.: Endodermal Sinus tumour of ovary and testes. *Cancer 12*: 1092-1105, 1959.

Teilum, G.: Personal communication.