ENDODERMAL SINUS TUMOUR (YOLKSAC TUMOUR) OF THE OVARY

(A Case Report)

by

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'Endodermal sinuses' is the name given by Duval (1891) to diverticulae of yolk sac growing into extraembryonic mesenchyme. The association of endodermal sinuses to certain tumours of gonadal origin in the human was suggested by Teilum (1959) after observing papillary formations in the rat placenta similar to that found in some of the tumours of the ovary classified as Schiller's mesonephroma. The morphological similarity between experimental yolk sac carcinoma and yolk sac tumours of testis demonstrated by Pierce *et al* (1970), clearly identifies endodermal sinus tumour (yolk sac tumour) as a distinct histopathological entity, separating it out from embryonal carcinomas and teratomas. These tumours are rare and seen in young children and rarely in older age groups. One such tumour was recently removed from an 1½ year old child in the T. D. Medical College Hospital, Alleppey. To our knowledge there is no report of a similar case in Indian literature.

Case history

A 1½ year old female child was admitted to the hospital about 9 months back with the complaints of fever and cough of two weeks' duration. She was the third child. The elder two are healthy. Parents are healthy. Weighed 8 kgs. On admission the child was ill looking and anaemic. Respiratory rate 38/minute, regular. Except a few fine crepitations at the infraclavicular region on both sides nothing special was detected in the respiratory system. Cardiovascular and central nervous systems were normal. Abdomen distended, liver and spleen not palpable. A mass was palpable on the right side at the level of the umbilicus. The mass measured approximately 8 x 10 cms. extending up to the epigastrium, mobile, firm and tender. Mobility more in oblique axis, not bimanually palpable. Fingers could be insinuated between the swelling and the costal margin. Dull on percussion. No shifting dullness or fluid thrill.

Haemoglobin 7.5 grms%, TLC 19,200/cmm. Poly 58%, lymph 42%; ESR 16 mm/hr. Urinalysis—nothing special. Stool: No ova or cyst present.

A provisional diagnosis of? Mesentric cyst? ovarian tumour was made. The patient was treated with antibiotics and general supportive measures. Two weeks after the admission a laparotomy was done. An ovarian tumour arising from the right ovary

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was seen. The tumour was soft, reddish-white with a nodular surface. Not adherent to any structure. An ovariotomy with appendicectomy was done and abdomen closed.

Post-operative period was complicated by diarrhoea and wound infection which was controlled by appropriate treatment and the patient was discharged two weeks after the operation. The patient was readmitted two months after with distention of abdomen and difficulty in breathing. Examination showed no abdominal mass. After passing a flatus tube the flanks were found to be dull on percussion. Two hundred and fifty ml of blood-stained fluid was aspirated. Since the patient was in a moribund condition the parents took the patient home.

Pathological Report: Gross: The operated specimen was a lobulated firm mass measuring 10 cm. in diameter. The fallopian tube was seen stretched over the surface. The surface of the tumour was smooth and showed dilated vessels. Cut section showed a lobulated whitish appearance. Small cystic spaces filled with colloid like material were seen. The surface presented a mucoid feel (Fig. 1).

Microscopically the neoplasm was composed of an irregular network of small and large spaces lined by flattened, cuboidal or low columnar cells, separated by loose myxomatous connective tissue stroma. Papillary projections were seen into some of these spaces which were covered by columnar cells. Papillae were composed of blood vessel and loose connective tissue. (Fig. 2 & 3) In some areas the cells were closely packed into small acinar groupings. In other places they were widely separated by loose tissue showing eosinophilic material which was weakly P.A.S. positive. Based on these histological findings a diagnosis of "endodermal sinus tumour" of the ovary was made. The aspirated ascitic fluid on Papacolou stain showed malignant cells.

Discussion

Endodermal sinus tumour is a rare neoplasm. There is still much confusion concerning the proper terminology and classification of these tumours. Shiller (1939) considered them to be neoplasms of ovary and called them mesonephroma, because they contained microscopic structures resembling mesonephric glomeruli. However, Kazancigil *et al* (1940) contended that these pseudoglandular structures were papillary projections forming a network. Teilum (1959) considered them to recapitulate stage in the phylogenetic development of extraembryonic structures, such as allantois and yolk sac. He observed papillary formations in the rat placenta that were similar to those found within this tumour. These structures had been called endodermal sinuses by Duval (1891) considering them to represent diverticulae of the yolk sac growing into extraembryonic mesenchyme. Pierce *et al* (1970) demonstrated a morphological relationship between yolk sac carcinoma of the mouse and the endodermal sinus tumour (yolk sac tumour) of human testis.

Endodermal sinus tumours are usually met with in the ovary and testis. Extragonadal occurrence of these tumours have been reported. These extragonadal cases are usually associated with a teratoma or an embryonal carcinoma. However, simple extragonadal endodermal sinus tumours unassociated with a teratoma or an embryonal carcinoma have been reported by many. Anterior mediastinum (Teilman *et al* 1967, Scully 1969), Sacro-coccygeal (Huntington *et al* 1970), in the region of pineal gland (Bestle 1968), pelvis (Huntington *et al* 1970) and vagina (Allyn *et al* 1971) are some of the sites reported. Gonadal endodermal sinus tumours occur in young children. Extra gonadal tumours occur in any age group, particularly in the first three decades.

Grossly these tumours of the ovary vary in size from a few cms. to 25 cms. or more, average being 10-15 cms, in diameter. The surface is usually smooth and nodular.

The cut surface is soft whitish or greyish-white with foci of cystic degeneration or necrosis. Microscopic structure is characterised by a discrete network of spaces lined by cuboidal epithelium into which papillary projections protrude. The papillary projections are supported by a delicate connective tissue stroma and shows thin walled capillaries and it is covered by a single layer of high cuboidal or columnar cells. This appearance is described sometimes as 'glomerulus like'. Formation resembling alveolar structure without papillary pattern tending to form a system of communicating channels may be seen. Suggestion of mucoid production is seen in PAS positive droplets in the alveolar spaces. The present case in the young girl showed all the characteristic features, both macroscopic and microscopic of endodermal sinus tumour. Section from multiple blocks of the tumour showed no evidence of teratomatous elements.

The prognosis of endodermal sinus tumour is uniformly bad in cases below the age of 2 years. Both the cases who died in those reported by Pierce *et al* (1970) were below the age of 2. The extragonadal endodermal sinus tumour reported by Thiele *et al*, (1971) who died of extensive secondary deposits, was a child of 14 months. In the present case even though the tumour was well encapsulated at the time of removal, must have produced definite metastases as evidenced by the presence of malignant cells in the peritoneal fluid.

Conclusion

A case of "endodermal sinus tumour" (Yolk sac tumour) in a child of 1½ years is described. The pathology of the condition is briefly discussed.

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See Figs. on Art Paper II