

# GONADOBLASTOMA ARISING FROM A GONAD OF UNCERTAIN ORIGIN

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## Introduction

Gonadoblastoma is a rare tumour first described by Scully in 1953. It is defined as a tumour almost invariably arising in a dysgenetic gonad and is composed of germ cells, sex-cord derivative of immature granulosa or Sertoli cell type and stromal elements resembling lutein or leydig cells. The case presented here is an atypical case of gonadoblastoma arising from a dysgenetic gonad of unknown origin in a Phenotypic female.

## CASE REPORT

Mrs. M. M. 25 years was admitted with a history of primary amenorrhoea and lump in the abdomen with pain for 1 year. The patient is married for 5 years, sterile. Her menstrual

history revealed that she used to get cyclic vaginal discharge without any obvious vaginal bleeding. Family history—The patient was having 3 sisters, all were married and having children.

On examination: Tall Phenotypic female, Height 5'6", span 5'5", weight 55 Kg., Secondary sex characters were not properly developed. Breasts were not developed. Axillary and pubic hair were scanty. Carrying angle of upper arm increased to certain extent. No webbing of the neck.

## Abdominal Examination

A firm mass of 3" x 3" x 2" occupying the lower abdomen, well defined mobile from side to side.

**Vaginal Examination:** External genitalia were infantile, female type with normal clitoris. Vaginal canal was dry, admitting one finger. Uterus was smaller than normal; cervix was small, directed backwards, Fornices were free.

**Investigations:**—Blood picture—normal. Plain x-ray abdomen—a soft tissue shadow in lower abdomen with no evidence of calcification X-ray skull and chest—showed no abnormality.

Vaginal cytology—mild oestrogenic feature.

Cervical mucus test—No mucus obtained.

Buccal smear—Chromatin (+ve) (18%).

Prior to laparotomy, E.U.A. and curettage was done. Uterus was found to be of 1½" length. No endometrium was obtained from the uterine cavity. On opening the abdomen, uterus was found to be smaller in size with normal fallopian tubes. The ovaries were smaller sized, whitish (2 cm. x 1 cm x ½ cm.) and normally situated. Two tumour masses

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were found in the pelvic cavity which had no attachment with the ovary. The Right one apparently looked like normal size testis. The left one was bigger in size (4" x 4" x 3") infiltrating into the neighbouring sigmoid colon. About 6" segment of the sigmoid colon along with the tumour masses (right and left) were removed. End to end anastomosis of the colon was done. Biopsy was taken from both the ovaries.

#### Histopathological Examination

The tumour masses were solid, soft to firm consistency, irregularly lobulated and partly encapsulated. On cutting, the tumour presented greyish-yellow fleshy homogenous surface with big pale areas of infarction. No haemorrhage or cystic degeneration was noticed. The segment of colon had markedly thickened wall which was infiltrated by greyish-pink to black soft tumour mass causing eccentric displacement of the lumen.

The tumour consisted of round, polygonal, uniform cells arranged in small groups, solid narrow cords or irregular sheets separated by connective tissue septa. Cell outline were well demarcated with moderate amount of cytoplasm. The nuclei were uniformly round and showed presence of prominent nucleoli. Fair number of mitotic figures were also seen. Connective tissue septa were infiltrated by lymphocytes. Lymphatic invasion by the tumour was also present.

The wall of the colon showed infiltration by the similar type of tumour cells. (Fig. 1). Right ovary showed dense wavy connective tissue with no primary and secondary follicles. Biopsy from left ovary had hyalinized fibrous tissue.

#### Discussion

Scully (1970) has commented on 74 cases of gonadoblastoma of which 55 were collected from the literature and 19 were personally studied but unreported cases. Hughesdon and Kumarasamy (1970) recently reviewed the literature and found 60 reported cases of which 45 were considered as authentic.

No true satisfactory theory of origin of this tumour has yet been put forward. Hughesdon and Kumarasamy (1970) suggested that the initial stage in the

development of a gonadoblastoma is germ cell neoplasia which ultimately evokes a stromal response in which granulosa Sertoli cells may develop. Teter (1960) had considered the gonadoblastoma as being one of a group of tumours that he classed as gonocytoma. He had subdivided these neoplasia into 4 categories. The tumour which has been described here falls under the category of gonocytoma Grade I which consists of only germ cell element.

Forty per cent of patients in whom a gonadoblastoma has been diagnosed are non-virilized phenotypic female (Jasso *et al* 1969) where primary amenorrhea was almost invariably the rule. On surgical exploration in these patients the uterus was found to be of infantile type. In approximately half of the reported cases of gonadoblastoma, the tumour was found to arise in a gonad of uncertain origin, where the opinion could not be made as to its true nature. The tumour is found to be bilateral in 30% of the cases. The present case presented as a non-virilized phenotypic female with history of primary amenorrhea and pain in abdomen. The gonads were of uncertain origin and the tumor was bilateral.

Though malignant potentiality of a pure gonadoblastoma has not yet been established, considering its invading nature in the present case and presence of mitotic figures in histopathological examination, the patient was given a course of radiotherapy. Sirsat *et al* (1965) and Scheuhas *et al* (1971) described cases of gonadoblastoma where there was local invasion of germ cell element.

It may be mentioned that in this case the tumour mass arising from the gonad of uncertain origin did not have any attachment with either of streak ovary. Though apparently it looked like testis,

exhaustive histopathological study could not demonstrate any testicular tissue. The authors feel it to be arising either from a foetal gonad undergoing malignant changes later on or from a dysgenetic testis where the tumour had completely replaced the gonadal tissue.

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See Figs on Art Paper II