

GONADOBLASTOMA ASSOCIATED WITH DYSGERMINOMA AND EMBRYONAL CELL CARCINOMA

(A Case Report)

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Introduction

The term gonadoblastoma was coined by Scully (1953) to a tumour capable of producing sex hormones. Later cases were reported to have association with dysgerminoma. It is only in 1970 that Scully reviewing 74 cases of gonadoblastoma showed an association with dysgerminoma in 50% of cases and a rare occurrence of this combination with more malignant germ cell tumour like embryonal cell carcinoma, endodermal sinus tumour, choriocarcinoma and malignant teratoma. These cases need reporting because clinically and histologically gonadoblastoma do not exhibit malignant characters, and even when associated with dysgerminoma the prognosis is not that bad, but, an association with embryonal carcinoma makes it a highly malignant tumour with poor prognosis.

Case Report

A 19 years old Hindu female was admitted to

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the hospital with amenorrhoea of 2 years' duration followed by long cycles with scanty bleeding for last one year and a lump in the lower abdomen. From menarche at 13 years, she had normal cycles till development of symptoms. She also noticed a small lump in left lower abdomen after the menstrual irregularity. The mass was gradually increasing in size, but for the last 2 months rapid increase in size was noted. The patient was of average built with well developed secondary sex characters. Her systemic examination was normal. On bimanual examination the uterus was of normal size, with a big mobile, non-tender lump in the left fornix, while right fornix was clear. Haemogram, urine analysis, X-ray chest were normal.

On laparotomy a well encapsulated nodular, ovarian solid mass, partially adherent to the left fallopian tube was seen. The uterus and right adenexa were normal. There was no free fluid in the peritoneal cavity nor any evidence of secondaries in the pouch of Douglas or omentum. Left salpingo-oophorectomy was done. Postoperative recovery was uneventful.

After 18 months of operation nodular secondaries were present in the operative scar as well as in the right ovary. X-ray showed metastases in the lungs.

Gross: Left sided ovarian mass measured 15 x 12 x 10 cms and weighed 758 gms. It was multinodular, greyish white and solid in consistency. The nodules measured 1 to 3.5 cms in diameter. Cut surface had a variegated appearance. Normal ovarian tissue could not be recognised. Yellowish white homogenous masses

were separated by brown friable areas. There were three firm bright yellow areas situated near the centre of the tumour. Slit-like spaces were also observed in the firm areas of the tumour.

Cytology: The imprint smear showed following types of cells. (1) large cells with clear foamy eosinophilic cytoplasm and small densely stained nucleus in the centre similar to Leydig cells. Second type of cells were smaller than previous ones with opaque, eosinophilic cytoplasm and large vesicular or compact nucleus similar to Sertoli cells. Along with these small lymphocyte-like cells were also seen. Some cells showed abnormal mitosis. At places structure like Call-Exner bodies were also noticed (Fig. 1).

Histology: Multiple sections from different regions were taken. The capsule was intact with bits of ovarian tissue. The tumour was pleomorphic, revealing typical areas of dysgerminoma having groups of cells separated by thin connective tissue septa with scattered lymphocyte-like cells (Fig. 2), at other places large dysgerminoma cells were intermixed with Sertoli-like cells and attempted formation of Call-Exner bodies (Figs. 3). Small hyaline bodies were also seen in between germ cells and/or Sertoli cells. In two of the sections there were cystic spaces having papillary projection lined with large cells having eosinophilic cytoplasm and vesicular nuclei with fair number of abnormal mitosis (Fig. 4). There was also vascular invasion. **Histological diagnosis:** Gonadoblastoma with dysgerminoma and embryonal cell carcinoma.

Discussion

Gonadoblastoma, a functionally active tumour of sex cord, represents gonadal development more completely than any other type of ovarian tumour (Scully, 1953). The patients are phenotype female in 80%, phenotype male in 20%, Talerman (1973). The exact incidence is not known but Scully (1970) after a critical review of literature, could find only 55 reported cases and added his own 19 cases. Upto 1971 only 81 cases were reported. Since then only few reports have come up in literature, (Talerman, 1974; Damjanow, *et al* 1975). Many

authors including Scully (1970), Teter (1970), Talerman (1971, 1973 and 1974) have reported that it bears a good prognosis and metastases do not occur even in bilateral tumour of quite big size. Therefore, a complete removal of tumour is sufficient.

Nearly 50% cases of gonadoblastoma are associated with dysgerminoma. In very rare instances it may be associated with more malignant tumours i.e. embryonal cell carcinoma, endodermal sinus tumour, choriocarcinoma or malignant teratoma. Uptil now only 4 cases have been reported with embryonal cell carcinoma and in all the cases prognosis was very poor and survival was less than 18 months. The present case was also associated with embryonal cell carcinoma with extensive infiltration in blood vessels, though the capsule was intact microscopically. The patient showed metastases in the lung within 18 months of removal, thus capsular infiltration can not be considered as an important factor in assessing prognosis and blood vascular invasion should be carefully looked for. Histopathologically, calcification is a common feature but in this case there was no gross, radiological or microscopic evidence of calcification, only areas of hyalinization were seen. It is possible that a rapid growing tumour may not be associated with calcification (Scully 1970).

In view of the poor prognosis whenever histological diagnosis of Gonadoblastoma with a germ cell tumour is made, not only the pathological gonad should be removed but a pan-hysterectomy should be done as is the standard procedure for any malignant ovarian tumour and precautions for distant metastasis should be taken.

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See Figs. on Art Paper IX