ENDODERMAL SINUS TUMOUR IN A GIRL WITH MIXED GONADAL DYSGENESIS

by

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Endodermal sinus tumour, as defined by Teilum (Fox, Longley) is a highly malignant germ cell tumour showing selective over growth of yolk sac endoderm, intimately associated with extra-embrayomic mesoblast.

CASE REPORT

Miss S. a Hindu girl of 15 years was admitted with fever for 1 month, dysuria and retention of urine of 6 days duration. She also noticed a painful swelling in the lower abdomen a week before admission.

She did not attain menarche. She is the second of the three daughters. Her elder sister attained menarche in her 14th year, and is menstruating regularly. Younger of the three is eight years.

On physical examination, the girl was found to be very much under nourished, anaemic with a low intelligent quotient. There was evidence of webbing of the neck (Fig. 1) and increased carrying angle at the elbow joints. Absence of development of breasts, axillary and pubic hair was noticed. Cardiovascular and respiratory systems were normal.

Abdominal examination revealed a distended bladder almost upto the umbilicus. After catheterisation, a firm swelling arising from the pelvis, occupying both the iliac fossae, left lumbar and umbilical regions was felt. On rectal examination the tumour mass was found to be filling the pouch of Douglas and pushing the posterior vaginal wall forwards.

Malignant ovarian tumour in a dysgenetic ovary was the diagnosis made as the tumour was very rapidly growing and there were stigmata of Turner's syndrome.

Investigations: Haemoglobin, 8.6 G. per cent. Urine—No albumin or sugar: 10 pus cells per high power field. Blood urea = 18 mg. per cent.

Bacteriological examination of urine was negative for pyogenic organisms, 17-Keto steroids—3 mg/24 hours urine excretion. Nuclear sex chromotin bodies were not seen in the buccal smear and her karyotype was 46. Xx with double deletion of both short and long arms of one X-chromosome. (Fig. 2). Tissue culture from skin and tumour mass was done for evidence of mosaicism, but the attempts were unsuccessful. X-Ray of skull revealed a small pituitary fossa and chest X-Ray was normal.

On Laparotomy, a highly vascular, tumour arising from the ovary, adherent to the pelvic wall on the left, was found. Towards the right side of the tumour, it was cystic containing (altered blood) dark-coloured fluid. The lower pole of the tumour was in the pouch of Douglas, quite adherent to it. Uterus was 1" x ½" in size stretched over the tumour mass. Right ovary was like a streak externally. While separating the tumour, the capsule ruptured and the soft friable tumour tissue escaped. Left ovariotomy was done. The other ovary was excised. Compatible blood was transfused pre and postoperatively. Wound healed by first intention. Mitomycin—

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10 mg. daily for 5 days was administered, as complete removal of the tumour was not possible because of adhesions. The tumour reappeared, and was growing very fast. The girl was reluctant to take food, complained all the time of vague pain in the abdomen. Methotrexate was administered orally for 5 days as that was the only drug available at that time. As the patient was unwilling to stay in the Hospital, her parents took her home. A month after she left the hospital, we were informed that she died.

Discussion

Endodermal sinus tumour occurs predominently in young women, mean age ranging from 12.9 to 18.6 years and no case has been reported above 38. (Fox and Langley 1976). The age of the girl reported here was 15 years. The most common presenting symptoms and signs are those resulting from the presence of an abdominal or a pelvic mass, though occasional manifestations of an endocrine disorder have been reported in combined tumours. Pain in the abdomen is a common symptom, either due to haemorrhage into the tumour, torsion or rupture, but the latter appeared to be the cause here. Fever was a conspicuous feature in 6 cases reported (Fox and Longle 1976) as in the case reported here.

Retention of urine due to a very rapidly growing tumour into the pouch of Donglas and lifting up the uterus and stretching the urethra was the presenting symptom in this patient. Karyotype of the peripheral lymphocytes showed a double deletion of X-chromosome, (46 Xx) a rare abnormality reported by Theakoske-Westphal *et al* (1974) in a 21 year old woman with primary amenorrhoea without the physical stigmata of Turner's syndrome. Streak and dysgenetic ovaries have a high potential for neoplasia, if the Karyotype contains 'Y' chromosome. Hence the tumour tissue, skin and a bit from the other ovary were subjected for tissue culture and Karyotype study but without success. Detection of Alpha—Fetoprotein in the serum or ascitic fluid is an evidence of functioning yolk sac tissue, a further evidence of the presence of endodermal sinus tumour.

The histological picture of the tumour in the present case was mostly of perivascular formations (Fig. 3). The contralateral ovary showed the presence of seminiferous tubules lined by atrophic epithelium surrounded by fibrous stroma (Fig. 4). There were no primordial folliceles. This was an other peculiar feature in the case suggesting-mixed gonadal dysgenesis, a form of intersex at the chromosomal level as was classified by Dewhurst (1976).

The primary mode of treatment is surgical excision of the tumour. If metastasis has already occurred, radiation therapy, following surgery and some times chemotherapeutic drugs may be of some use. Smith and Rutledge (1975) reported remission in 15 out of 20 patients treated with a combination of Vincristine, Actinomycin-D, and Cyclophosphomide.

Remmission for nearly 4 years was reported with intermittent Chlorambucil therapy following surgical excision of the tumour by David *et al* (1977). In general the prognosis is very poor and even in the few cases with remmissions reported so far, the survival was only for few months.

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References

1. David, S. E., Pamley, T. H. Owellen, R. J. and Davis, E. E.: Obstet. Gyneac. 49: 535, 1977.

2. Dewhurst, C. J.: Integrated Obstetrics and Gynaecology ed. ii, Oxford 1976; Blackwell Scientific Publications.
3. Fox, H., Longley, F. A.: Tumours of the ovary Ed. 1; 1976 Williams Hanemeum Medical Books Ltd., London.
4. Harilal, K. R. George, K. P. and Pillai, N. N.: Endodermal sinus tumour of the oary: J. Obstet. Gynaec. India **22**: 715, 1972.
5. Smith, J. P. and Rutledge, R.: Cancer. **36**: 669, 1975.
6. Theakoske-Westphal, R. E. and Seal, E.: The Lancet. **11**: 1577, 1975.

See Fig. on Art Paper VI