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Situs Inversus Totalis with Embryonal Cell Carcinoma of Ovaries

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A very rare association between situs inversus totalis and embryonal cell carcinoma of the ovary is presented in a 20-year-old Saudi female. Recurrence of the tumor in the form of metastases to the left lobe of the liver was successfully treated using a combination of bleomycin, vinblastin and *cis*-platinum. Second-look laparotomy has confirmed the complete remission.

INTRODUCTION

Situs inversus totalis is a rare congenital condition occurring in 1 in 5000 to 1 in 10,000 of all births and in this there is mirror image transposition of both abdominal and thoracic viscera. Several disease entities have been reported with this condition, for example amebic liver abscess [1], duodenal ulcer [2], acute appendicitis [3], cholecystitis [4], and jejunal and ileal atresia [5]. Several tumors have also been reported in association with situs inversus totalis such as carcinoma of the caecum [6] and stomach [7]. To our knowledge only one case of ovarian carcinoma has ever been reported in association with situs inversus totalis [4]. Association with embryonal ovarian carcinoma, which is a rare type of ovarian neoplasm, has never been reported before. The rarity of this condition has prompted us to report this case.

CASE REPORT

A 20-year-old unmarried Saudi female presented in October 1980 with lower abdominal pain. Examination and investigations revealed that she had situs inversus totalis. In addition a pelvic mass was found. In November 1980 exploratory laparotomy was carried out. The left ovary was replaced with a large mass. Only left salpingo-oophorectomy was done. The mass was extensively hemorrhagic and necrotic on sectioning. Slides were sent to Montreal, Canada, for review. The tumor was composed of a network of tubular spaces lined with embryonal

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cells. Papillary structures containing blood vessels protruding into these spaces were also demonstrated. Extra- and intracellular hyaline bodies were found. A diagnosis of embryonal cell carcinoma of the ovary was established. No post-operative chemotherapy was offered. In March 1981 she again presented with recurrence of her abdominal pain. Recurrence was suspected. On reexploration the right ovary was found to be involved and in addition liver metastases were detected. The patient was started on monthly courses of melphalan; however, in September 1982 she demonstrated clear evidence of progression.

Clinical examination revealed a thin female, who was not in acute distress. Apart from her dextrocardia, her only abnormal finding was confined to the abdomen. The liver was palpable in the left hypochondrial region, 7 cm below the left costal margin, with a total span of 17 cm. The liver was firm with a sharp edge and a smooth surface.

Her CBC, urine analysis, BUN, electrolytes, calcium, and creatinine were normal. Her alkaline phosphatase was 15 IU/liter (normal up to 9.7), SGOT 60 IU/liter (normal up to 41), GGTP 200 IU/liter (normal up to 83), and LDH 410 IU/liter (normal up to 190). She had normal CPK, SGPT, and serum bilirubin.

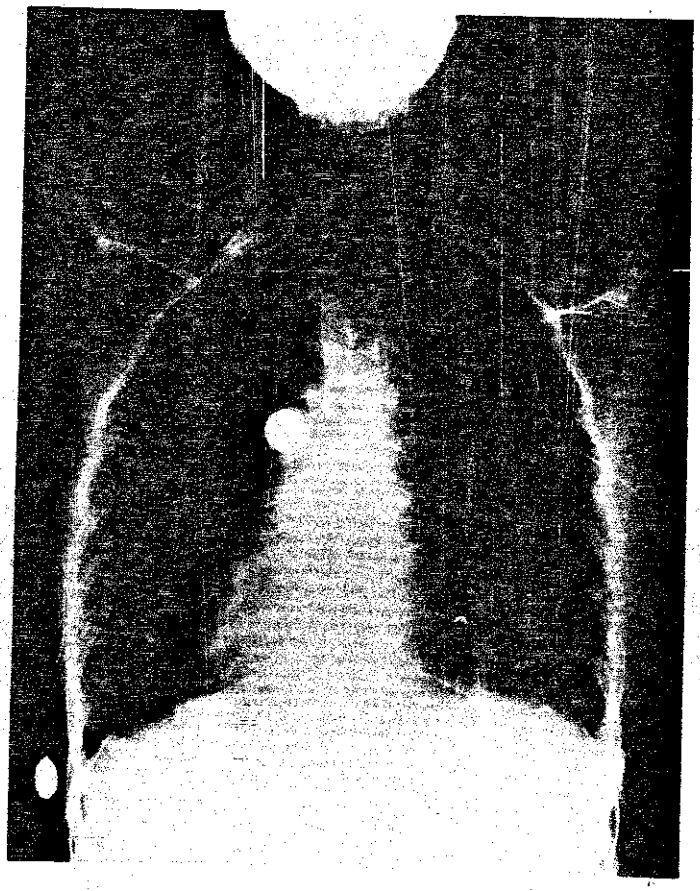

FIG. 1. Chest X-ray showing true dextrocardia.

Her FSH was 74.4 mIU/ml, LH 143 mIU/ml and estradiol less than 10 Pg/ml. Her α-fetoprotein was 650 ngm/ml, and carcinoembryonic antigen was normal. The serum copper was 1cmu 288 μg/dl (normal up to 155), and B-HCG was normal. The chest X-ray was normal apart from dextrocardia (Fig. 1), and pulmonary function tests were normal. The CT scan of the abdomen (Fig. 2a) demonstrated the enlarged left lobe of the liver together with liver metastases.

In October 1982, the patient was started on her first course of chemotherapy using the nonseminomatous testicular protocol. Vinblastin was given as 6 mg/mm on Days 1 and 2. Bleomycin was given as 90 units infused over 36 hr starting from Day 1. *CIS*-platinum was given as 100 mg/mm infused over 6 hr in Day 2. Administration of *CIS*-platinum was preceded by the usual hydration and was accompanied and followed by osmotic diuresis using mannitol together with adequate IV fluids.

The patient received three courses of this regimen at 3-week intervals. Apart from the expected degree of nausea and vomiting the patient did not develop any other toxicity.

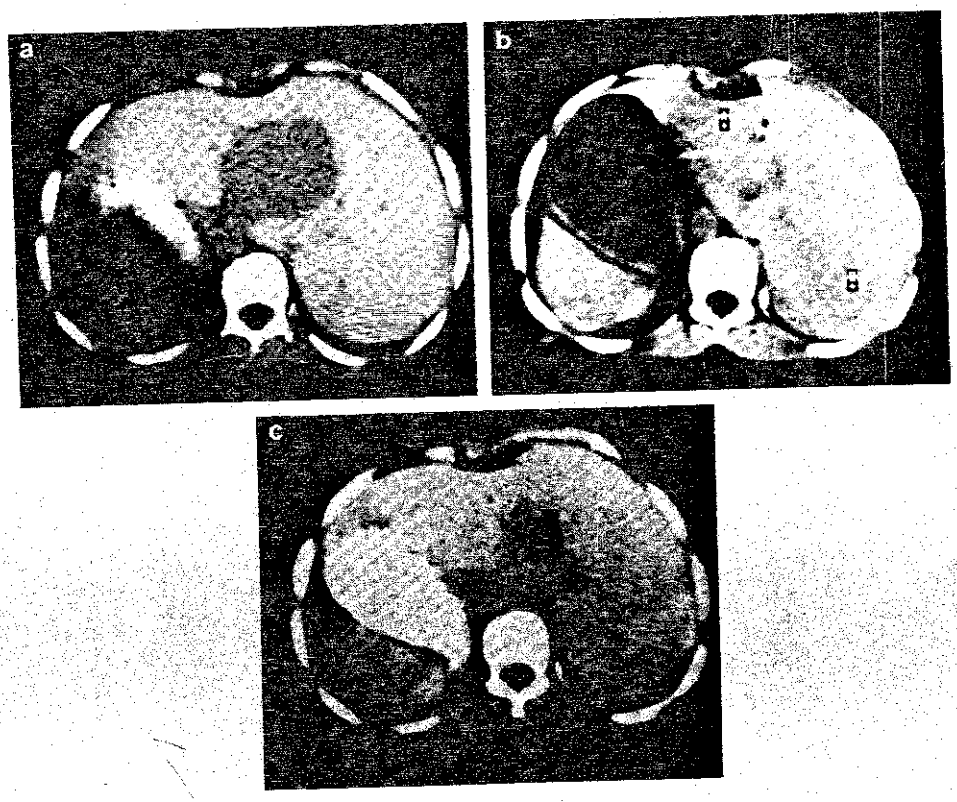

FIG. 2. CT scan examinations of the abdomen. (a) CT scan prior to therapy showing reversed position of organs due to situs inversus totalis. The lobe of the enlarged liver shows a large area of decrease radiolucency consistent with metastatic lesion. (b) repeat CT scan done after the first course of therapy. There is evidence of significant response to combination chemotherapy. (c) CT scan done after the third course. Only an area of necrosis is shown in the liver with no radiological evidence of active metastatic disease.

After the third course her liver function tests and α-fetoprotein were normal. A repeat of the CT scan during therapy (Fig. 2b), and after the third course (Fig. 2c), demonstrated significant remission, with only evidence of necrosis in the liver parenchyma demonstrated.

In January 1983, a second-look laparotomy was carried out and did not reveal any residual disease. The patient refused any further consolidation chemotherapy. In a recent follow-up in March 1983, the patient was in complete clinical remission.

DISCUSSION

Situs inversus totalis should not be considered in any way as a premalignant condition. The occurrence of embryonal carcinoma in our patient with situs inversus totalis is most likely a coincidence. More attention, however, should be directed to the other intra-abdominal anomalies that have frequently been found to accompany complete situs inversus [5].

Embryonal cell carcinoma, also known as endodermal sinus tumor, is a rare type of ovarian neoplasm. It represents probably less than 1% of all ovarian carcinomas [8]. This tumor is highly malignant with a distinctive histological pattern.

This tumor was uniformly fatal until the advent of effective combination chemotherapy [9,10]. Embryonal cell carcinoma of the ovary is analogous to the embryonal carcinoma of the testis. Thus, it was not unexpected that our patient would demonstrate an excellent response to nonseminomatous chemotherapy protocol utilizing vinblastin, bleomycin, and *cis*-platinum.

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