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Case Report

Pure extragonadal dysgerminoma - a rare cause of abdominal mass

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Introduction

Extragonadal germ cell tumors (GCT) are rare with a prevalence of 1% - 2.5% of all GCTs [1]. Pathologic subtypes vary, and the prevalence of mixed tumors is high[2]. We report a case of a young female with pure extragonadal dysgerminoma with supraclavicular lymph node metastasis who presented with abdominal symptoms.

Case report

A 27-year-old woman was admitted on 20th September 2003, with abdominal distension for six months and diffuse dull aching abdominal pain for three months. Pain was nonradiating, with no relation to meals, posture, bladder or bowel function, and no diurnal variation. There was a history of anorexia and loss of weight. She had amenorrhea since two years. Earlier she had normal menstrual cycles. There was no history of fever, pedal edema, vomiting, or gastrointestinal bleeding. She had lower segment cesarean section with tubal ligation two years back. She had a normal delivery earlier and that child is living. She had no other pregnancy. General examination revealed a moderately built, but ill nourished woman with a body mass index of 17.5. Pallor was present along with firm, mobile, palpable left supraclavicular lymph node.

On abdominal examination three lumps, each of 8 x 10-12 cm were palpable in the left hypochondrium, periumbilical region and hypogastrium. The masses were lobulated with irregular margins, and variegated in consistency; edges were not prominent and overlying skin was free. Masses in the left hypochondrium and hypogastrium were fixed while the periumbilical mass was mobile. Flanks were resonant. Liver and spleen were not palpable. Investigations revealed Hb of 8.5 g/dL, leucocyte count of 8500/cmm with a normal differential count, ESR of 115 mm at one hour. Fasting blood sugar and liver function tests were normal, serum proteins – 7.1 g/dL, serum albumin 3.4 g/dL, blood urea nitrogen 53 mg/dL serum creatinine 2.7 mg/dL, serum calcium 10.3 mg/dL, inorganic phosphorous 3.4 mg/dL and serum uric acid 23.9 mg/dL. Electrolytes were within normal limits.

Abdominal ultrasound showed normal liver and a 17x7 cm mixed echogenic lesion in left hypochondrium, another 19x9 cm mixed echogenic lesion in epigastrium extending to umbilical region encasing aorta, another

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19x8 cm mixed echogenic lesion in left lumbar region extending to pelvis, and 8x4.5 cm mixed echogenic lesion in umbilical region. Also present was bilateral hydronephrosis.

CT of the abdomen and chest showed multiple, large, mildly enhancing soft tissue lesions in the peritoneal cavity, retroperitoneal region and the pelvis with mass effect and encasement of vascular structures, uterus and other solid organs, besides bilateral mild pleural effusion and enlarged left supraclavicular lymph node. Ovaries were normal.

Tumor markers showed LDH (lactate dehydrogenase) of 5336 IU/L (normal 100-190 IU/L), CA 125 123.6 µg/dL (normal range 0-35), CEA (carcino embryonic antigen) of 4.09 (normal < 3.4), AFP (á-feto protein) 1.2 µg/dL normal (<5).

Excisional biopsy of left supraclavicular lymph node showed high grade malignant tumor with negative lymphoid and epithelial markers.

Immunohistochemistry showed c-kit (CD-117) positive, other markers like Calretin, CK (creatinine kinase), CEA (carcino embryoanic antigen), PLAP (placental lactogen), HMB (marker for melanoma) 45, S100, CD 3, LCA (leukocyte common antigen), TTF-1 (thyroid transcription factor), CK7 were negative. Morphology and immunohistochemistry were suggestive of dysgerminoma. On 6th October 2003, the patient underwent bilateral DJ stenting for relief of hydronephrosis. Unfortunately she developed infection of the stent, and succumbed to septicemia on 11th October, 2003 despite intensive antibiotic therapy before any definitive treatment for dysgerminoma could be considered. Due to refusal of the relatives postmortem could not be carried out.

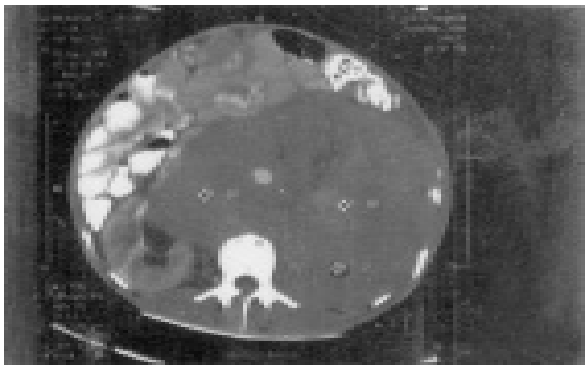

Figure 1. CT abdomen showing dysgerminoma occupying most of the abdominal cavity

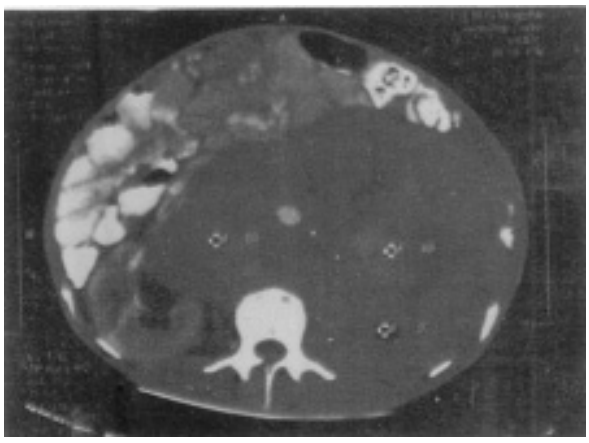

Figure 2. CT abdomen with Contrast showing encasement of aorta by tumor.

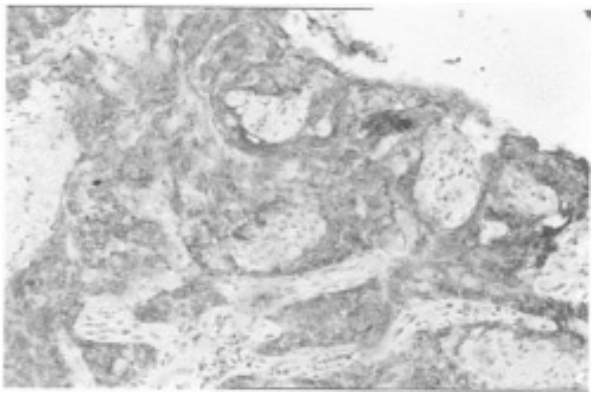

Figure 3. Lymph node biopsy showing lobules of large malignant cells with light staining cytoplasm, vesicular nuclei and prominent nucleoli, H & E stain, 40 X magnification.

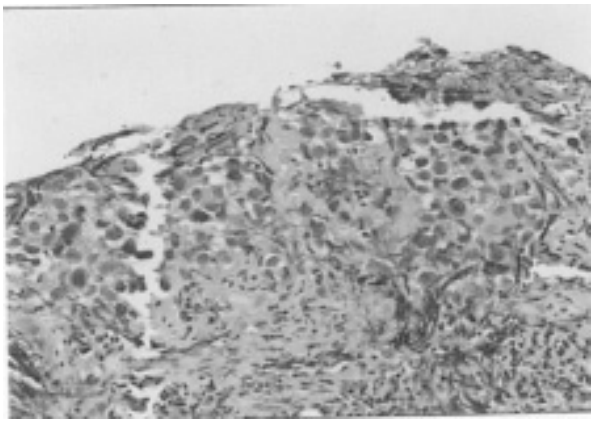

Figure 4. Immunohistochemistry with antibody to C-Kit showingstrong expression in tumor cells. 20x magnification.

Discussion

Dysgerminoma is the most common malignant GCT, accounting for 1-3% of all ovarian cancers, occurring usually in the second and third decade [3]. Extra gonadal dysgerminomas are rare with mediastinum being the most common site of involvement (50-70%), followed by retroperitoneum (30-40%), pineal gland (5%), and sacrococcygeal region (<5%) [4]. Our patient had predominantly retroperitoneal involvement with no lesion in the mediastinum.

Five percent of the cases are seen in women with abnormal gonads like pure gonadal dysgenesis, mixed gonadal dysgenesis and androgen insensitivity syndrome[3]. Our patient was phenotypically and genetically normal female with normal ovaries and normal fertility though we did not do karyotyping. Over 95% of the ovarian GCTs are mature cystic teratomas [1]; however our patient had no cystic component. Unlike testicular tumors, most ovarian malignant GCTs occur in a pure form and like other ovarian neoplasms are notorious for manifesting with mild early symptoms. The most common initial presentation is abdominal distention, which is produced by the tumor itself as in our patient, and ascites which was not seen in our patient. Complications such as rupture or torsion can also occur. Size is generally 5-15cm with bosselated capsule, and spongy consistency. The GCTs exhibit various pathologic features according to the totipotentiality of the tumor cells. Therefore, one-third of the tumors have mixed patterns, and tumors arising from germ cells at different stages of their development exhibit different features [5].

Dysgerminomas have a solid, lobulated, flesh-like gross appearance with a smooth surface. Microscopically cells are round and ovoid, contain abundant cytoplasm, and have irregularly shaped nuclei with more than one prominent nucleolus. These cells have a propensity to aggregate forming cords and sheets. Lymphocytic and granulocytic infiltrations of the fibrous septa are often evident. Extragonadal GCTs are thought to develop from primordial germ cells, which are misplaced during the long trip to the gonads. Recent progress is chromosomal and imprinting analysis strongly supports this theory [5,6].

A pure dysgerminoma shows a normal serum α-fetoprotein level, as seen in our patient. Mixed GCTs containing endodermal sinus tumor elements have elevated serum α-fetoprotein levels. Dysgerminomas have elevated LDH (95%), CA-125 and hCG (3%), C kit (92%) ALP, NSE positivity and elevated PAS, PLAP. Vimentin can also be positive but was negative in our patient. PLAP, in association with AFP and hCG, is of value in the diagnosis and monitoring of gonadal and extragonadal germ cell tumors.

Basic modality of therapy is surgical excision. Tumors are highly radiosensitive. All patients irrespective of tumor histology should received postoperative chemotherapy, for adjuvant or curative purposes. Combination therapies include vinblastine, bleomycin, and cisplatin (VBP); and bleomycin, etoposide and cisplatin (BEP). Combination chemotherapy is given to patients with bulky residual disease or extra abdominal metastases, or those who failed primary treatment with a curative intent. Our patient could not undergo surgery or chemotherapy as she had renal failure, aortic encasement of tumor and fatal septicemia secondary to infected DJ stent despite intensive antibiotic therapy.

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