Retroperitoneal Germ Cell Tumor

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Introduction

Primary retroperitoneal teratomas are rare, representing 5-10% of primary retroperitoneal tumors. The mean age of patients ranges between 13 and 16 years, with cases reported in a fetus and in patients up to 82 years old. Fewer than 10-20% occur after the age of 30 years, with 43-55% diagnosed in the first decade of life[1]. Retroperitoneal mixed germ cell tumors in adults are normally rare in the sacrococcygeal region. Most of these tumors are benign and are probably present at birth. A few show malignant foci, either in the form of germ cell components (trophoblastic, yolk sac) or of adult type carcinornatous tissues[2,3]. Chromosomal analysis of these tumors suggests that they have arisen from post-mitotic or pre-mitotic cells[4]. These tumors may be present at birth in the sacrococcygeal region or can protrude through the abdominal cavity[5,6]. The tumor mass is usually very large and may be cystic and multilobular. There may also be a stubborn adherence of an inflammatory nature to neighboring structures (i.e., pancreas, kidneys, ureters and adrenals)[7]. A knowledge of the neoplasms to be expected in the retroperitoneal site may indicate which pre-operative investigations should be undertaken[8].

Case Report

25 year old man from Rawalpindi presented in the surgical out-patient department of Federal Government Services Hospital, Islamabad. His chief complaint was a hard swelling in the lower abdomen for the last 3 months. The swelling was increasing at a rapid rate. He also complained of persistent lower abdominal pain for the last month. He had no bowel or urinary complaints. Initial diagnosis was of a pelvic growth. Investigations revealed an elevated erythrocyte sedimentation rate (ESR). Blood sugar, urea, liver function test (LFT), creatinine and electrolytes were normal. At that time no serum tumor markers were sent. Abdominal ultrasound investigations revealed a large lobulated mass in the pelvic area. The mass was irregular and of mixed echotexture. Bowel loops were displaced by the mass. Right kidney was echogenic, showing an accumulation of fluid around it with branched renal pyramids and hydronephrotic changes. Liver, spleen and left kidney were normal on ultrasound. Sonography showed that the mass was present in the retroperitoneal space. The patient was advised to have a barium enema and it revealed that the bowel was merely displaced, not obstructed. Testicular ultrasound and biopsy were performed to rule out any primary foci of disease. The testicular biopsy was taken from two random sites. Ultrasound was normal and the biopsy was negative for any malignant changes. Based on these results, the differential diagnosis included the possibilities of retroperitoneal germ cell tumor, lymphoma, or a cyst.
The patient was admitted and prepared for surgery. A complete radiographic workup was done; which included chest, abdomen, peLvis, kidney, ureter, bladder (KUB) and intravenous pyelogram (IVP). His electrocardiogram was normal. Eventually a laprotomy was performed under epidural anesthesia. A large football sized retroperitoneal tumor originating in the pre-sacral area and extending rostrally into the abdomen was discovered. Enlarged intra¬abdominal para-aortic lymph nodes were also present. Lymph node histology was not reported due to

laboratory error. Only 95% of the tumor was removable due to adherence to the adjacent structures. Histopathology report described the specimen to be consisting of multiple irregular pieces of soft tissue measuring collectively 7x7x5 cm. On microscopic description sections showed foci of immature neural tissue, immature cartilage, spaces lined by respiratory type of epithelium and foci having the features of embryonal cell carcinoma. Few areas showed multinucleated giant cells. Areas of necrosis were also seen. The pathology is consistent with a Mixed Germ Cell Tumor-retroperitoneum (immature teratoma and embryonal carcinoma). This patient was not give any additional chemotherapy and radiotherapy as he was lost to follow-up and he did not come back for any post-operative reviews.

Discussion

Primary retroperitoneal tumors are rare, with a reported incidence of 0.3 to 3 percent[9.10]. These tumors are derived from germ cells that failed to migrate to normal gonadal locations. Germ cells are totipotential, undergoing variable differentiation into tissue components that represent derivatives of ectoderm, mesoderm and endodern[11,13], Macroscop ical ly, the cystic teratomas are generally benign, containing sebaceous material and mature tissue. The solid teratomas are frequently malignant, composed of immature embryonic tissue in addition to fatty, cartilaginous, fibrous and bony elements[14,16]. These teratomas occur in many locations including the ovaries. testes, anterior mediastinun, retroperitoneal space, presacral and coccygeal areas, intracranial sites, neck and abdomen.

The commonest primary tumor of the retroperitoneal space are of kidneys, adrenais and lymphomas (non¬Hodgkin's type and B-cell deviation). Symptoms secondary to retroperitoneal neoplasms are vague and appear late in the course of the disease. These are related to the displacement of the organs and obstructive phenomena. The classic radiological methods for the evaluation of the retroperitoneal tumors are plain roentgenograms, barium studies of the Gil and intravenous retrograde pyelograms. Plain radiographs usually show a soft-tissue mass that displaces the bowel. Ultrasound, CT and MRI have largely superseded these techniques. A study conducted by Ellis et al. on 25 patients of metastatic retroperitoneal lymphadenopathy from testicular carcinoma showed that computed tomography correctly predicted the presence or absence of adenopathy in 88% and assigned the correct stage of the tumor in 84%. Nuclear magnetic resonance (NMR) had comparable figures of 84% and 80% respectively[17].

Primar retroperitoneal germ cell tumors are formed by a single mass whereas those, which have metastasized from the testis, tend to involve several groups of lymph nodes on both sides of the peritoneum[18]. These tumors can comprise of mature or immature tissue microscopically. Presence of' immature elements should he evaluated with care in regard to amount and microscopic type. If immaturity is restricted to neuroectodermal components, the tendency is towards spontaneous differentiation. As a result, the behavior of this type of immature teratorna is usually benign, although occasional cases will recur or metastasize[19]. These teratomas can also grow within the abdominal cavity. this being responsible for the clinical observation that teratomas are associated with marked bowel or bladder dysfunction are often malignant. Exceptions in both directions certainly occur, but the majority of teratomas in this region fit into this scheme. Malignant teratomas tend to progress rapidly and are associated with acute symptoms usually including pain. Successful treatment of these tumors remains primarily surgical. An enbioc resection of these malignancies provides the most favorable 5-year survival of 67%[20]. The same was done in this case.

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