CASE REPORT

Primary retroperitoneal seminoma: A case report

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Abstract

Primary retroperitoneal seminomas account for approximately 2% of all seminomas. Despite the availability of ultrasonic examination, differentiating the primary retroperitoneal tumor from a metastatic tumor with an occult testicular primary remains difficult. We present a case of primary retroperitoneal seminoma with totally absent testicular tissue in ultrasonography.

Keywords: Primary; Retroperitoneal; Seminoma

Introduction

A 38 year old, married man from Somali with severe acute abdominal pain referred to Iranian Hospital, Dubai, UAE. On initial examination in the Emergency Room, his temperature was 39°C, heart rate 95/min, respiratory rate 22/min and blood pressure 118/80 mmHg. In physical examination, his chest was clear to auscultation, and no cardiac murmur was audible. The abdomen was tender with rebound. The liver was palpable one cm below the costal margin with no splenomegaly. Neither genital malformation nor gynecomastia was seen. Clinical laboratory studies revealed a white blood count of 10000 cell/mm^3, with 89% neutrophils, 7% lymphocytes, 4% monocytes, hemoglobin of 10.5 g/dl, and a platelet count 230000/mm^3. The aspartate aminotransferase was 30 IU/L, and the alanine aminotransferase was 28 IU/L. The erythrocyte sedimentation rate was 98 mm/h and C-reactive protein was positive. Sodium and potassium serum levels were 129 and 2.6 mmol/l, respectively. The liver and kidney function tests were within normal limit. Tumor markers (HCG, AFP) were normal. Computed tomography showed huge solid tumor, located in the pelvis, with irregular contours (Figure 1), and a significant right hydronephrosis. CT scan of the chest was normal. No clinical features in favor of Klinefelter's syndrome or Down syndrome were present. Past medical and family history were not significant. The tumor was excised and was found to contain multiple fragments of creamy-whitish tissue in the cut section measuring about 5 **Í** 5 **Í** 4 cm (Figure 2). In pathologic examination, there was a diffuse neoplastic proliferation of large, uniform tumor cells arranged in sheets, nests and cords separated into lobules by a supporting stroma containing numerous lymphocytes (Figure 3). Metastatic carcinoma and malignant lymphoma were considered as two main differential diagnoses. In the immunohistochemical study, the malignant large cells were positive for placental alkaline phosphates (PLAP) (Figure 4) and the scattered lymphocytes were immunoreactive for CD45 (Figure 5). The negative immunostaining for cytokeratin, epithelial membrane antigen (Figure 6) and CD30 ruled out the possibility of lymphoma, metastatic carcinoma and embryonal carcinoma. As these findings most likely represent metastatic testicular tumor, a scrotal ultrasound was obtained and revealed no testicular tissue. According to the large residual mass, the patient received systemic chemotherapy with bleomycin, etoposide and cisplatin (BEP). The retroperitoneal mass decreased to a poorly defined plaque in the retroperitoneum. During the follow up, the patient was found to be free from any progression.

Discussion

Extragonadal germ cell tumors (EGCTs) are rare

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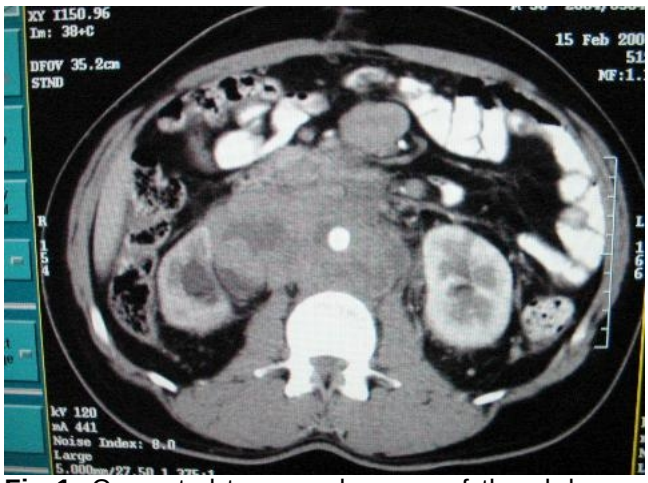

Fig 1: Computed tomography scan of the abdomen show large retroperitoneal mass with right sided hydronephrosis.

Fig 2: Multiple fragments of whitish rubbery tissue.

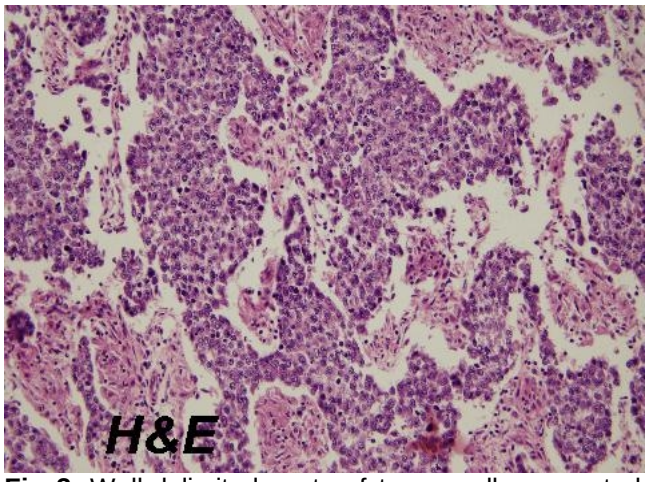

Fig 3: Well-delimited nests of tumor cells separated by fibrous strands containing inflammatory cells (H & E Í 100).

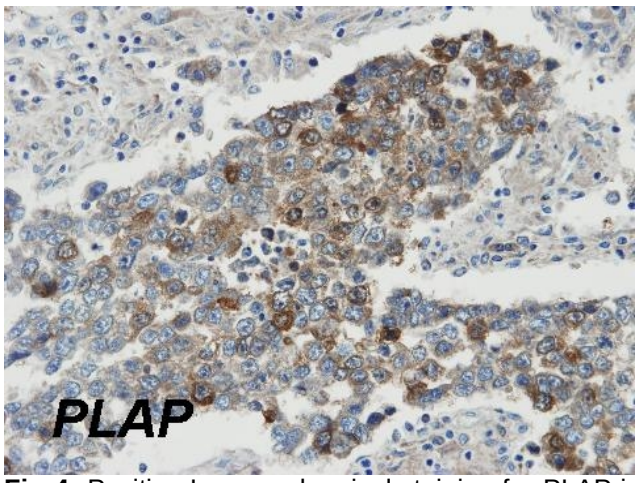

Fig 4: Positive Immunochemical staining for PLAP in tumor cells (Avidin-Biotin complex Í 400).

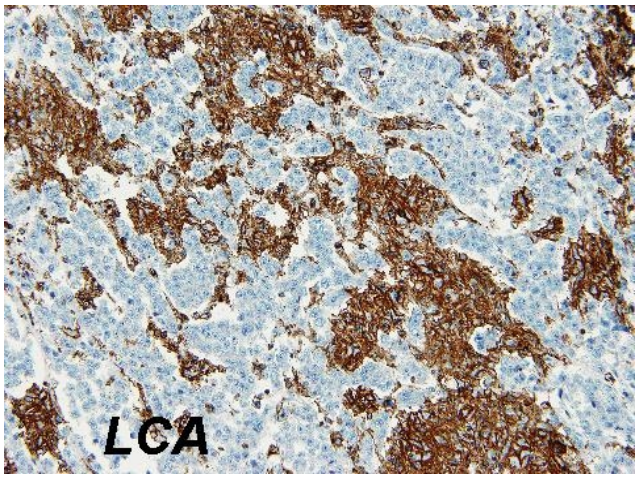

Fig 5: Positive Immunochemical staining for CD45 in lymphocyte (Avidin-Biotin complex Í 400).

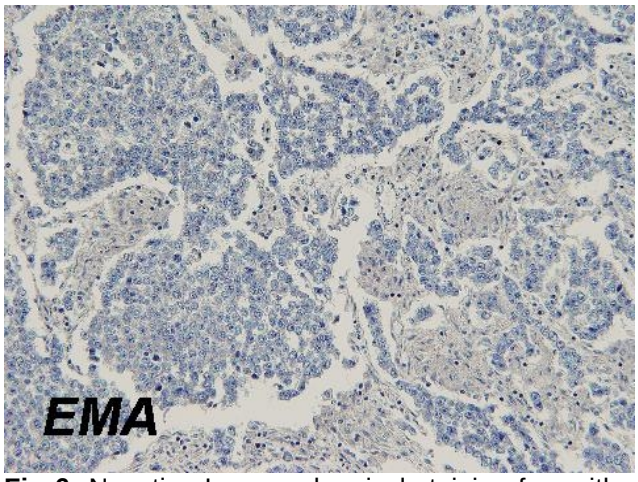

Fig 6: Negative Immunochemical staining for epithelial membrane antigen in tumor cells (Avidin-Biotin complex Í 400).

lesions that have been reported to arise in the pineal body, the thymus, the anterior mediastinum, and the retroperitoneum.[1-4] Pure seminomatous tumors account for approximately 35% of all germ cell tumors but represent approximately half of extragonadal primary tumors found in the mediastinum and retroperitoneum. The peak incidence of seminoma is between 34 to 45 years of age. Before puberty, seminoma is an extremely rare tumor and, in fact, it does not occur in the first decade of life especially in children younger than 5.[5] Grossly, seminomas are firm when intact, but after sectioning, they are soft at palpation with a light tan, moist, and homogenous appearance. In histologic examination, seminoma cells typically have a solid or nested growth pattern separated by thin fibrovascular trabeculae rich in T-cell lymphocytes. The individual tumor cells are large, uniform, round to polygonal, and evenly spaced without any nuclear overlapping. Seminoma cells display a distinct cell membrane and are usually lightly eosinophilic to clear cytoplasm because of the presence of glycogen, demonstrable by a PAS stain. The nuclei contain one or more prominent nucleoli. Mitoses are variable but usually readily identifiable.[5] Immunohistochemically, seminomas are positive for PLAP and c-Kit (CD117) but are negative for epithelial membrane antigen, Ki-1 (CD30), AFP, and hCG.[1,5] The transcription factor OCT3/4 is a robust diagnostic marker for seminoma as well as embryonal carcinoma. Recent studies have reported that podoplanin, recognized by the commercially available D2-40 monoclonal antibody,[6] is a specific marker for seminoma because unlike OCT3/4, it is not expressed by embryonal carcinoma. Primary EGCTs are rare, and their existence is still met with caution. The possibility that retroperitoneal tumor represents a metastatic site from a clinically occult testicular primary source should be ruled out.[7,8] Nowadays, with the availability of modern scrotal ultrasound, the likelihood of missing a testicular abnormality is markedly reduced.

Primary extragonadal germ cell tumors should be differentiated from the 'burned out' phenomenon in germ cell tumors.[7] This condition is defined as the presence of an extragonadal germ cell tumor with no evidence of neoplasm at the testis, where a series of distinctive histological lesions, indicative of the earlier presence of a completely regressed testicular malignancy, can be detected.[7] It is important to distinguish true extragonadal germ cell tumors from burned out tumors of the testis because the primary removal of the testicular tumor is essential for a satisfactory outcome.

Chemotherapy is the treatment of choice for patients with bulky retroperitoneal involvement and all stage III cases result in a complete response rate of approximately 90%. Despite several recognized risk factors in the development of germ cell tumors (GCTs), (e.g. cryptorchidism or a prior history of GCT), the pathogenesis of germ cell neoplasms including the contributing role of environmental factors or genetic susceptibility remains unknown.[5] During the migration to the scrotum, some primordial germ cells from the urogenital ridge are left behind during embryonic development and this embryologic theory can explain this phenomenon.[3] The therapeutic outcome for patients with extragonadal germ cell tumors is similar to that for patients with very advanced testicular cancer.[5] If the history and physical examination as well as a testicular sonogram are completely normal, it means that there is no history of cryptorchidism, testicular atrophy and palpable mass. So, attention should be turned to the retroperitoneal mass as a likely primary site. Otherwise, testicular biopsy is mandatory. It also suggests that all cases with these tumors should be screened for the presence of clinical features of Klinefelter's syndrome.[8]

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