Primary Retroperitoneal Mucinous Tumor of Low Malignant Potential in a Persian Woman

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Abstract- Primary retroperitoneal mucinous tumor (PRMT) of low malignant potential (border line) is an uncommon neoplasm with fewer than 50 reported cases. Uncertain diagnostic imaging results make diagnosis of its origin difficult, preoperatively. Later treatment planning and prognosis would be affected by exact diagnosis of the tumor origin. This study presents a case of Persian woman with diagnostic, histological and immunohistochemical specifications.

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Keywords: Primary retroperitoneal mucinous tumor; Low malignant potential; Borderline

Introduction

Primary retroperitoneal mucinous tumor (PRMT) of low malignant potential (borderline) is a rare neoplasm. Till now, less than 50 cases of PRMT have been reported in English literature. Almost always, the reported patients were females (1). PRMT has the same gross and histomorphologic characteristics than the ovarian or pancreatic mucinous tumors (1). So the major issue of this condition is how to distinguish the exact origin of the tumor preoperatively; because it can change the treatment planning, survival and prognosis (1). As far as we know, similar cases reported in Iran and Middle East region very rarely.

Case Report

A 26-year-old woman with unremarkable medical, surgical and familial histories, complaining of abdominal distention and abdominal pain of few months duration was referred to Cancer Institute of Tehran Imam Khomeini Medical Complex. On clinical examination a large mass at left lower quadrant (LLQ) of the abdomen was detected. Primary abdominopelvic CT scan show pelvic mass measuring about 13*10*8cm with extension to LLQ with no evidence of pancreatic or ovarian origin of the mass (Figure 1). Serum CEA, α-FP, CA19-9 and CA125 were in normal limits and was 3.5 U/ml, 5.3 U/ml, 35.17 U/ml and 20 U/ml, respectively.

A laparotomy was performed and retroperitoneal mass in LLQ, without attachment to other organs, completely resected; No significant changes in other abdominal or pelvic organs such as the pancreas were seen.

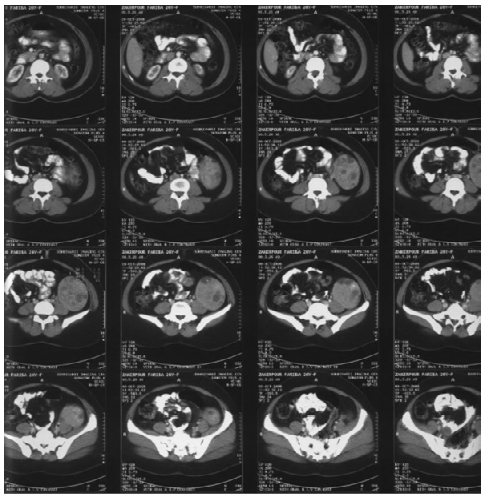

Figure 1. Triple-contrast abdominopelvic CT scan shows a large heterogeneous mass at LLQ of abdomen.

Pathological findings

Gross examination of the received specimen revealed an ovoid shape encapsulated creamy-grayish piece of tissue measuring 12*10*7cm in diameters. Capsular surface was unremarkable, but cut sections showed heterogeneous solid and cystic areas with pasty greenish materials in some foci. Microscopic examination shows

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multiloculated and capsulated mucinous tumor with mild to moderate nuclear atypia. In some areas proliferating, epithelium shows epithelial bridging and delicate papillary structures with filigree pattern. There is no evidence of stromal invasion in prepared slides (Figures 2,3).

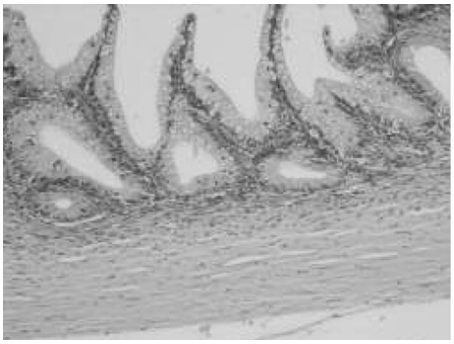

Figure 2. Papillary structures lined by mucin containing cells; H&E, x100

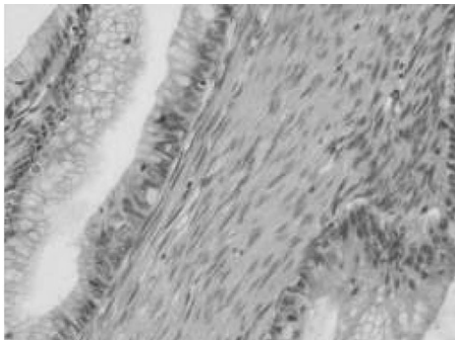

Figure 3. Mild to moderate nuclear atypia of epithelial cells; H&E, x200

Immunohistochemistry findings

In order to elucidate the exact origin of the tumor IHC study was performed. Results were positive for Cytokeratin(CK) 7, CK20, Ki-67 (about 40%) and Carcinoembryonic Antigen (CEA) and negative for Estrogen Receptor(ER) and progesterone receptor (PR). (Figures 4 - 9, respectively).

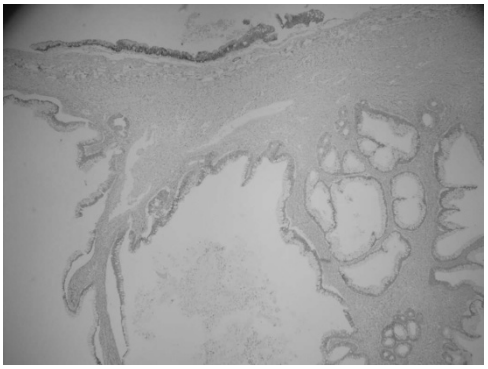

Figure 4. IHC study shows positivie reaction for CK7

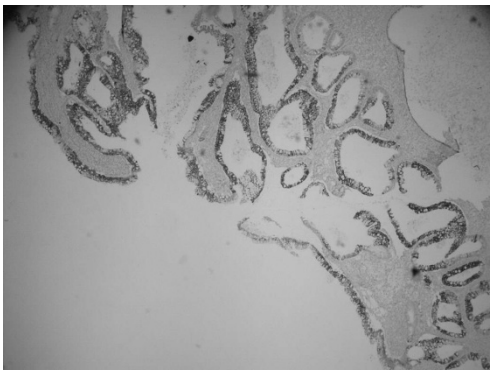

Figure 5. IHC study shows positive reaction for CK20

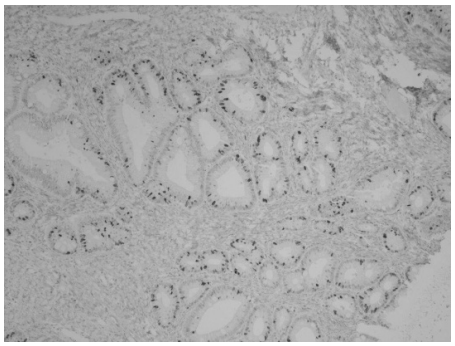

Figure 6. IHC study shows positivie reaction for Ki-67

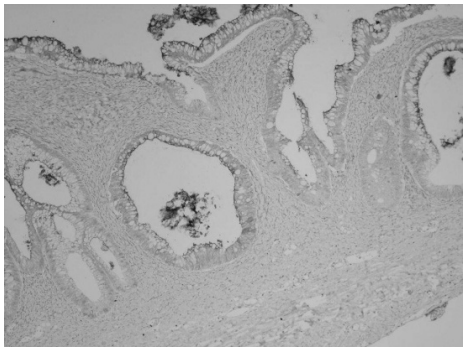

Figure 7. IHC study shows positivie reaction for CEA

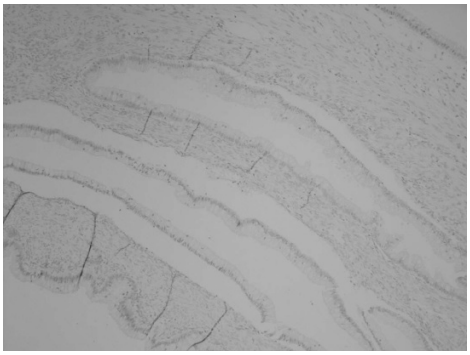

Figure 8. IHC study shows negative reaction for ER

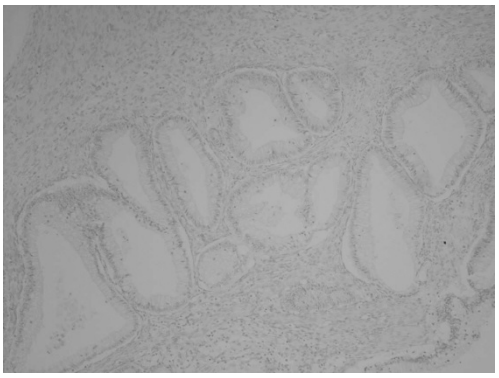

Figure 9. IHC study shows negative reaction for PR

Discussion

Prevalence of primary retroperitoneal tumors is about 0.01-0.2% of all neoplasms (2,3). Since most of these tumors show no significant symptoms, delayed diagnosis is common. The more common primary tumor of retroperitoneum includes soft tissue tumor such as liposarcoma and leiomyosarcoma, germ cell tumor and tumors of sympathetic nervous system such as neuroblastoma or ganglioneuroblastorna and tumors of Müllerian type. The latter consisting of primary retroperitoneal mucinous cystadenomas, mucinous cystadenomas of borderline malignancy and cystadenocarcinomas, is extremely rare (4).

Reviewing the literature from 1966 till 2006 by Baker et al revealed 45 cases of primary retroperitoneal mucinous cystadenomas and 25 cases of primary retroperitoneal mucinous cystadenocarcinoma (5). Seven cases of primary retroperitoneal mucinous cystadenoma of borderline malignancy were identified in a retrospective analysis of 18 retroperitoneal mucinous tumors by Roma *et al.,* (6).

Epithelial components do not exist in the retroperitoneal region. Hence, tumors derived from epithelial cells are rare in this area. Despite that, some Müllerian type epithelial tumors in the retroperitoneal region are reported by Roth *et al.,* (7).

Since PRMC shows identical histological findings to the mucinous tumors of the ovaries, the theory that tumor's origin is an ectopic or aberrant ovarian tissue seems to be more reliable. However, ovarian tissue has been found histologically in these tumors rarely (8). Other theories include the presence of potential of Müllerian differentiation in the peritoneal epithelium, overgrowth of mucinous epithelium on teratoma or genitourinary remnants (9,10) and mucinous metaplasia of the coelomic mesothelium (11), and some have shown evidence of gastric mucosal differentiation, suggesting a totally different histogenesis (12).

Nonetheless, origination of these mucinous tumors from the retroperitoneal entrapping of multipotential mesothelial cells during the development is agreed by most of the authors.

Non-specific presenting symptoms of retroperitoneal mucinous tumors such as abdominal discomfort or distention make the preoperative diagnosis difficult (2,3,13,14). Although diagnostic imaging by ultrasound (US), computed tomography (CT) or magnetic resonance imaging (MRI) can identify the retroperitoneal masses especially whether organ displacement caused by the mass is present, the exact origin of them remains vague (13,15).

US have low diagnostic value in the determination of origin or extension of these tumors, but MRI, and particularly CT results are more valuable. CT can reveal the mural calcification and extension of the mass better. Former can be assumed as a key finding in differentiation cystadenoma from cystic teratoma. The latter shows more calcification within the mass rather than cyst wall (16,17). On the other hand, MRI is more helpful to determine the correlation between the mass and adjacent organs (13,15,17).

First, our case was detected by US. Then performed CT just verified that there was no correlation between the mass and the pancreas but not any more data. Checked tumor markers including serum CEA, α-FP, CA19-9 and CA125 were within normal limits before the surgery and the patient underwent diagnostic laparotomy.

Since rare, similar cases reported around the world, and the preoperative diagnosis is difficult in current hospital diagnostic settings, laparotomy could be the standard approach for both diagnosis and treatment in these patients (18).

Assessment of prepared slides of our tissue sample revealed the histopathological findings as discussed earlier; an ovarian-like mucinous tumor with low malignant potential. Thus, more investigations to determine the exact origin of the tumor (primary vs. secondary tumor) were done by IHC; Positive reactions for CK7, Ck20 and CEA, are rather the same as the results of IHC panel of ovarian mucinous tumors, but negative reactions for ER and PR ruled out the ovarian origin of the tumor. On the other hand, CK7 positivity is out of favor of gastrointestinal origin (19). Furthermore, pancreatic origin of the tumor excluded radiologically. In addition to histopathological findings, Low reaction of Ki-67 is in favour of borderline nature of the tumor rather than carcinoma. Correlation of histopathological,

IHC findings, clinical and imaging results led us to eliminate other origins of the tumor rather than retropeitoneum itself.

Although the behaviour of these tumors generally is similar to the ovarian counterparts, but there is a case report of one otherwise typical borderline tumor metastasized four years after its removal (20).

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