



Management of Cervical Lymph Nodes Metastasis of Nonseminomatous Extra Gonadal Carcinoma - Report of a Rare Case

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Abstract

Presence of supraclavicular lymph node metastasis as first symptoms in retroperitoneal Extra Gonadal Germ Cell Tumor (EGGCT) is not common but this phenomenon has been well recognized and does not mean inoperable stage of disease. While there is limited literature on the management of cervical lymph node metastases in retroperitoneal EGGCT, we present our case who underwent selective neck dissection for post-chemotherapy residual disease. 6 months evaluation revealed no finding of recurrence in imaging modalities and also the level of tumor markers.

Keywords: Extra-gonadal germ cell tumor; Nonseminomatous tumor; Lymph node metastasis; Selective neck dissection

Introduction

A germ cell neoplasm who is originated outside of the gonads is named an Extra Gonadal Germ Cell Tumor (EGGCT) [1,2]. 5% of the malignant germ cell tumors are of extra gonadal origin [3]. After mediastinum, retroperitoneum is the most common site of EGGCT. Like testicular carcinoma, in EGGCT, although supraclavicular lymph node metastasis is not common, this phenomenon has been well recognized and despite other infra diaphragmatic malignancies does not mean inoperable widespread disease [4-6].

Although the incidence of neck lymph node metastases in testicular tumors range from 4.5% to 15%, in retroperitoneal EGGCT the amount is higher and is about 18% [1,5,7,8]. While there is limited literature on the management of cervical lymph node metastases in retroperitoneal non-seminoma EGGCT, we present our case to evaluate the role of selective neck dissection in the treatment of metastatic lymph nodes which be performed for post-chemotherapy residual disease.

Case Presentation

The patient was a 36 year-old male complained from a left supraclavicular mass. He felt mild fatigue, low back pain and abdominal discomfort from 2 months ago. The neck mass was about 4 centimeters in great diameter, fixed, painless and behind the sternocleidomastoid muscle in the left supraclavicular fossa. Imaging (CT scan and ultrasonography) showed well defined mass measuring 33 mm × 17 mm at the left lower neck along the margin of trapezoid muscle and anterolateral to brachial plexus without obvious internal vascularity which suggest metastatic lymph node. In addition, there were several para aortic and retroperitoneal pathologic lymph nodes. Core needle biopsy of the neck mass was done and after microscopic and immunohistochemistry evaluations and combination of those data with elevation of germ cell tumor markers, diagnosis of metastatic germ cell tumor was approved. Ultrasonography of testicles was normal. Spiral chest CT scan realized no lung or mediastinal metastasis. Evaluating of all these data concluded that we faced a primary retroperitoneal non-seminoma cancer patient who presented with neck metastasis.

He was treated by four course of BEP chemotherapy (cisplatin, etoposide and bleomycine) which although disappeared all signs and symptoms, in imaging multiple small nodes were seen in retro peritoneum and left lower neck. The patient underwent Retro Peritoneal Lymph Node Dissection (RPLND) and neck selective lymph node dissection (level III, IV and V) for residual disease 6 months ago which histologic examination revealed mature teratoma. The physical examination and tumor markers were normal during this period of time.

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Discussion

Although Testicular cancers (98% are germ cell tumor) include only one percent of all malignancies in men, they are the most common neoplasms in the age between 15 to 34 years [8]. The median age of onset for retroperitoneal non seminoma EGGCTs is 30 years which is similar to primary gonadal.

Germ cell tumors divided to gonadal and extra gonadal (EGGCTs) which EGGCTs are approximately 2% to 10% of all germ cell tumors [9-12]. The most common site of EGGCTs are mediastinum and retro peritoneum. However, it can also be seen in sacrococcygeal region, central nervous system, prostate, orbital cavity, liver, vagina, and gastrointestinal tract [1,9,10,13,14]. Although in some cases of EGGCTs the imaging of testicles or histologic examination of testis needle biopsy realize a scar in the testis which representing a 'burn-out' or 'auto infarcted' primary tumor and based on histological, serological and cytogenetic characteristics of EGGCTs we can say that they are similar to testicular germ cell tumors, their clinical behaviors are differ [1,12,15]. Like primary gonadal germ cell tumors, the common tumor marker expression of these tumors includes α -fetoprotein (AFP), β human chorionic gonadotropin (β -HCG) and LDH [1,9,14]. Evaluating these tests after chemotherapy completion and also during the closed observation period after retroperitoneal and neck dissection, has been in normal range in our patient.

Because of small grow up of primary retroperitoneal non seminomatous germ cell tumors; they may be very large without overt symptoms [9]. Common sign and symptoms of these tumors are abdominal (29%) or back pain (14%), weight loss (9%), fever (8%), vein thrombosis (9%), palpable abdominal mass (6%), cervical nodes (4%), scrotal edema (5%), gynecomastia and dysphagia (3%) [1,9,16]. The symptoms of our patient were fatigue, low back pain and cervical mass which Adhered from classic presentation. Like our case, 76% for retroperitoneal non seminomatous EGGCTs represent with metastasis at diagnosis time [1,9]. The most common site of metastasis are lungs (49%), intra-abdominal lymph nodes (34%), cervical lymph nodes (18%) and liver (25%) [1,8,9,16,17]. The pathway of tumor to reach the cervical nodes without involvement of the mediastinum is not recognized well. Although some researchers believe that this type of metastasis is hematogenous, the majorities indicate that the metastatic cells spreads to the junction of internal jugular and subclavian veins via thoracic duct and from that location it may spread to cervical lymphatic nodes [4,18,19]. Based on this explanation, like our patient, neck metastases are almost always in the left-side of neck [18].

The rate of metachronous testicular cancer in EGGCT patients is about 10% over 10 years which confirm that excluding of a testis primary tumor is by sonographic investigation alone and performing a routine bilateral testicular biopsy of patients with EGGCT is not necessary [1,9,12,20-23]. The ultra sonographic evaluations of testis in our patient in the early stage of diagnosis and during the closed observation were normal, up to now.

Residual tumors after chemotherapy (4 courses of BEP chemotherapy) in non seminomatous retroperitoneal EGGCT must be extracted surgically, because in 25% of patients the histologic examination revealed viable undifferentiated tumor and also there is always some risk of eventual reversion of residual teratoma to its malignant counterpart [1,7,9,24,25]. Based on this conception, we performed Retroperitoneal Lymph Node Dissection (RPLND) for

our patient and the final histologic examination revealed only mature teratoma in the residual tissues. In addition, behavior around this rule is logical for neck metastasis and neck dissection has a demonstrated role in the treatment of these conditions which we did it by a selective neck dissection. Since generally it is accepted that neck metastasis in retroperitoneal non-seminoma tumors is via thoracic duct, it seems that dissection of neck nodes which are located or near the insertion of thoracic duct to the subclavian vein (level III, IV and V) are enough to control the local disease and expanding this to the other neck levels and/or mediastinum should be considered in patients with metastasis in those sites [5,7,18,24,26]. This procedure provides excellent local control of the neck disease by eradicating microscopic metastases. The only contraindication for surgical clearance of node metastasis is the presence of persistently elevated serum tumor markers after chemotherapy. These patients are considered non-responder and it is recommended to manage by second-line salvage chemotherapy [18,27].

Unlike other infradiaphragmatic cancers, the cervical lymph node metastases in EGGCT are curable if appropriate treatment strategy is designed [6,8]. 5-year survival rate of Nonseminomatous retroperitoneal EGGCTs is 62% that is worse than seminomatous EGGCTs [1,9,12,16].

We conclude that, performing selective neck dissection in neck node metastasis from retroperitoneal non-seminoma tumors after completion of chemotherapy is effective with acceptable complications. Performing extensive operations have more probable complications and are not necessary for local control.

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