J Rural Med 2011; 6(1): 32–34

Case report

Seminomatous Extragonadal Germ Cell Tumor with Complete Obstruction of the Superior Vena Cava Responding to Intensive Chemotherapy

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Abstract

A 20-year-old man was admitted to our hospital with persistent cough and dyspnea. He had bilateral distention of the jugular veins, and swollen lymph nodes were palpable in the right subclavicular region. Plain X-ray and computed tomography (CT) of the chest showed a solid soft tissue mass in the upper mediastinum, with leftward displacement of the trachea and complete obstruction of the superior vena cava. Mediastinal radiotherapy (1.8 Gy/day) and methylprednisolone (100 mg/day) were started immediately. Biopsy of the right subclavicular lymph nodes revealed metastatic seminoma. The patient was referred for chemotherapy, which was performed with a combination of cisplatin, bleomycin and etoposide (BEP). A partial response was observed after completion of 3 cycles of chemotherapy, but there was no further tumor shrinkage after additional salvage chemotherapy. The patient is being followed up on an outpatient basis and has been free of recurrence for 32 months after intensive treatment.

Key words: Extragonadal germ cell tumor, intensive chemotherapy, superior vena cava obstruction

(J Rural Med 2011; 6(1): 32–34)

Introduction

Extragonadal germ cell tumors (EGGCTs) have the same histological components as their gonadal counterparts, but a gonadal mass is not detectable by palpation or high-resolution ultrasonography[1]. It has been estimated that only 2–5% of all germs cell tumors are extragonadal. Primary mediastinal seminomas, in contrast, comprise about 25% of all primary germ cell tumors (GCTs)[2]. They are frequently associated with extensive local invasion, and chemotherapy has been reported to achieve a favorable outcome as initial treatment[1,2]. In a minority of patients, the primary tumor is located extragonadally, i.e., in the retroperitoneum or in the mediastinum. About one-third of these patients will harbor intratubular germ cell neoplasia (TIN) (synonym: carcinoma in situ, CIS). In another one-third, ultrasonography of the testes reveals scar tissue indicating burned out testicular tumors, which also have to be removed. Therefore, only one-third of these patients have definitive primary EGGCTs[3]. The most common primary sites for EGGCTs are the mediastinum and the retroperitoneum, while these tumors have also been reported to occur in the sacrococcygeal and pineal regions[2]. However, management of residual seminomatous GCTs after chemotherapy remains an issue needing clarification.

Here, we report a case of primary mediastinal seminoma with complete obstruction of the superior vena cava and no recurrence of the residual mass after intensive chemotherapy.

Case Report

A 20-year-old man was admitted to our hospital with persistent cough. His bilateral jugular veins were distended, and swollen lymph nodes were palpable in the right subclavicular region. A chest X-ray showed a mediastinal mass. Computed tomography (CT) of the chest revealed a solid soft tissue mass (95 × 75 × 112 mm) [in diameter] in the upper mediastinum, as well as a leftward displacement of the trachea and complete obstruction of the superior vena cava (Figure 1). Based on the suspicion of a mediastinal tumor with lymph node metastasis, mediastinal radiotherapy and methylprednisolone were immediately started. Biopsy of the right subclavicular lymph nodes revealed metastatic seminoma (Figure 2), and the patient was referred for chemotherapy.

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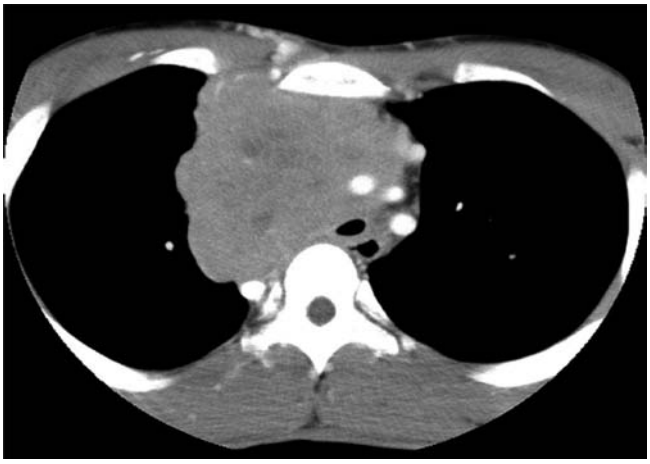

Figure 1 Pretreatment CT shows a solid soft tissue mass (95 × 75 × 112 mm) in the upper mediastinum, with leftward displacement of the trachea and complete obstruction of the superior vena cava.

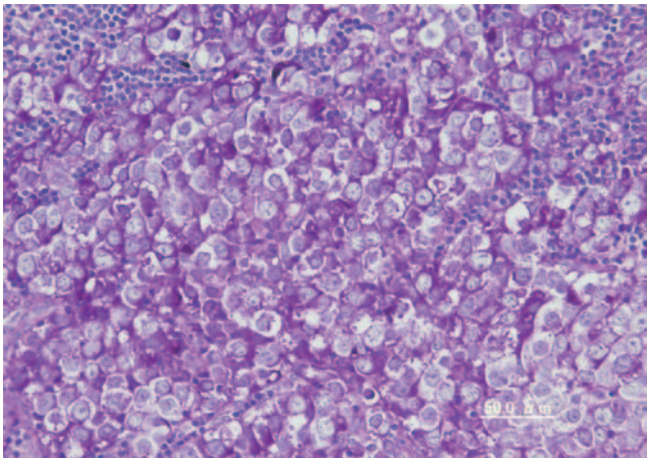

Figure 2 Biopsy specimen from a right subclavicular lymph node. Well-defined tumor cells with large nuclei and clear nucleoli and a large clear reticulum are formed as map-like alveolar tumor cells. Cobblestone growth can be seen with lymph node involvement surrounding the alveolar tissue.

Lactate dehydrogenase, β-human chorionic gonadotropin and alpha-fetoprotein were all in the normal ranges. Ultrasound revealed a uniform, echogenic structure of the bilateral testicles, without any findings suggestive of malignancy.

Radiotherapy and methylprednisolone were discontinued, and systemic chemotherapy was simultaneously started with a combination of cisplatin, bleomycin and etoposide (BEP). Since lung function test values deteriorated after one cycle of BEP, bleomycin was excluded from subsequent chemotherapy, and the regimen continued with etoposide plus cisplatin (EP). A partial response was observed after completing 3 cycles, but no further shrinkage was observed on subsequent examinations.

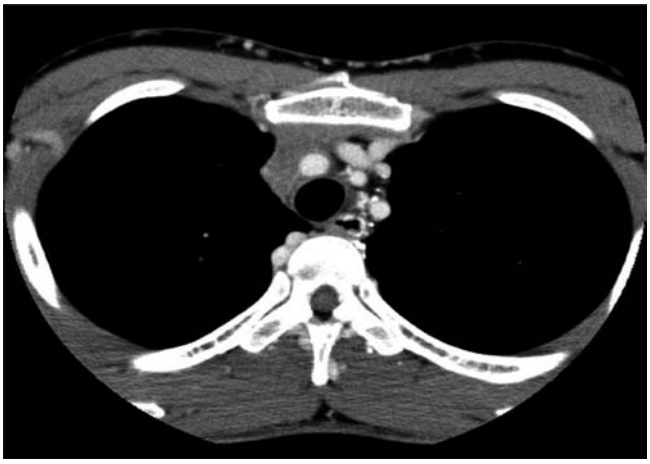

Figure 3 Posttreatment CT shows less enhancement of a smaller soft tissue mass (30 × 18 × 36 mm).

Salvage surgery was then considered for the residual mass because the superior vena cava was still completely obstructed by the tumor. Although implantation of a prosthetic vessel was required, the procedure was not performed because it was considered to be a high risk surgery. Instead, the patient received 2 courses of vinblastine, ifosfamide and cisplatin (VeIP) as salvage chemotherapy. On completion of the chemotherapy, the evaluable lesions were considered to represent a "stable disease" (Figure 3). As of December 2010, there is no evidence of residual tumor growth, new masses or metastatic foci. The patient is being closely followed up on an outpatient basis and has been free of recurrence for 32 months after intensive treatment.

Discussion

Our patient was first diagnosed as having an EGCT arising from the mediastinum when he presented with chronic cough and complete obstruction of the superior vena cava.

Since few patients with early germ cell tumors arising in the mediastinum have any symptoms, these tumors are often quite large by the time they are diagnosed and the mean maximum diameter of such tumors is reportedly 4.6 cm[2]. Seminoma accounts for about 25% of EGGCTs[4]. When patients with seminoma or nonseminomatous tumors are treated with platinum-based chemotherapy (cisplatin or carboplatin) and radiotherapy, the 5-year progression-free survival rate is 80% for seminoma versus 35% for nonseminomatous

tumors[5]. According to the International Germ Cell Cancer Collaborative Group (IGCCCG) criteria, primary mediastinal seminoma without any nonpulmonary visceral metastases is classified as a good risk tumor with favorable sensitivity to chemotherapy and radiotherapy, and these patients have progression-free survival similar to those with primary testicular seminoma. However, treatment of the residual mass differs between these two groups. For primary mediastinal tumors, all residual radiological lesions should be resected, if possible[3], and the presence or absence of viable tumor cells should be determined.

In 2008, Kesler *et al.* reported that 10 surgical deaths occurred among 158 patients who underwent additional resection of residual primary mediastinal nonseminomatous tumors[6]. Kang *et al.* also reported in the same year that marked fibrosis and surrounding invasion of residual tumor cells were detected in the majority of 29 patients who underwent surgery for residual nonseminomatous tumors[7]. They also reported that it may be very difficult to remove the tumor completely, particularly if it has invaded the great vessels. In our patient, salvage surgery was considered for the residual mass after chemotherapy, but the superior vena cava was completely obstructed by the tumor, suggesting that the operation constituted a high risk. In addition, residual tumor cells were suspected to have invaded the surrounding mediastinal tissue, adding further evidence for an unlikely successful operation.

Like primary testicular seminoma, primary mediastinal seminoma has a good prognosis, and the rate of necrosis of the residual mass is as high as 92% when partial remission is obtained after chemotherapy[2]. When the residual mass is considered inoperable in patients with primary mediastinal seminoma, additional salvage chemotherapy can be considered for careful follow-up of patients after initial chemotherapy as described in this report.

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