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Case Report

Clinical Presentation of Advanced Extragonadal Embryonal Carcinoma Mimicking Classical Hodgkin Lymphoma

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Abstract

A 22-year-old man presented with a 2-week history of abdominal pain, fatigue, fever, and night sweats. Enlarged neck and paraaortic lymph nodes were noted, but no testicular lesions were found using imaging tools. Pathology revealed poorly differentiated embryonal carcinoma (EC) of the metastatic neck lymph node. The patient presented with clinical symptoms similar to Hodgkin lymphoma (HL), but the final diagnosis was advanced-stage extragonadal EC. To our knowledge, this is a rare case report of EC presenting as classic HL.

Keywords: Embryonal carcinoma, extragonadal, Hodgkin lymphoma

Introduction

In general, embryonal carcinoma (EC) occurs at a young age, in the second to third decades on average.[1] EC is aggressive and results in early hematogenous spread. Approximately 66% of patients with EC have metastasis at the time of diagnosis.[2] It can be difficult to differentiate from lymphoma, with both diseases presenting with lymphadenopathy. Early histological proof and quick treatment are important factors for disease outcomes.

Case Report

We present the case of a 22-year-old male patient who was admitted to our hospital with abdominal pain and fatigue. He had no underlying diseases or operation history. A clinical examination revealed a left supraclavicular fossa mass without painful sensation. No aberrant palpable masses over the scrotum or the inguinal canal were noted. Blood test results were normal (white blood cell count: 9.8 × 10^9/l, Hgb: 13.7 g/dl, blood creatinine: 0.8 mg/dl, C-reactive protein: 0.9 mg/dl). However, serum tumor markers were elevated: alpha-fetoprotein (αFP) (26 ng/ml), free human chorionic gonadotrophin (hCG) (0.17 ng/ml), lactate dehydrogenase (LDH) (485 U/l).

Abdominal contrast-enhanced computed tomography (CECT) showed weakly enhancing soft-tissue lesions in the paraaortic

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space [Figure 1]. Splenomegaly was also noted. Neck CECT showed a mass lesion in Level III and also in some small lymph nodes in the bilateral neck [Figure 2]. Scrotal sonography showed no significant varicocele or abnormal findings. Positron emission tomography (PET) showed an enhancing signal over the right subclavicular node and a paraaortic mass lesion [Figure 3a].

An excisional biopsy of the neck mass was done, and a histopathological examination showed poorly differentiated carcinoma. Immunohistochemistry staining was positive for cytokeratin (CK), CK7, CD30, sal-like protein 4, and octamer-binding transcription factor 3/4 (OCT 3/4) [Figure 4]. Metastatic EC of the neck mass was diagnosed. According to the American Joint Committee on Cancer staging system, cT0pN2M1aS2, Stage IIIB was diagnosed.

Following the NCCN guidelines, systemic chemotherapy with bleomycin, etoposide, and cisplatin was carried out at full dose for four courses. A follow-up PET showed a partial response [Figure 3b]. Further surgical interventions were needed, but the patient has so far refused.

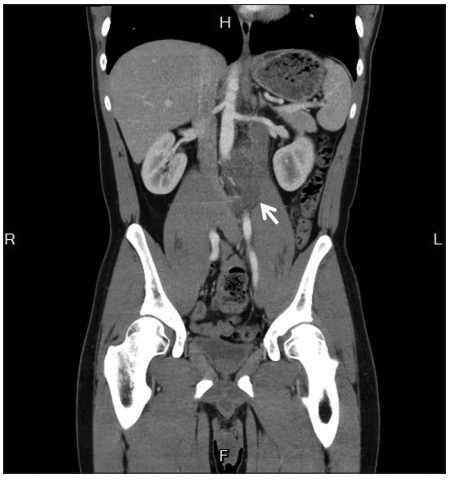

Figure 1: Abdominal contrast-enhanced computed tomography showed weakly enhancing soft tissue lesions in the paraaortic space (white arrow)

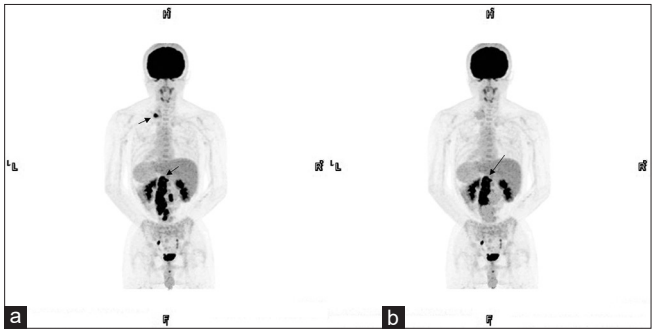

Figure 3: (a) Positron emission tomography scan before treatment showed an enhancing signal over the right subclavicular node and paraaortic mass lesion (black arrow). (b) Positron emission tomography scan after four courses of bleomycin, etoposide, and cisplatin showed a residual enhancing lesion over the paraaortic region (black arrow)

Discussion

EC is the second most common histological type of testicular tumor after seminoma.[3] In general, it occurs at a young age.[1] Germ cell tumors (GCTs) originate in the gonads in most clinical cases. Extragonadal germ cell tumors (EGGCTs) commonly arise in the midline of the retroperitoneum or the mediastinum but are not common.[4] Only approximately 2%–5% of all GCTs are of extragonadal origin.[5] The overall incidence ranges from 1.8 to 3.4/1 million.[6] According to this low incidence and nonspecific clinical features, diagnosing EC is a challenge, especially to differentiate it from Hodgkin lymphoma (HL).

The clinical presentations of EGGCTs depend on the location and size of the tumor. The symptoms of mediastinal GCTs include dyspnea, chest pain, fever, cough, and hemoptysis. In retroperitoneal GCTs, abdominal pain, lumbago, backache, and body weight loss are usually noted.[7]

Clinical symptoms and signs of HL in young adolescents include lymphadenopathy, systemic complaints, and

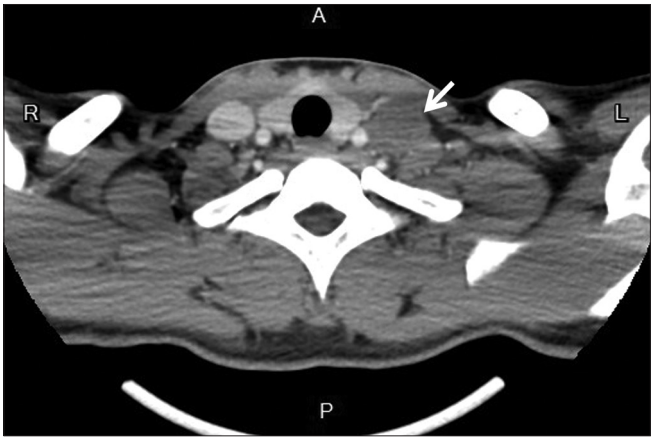

Figure 2: Neck contrast-enhanced computed tomography showed a mass lesion over the left neck (white arrow)

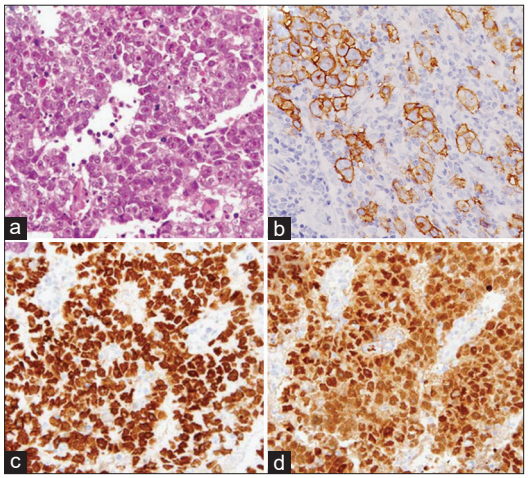

Figure 4: Immunohistochemistry staining. (a) H and E stain, (b) CD30, (c) Sal-like protein 4, (d) Octamer-binding transcription factor 4

mediastinal mass.[8] Patients may present with symptoms including fatigue, anorexia, and weight loss. Other symptoms such as fever and night sweats are also common.[9] Both EC and HL have similar clinical presentations, however, they differ in primary location (i.e. the testis for EC but not for HL). It is difficult to differentiate quickly when extragonadal EC occurs.

Serum tumor markers include αFP, β-hCG, and LDH, which are a common tool for diagnosing EGGCTs. In EC, serum tumor markers including αFP, β-hCG, and LDH may be only slightly elevated and are thus hard to detect initially.[10]

As CD30 immunostaining is often observed in EC and also in a variety of lymphomas (e.g. HL, diffuse large B-cell lymphoma, and anaplastic large-cell lymphoma), CD30 should not be used as a marker to distinguish between EC and lymphomas.[11] The HL H and E stain may mimic EC closely because of its sheet-like growth pattern. In contrast to HL, CK immunostaining should highlight EC, and OCT4 may be used as a marker of either EC or seminoma.[12]

An early confirmatory diagnosis of EC is advantageous because of the high risk of lymphatic and vascular invasion, as well as the increased possibility of spread into the paratesticular tissue in EC.[13]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal the identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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