

Case Report

Computed tomography imaging features of primary extragonadal seminoma-nine cases report and literature review

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Received July 26, 2016; Accepted June 20, 2017; Epub August 15, 2017; Published August 30, 2017

Abstract: Primary extragonadal seminoma is relatively rare, which belongs to primary extragonadal germ cell tumor. Up to date, only a small number of literatures have been reported about its Computed Tomography (CT) features. In the present study, we reported 9 cases of primary extragonadal seminoma proved by histopathology. This report aims to demonstrate the CT features of infrequently occurred primary extragonadal seminoma. Based on the existing literatures and our 9 cases, we make a conclusion that primary extragonadal seminoma usually occurs in young or middle aged adults with no specific clinical manifestations, and CT is of great diagnostic and clinical values for this kind of tumor according to the configuration, size, density, style of enhancement and surrounding infiltration of the lesions, but the final diagnosis depends on pathology.

Keywords: Computed tomography, primary extragonadal seminoma, case report, literature review

Introduction

Primary extragonadal germ cell tumors are relatively rare, comprising 5% to 10% of all germ cell tumors [1]. They are identical histologically to their testicular counterparts and, thus, can be divided into seminomatous tumors and non-seminomatous tumors. Pure seminomatous tumors are extremely rare, and majority of reports regarding such tumors are case reports and do not enable the summary of their characteristic CT imaging findings [2-5], which leads to great difficulty of preoperative diagnosis. Hence, we retrospectively reported the CT imaging findings of 9 patients with primary extragonadal seminomas and reviewed the literature, so as to raise the awareness of this tumor.

Case presentation

Nine cases from April of 2008 to May of 2014 were retrospectively studied. The physical and ultrasonic examinations of all the patients' testes were normal.

All of the patients underwent non-enhanced and contrast-enhanced CT examinations simultaneously. CT imaging was performed using a Siemens SOMATOM Definition 64 spiral CT machine, with the following parameters: 5 mm slice thickness, 120 KV voltage, 250 mA current, and 256×256 matrix. After non-enhanced CT scan, an intravenous bolus dose of 100 mL of nonionic iodinated contrast agent (iopromide; Ultravist; Schering) was administered at a rate of 3.5 mL/s. Enhanced CT images were obtained at 60 s after the contrast agent injection.

Two experienced radiologists evaluated the CT images, including the location, size, shape, edge, CT attenuation on non-enhanced and contrast-enhanced CT images, involvement of adjacent structure, and local or distant metastasis of each lesion.

This retrospective study was an audit of existing practice and did not require local ethical and research committee approval.

CT features of primary extragonadal seminoma

Table 1. Clinical features of 9 patients with primary extragonadal seminoma

Case	Sex	Age (year)	Location	Clinical manifestation	Maximum diameter (cm)
1	Male	28	Anterior mediastinum	Chest pain and dry cough for 1 year	5.1
2	Male	35	Anterior mediastinum	Fever for 1 week	12.8
3	Male	29	Anterior mediastinum	Asymptomatic	10.8
4	Male	31	Anterior mediastinum	Asymptomatic	7.8
5	Male	32	Retroperitoneum	Stomachache, Progressive nausea and vomiting for 11 days, anuria for 24 hours.	8.5
6	Male	62	Retroperitoneum	Asymptomatic	10.8
7	Male	28	Pelvis	Asymptomatic	12.2
8	Male	30	Pelvis	Asymptomatic	10
9	Male	67	Pelvis	Asymptomatic	8.4

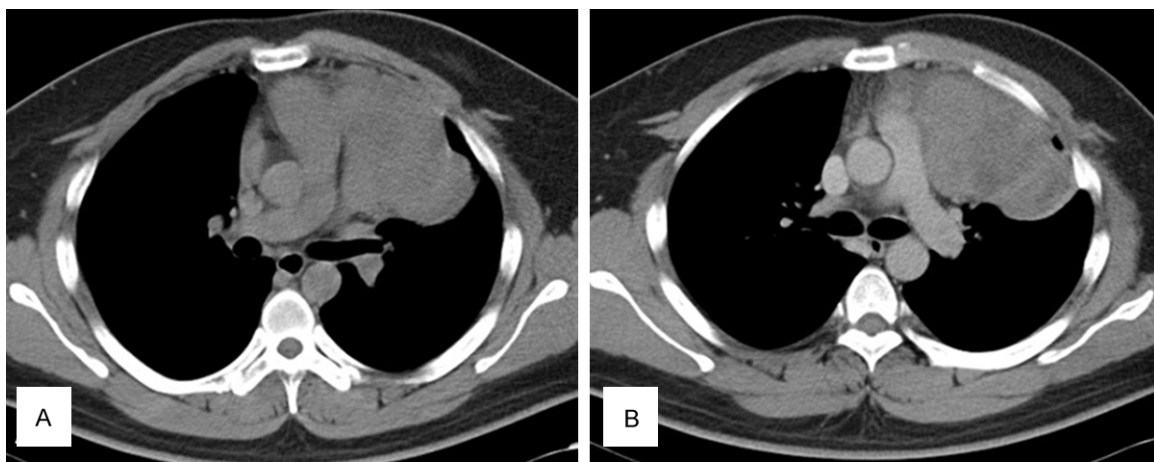


Figure 1. 28-year-old male with seminoma in the mediastinum. A. Non-enhanced CT image shows a bulky, lobulated mass with heterogeneous attenuation. B. The mass reveals moderately heterogeneous enhancement after contrast administration. And the mass compresses the pulmonary artery, the adjacent chest wall and pericardium was involved.

Clinical data

The clinical data of the 9 patients are summarized in **Table 1**. The clinical manifestations were different with its different locations.

CT findings

Four cases arose within the anterior superior mediastinum, and the maximal diameter of the biggest one was 12.8 cm. 3 masses were lobulated and ill-circumscribed, which showed heterogeneous attenuation on non-enhanced CT with patchy low-attenuation foci, and presented heterogeneous enhancement after contrast administration. Moreover, adjacent chest wall and pericardium involved were observed (**Figure 1**). The mediastinal vessels, such as pulmonary artery were compressed. 1 mass

was oval-like and well circumscribed, that presented homogeneous attenuation on non-enhanced CT and showed lightly homogeneous enhancement after contrast administration (**Figure 2**).

Two cases arose in the retroperitoneum, the maximal diameter of the bigger one was 10.8 cm. The masses were lobulated with intratumoral necrosis, presented infiltrating growth, pattern with the abdominal aorta, renal artery and renal vein being wrapped and compressed. One of the mass compressed the left ureter, causing moderately hydronephrosis of the left kidney (**Figure 3**), with adjacent lymph nodal involved. The two masses showed heterogeneous enhancement after contrast administration.

CT features of primary extragonadal seminoma

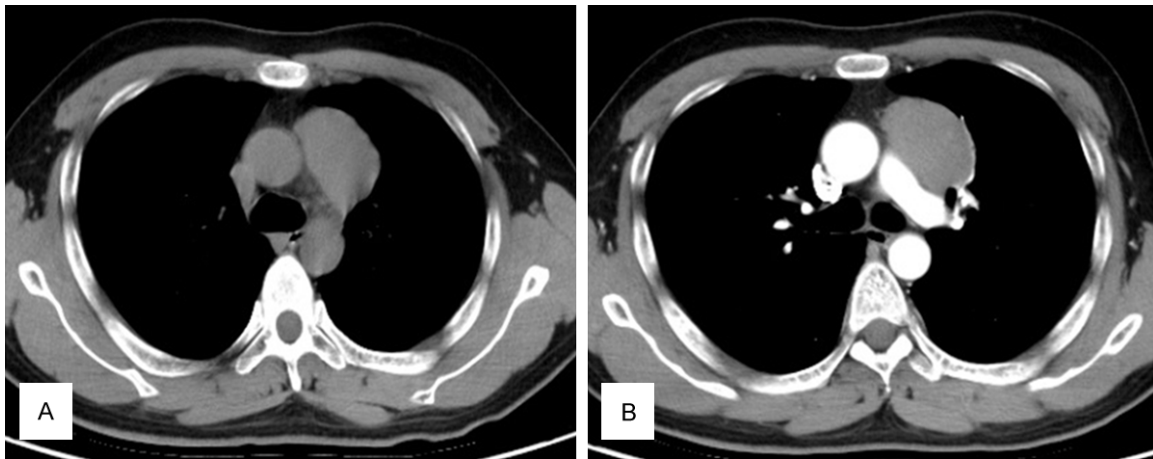


Figure 2. 35-year-old male with seminoma in the mediastinum. A. The mass shows homogeneous attenuation and well-circumscribed on non-enhanced CT image. B. The mass reveals lightly homogeneous enhancement after contrast administration.

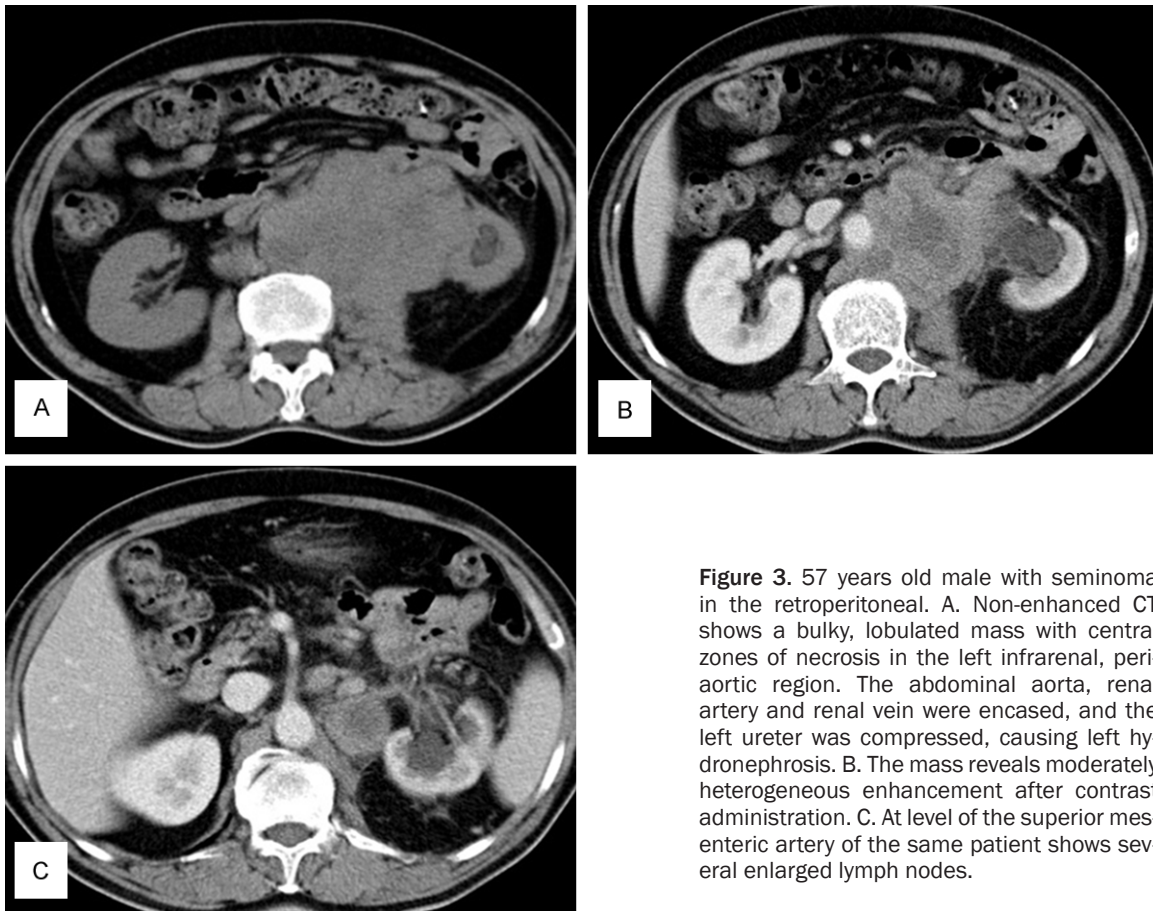


Figure 3. 57 years old male with seminoma in the retroperitoneal. A. Non-enhanced CT shows a bulky, lobulated mass with central zones of necrosis in the left infrarenal, peri-aortic region. The abdominal aorta, renal artery and renal vein were encased, and the left ureter was compressed, causing left hydronephrosis. B. The mass reveals moderately heterogeneous enhancement after contrast administration. C. At level of the superior mesenteric artery of the same patient shows several enlarged lymph nodes.

Three cases of extragonadal seminoma in the pelvis were oval-like and well-circumscribed, and the maximal diameter of the biggest one

was 12.2 cm. These cases presented central necrosis, and showed heterogeneous enhancement after contrast administration (**Figure 4**).



Figure 4. 42 years old male with seminoma in the pelvis. A. Non-enhanced CT image shows an oval-like, well-circumscribed mass with central necrosis. B. The mass shows heterogeneous enhancement after contrast administration.

All the cases were pathologically diagnosed by surgical or percutaneous needle biopsy.

Surgical excision was performed in 6 seminomas (3 cases located in the mediastinum, 3 cases located in the pelvis), followed by postoperative radiotherapy, and all of these 6 patients survived without relapse. 1 case located in the mediastinum received two courses of chemotherapy, then referred to radiotherapy, to a dose of 30 Gy in 17 fractions. The patient was disease-free after 20 months. 1 case located in the retroperitoneum only received radiotherapy, and prolong palliation was achieved. 1 case located in the retroperitoneum refused therapy for hepatic metastasis and was dead 3 months later.

Discussion

Germ cell tumors in men usually arise from the gonad. Extragonadal germ cell tumors are rare, and are usually located along the midline structures, which have been reported to arise in the pineal gland, the anterior mediastinum and the retroperitoneum [6, 7]. They comprise 5% to 10% of all germ cell tumors [1]. 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 [8].

Two embryologic theories have been proposed to explain the development of primary extrago-

nadal germ cell tumors. The first posits a failure of some primordial germ cells to complete their midline migration from the urogenital ridge to the scrotum. During the migration, germinal epithelium may be sequestered along the route and ultimately undergoes malignant transformation, then may form primary extragonadal seminoma [9]. The second theory proposes there are rests of totipotent cells left behind during embryonic development [10]. The histology of these tumors is similar to that of the tumors arising in the gonads.

Primary extragonadal germ cell tumors can be teratomatous and non-teratomatous tumors, and the latter tumors are malignant entities, and are extremely rare. Reports revealed that non-teratomatous tumors usually occurred in young males, and patients with such tumors had no special clinical manifestations, which resulted in bulky mass when they were referred to hospital for treatment [11, 12]. In our study, except for one male patient of 67 years old, all of the rest patients were young or middle aged males with a mean age of 38 years, which was consistent with previous reports. The clinical manifestations are different for its different locations. One patient with mediastinal seminoma complained of chest pain and cough, and one patient with mediastinal seminoma had a fever for one week. One patient with retroperitoneal seminoma complained of stomachache and anuria, because of hydronephrosis of the left kidney, which were consistent with the literature.

It is often difficult to differentiate a primary extragonadal germ cell tumor from a metastatic disease, due to an undetected or spontaneously regressed primary gonadal tumor. A valuable imaging sign is that a primary extragonadal germ cell tumor should be a midline mass. If there is predominantly a right- or left-sided adenopathy, this is compatible with an occult primary site of origin in the ipsilateral testis [9]. In our cases, 4 mediastinal masses with 3 deviated to left and 1 deviated to right, one retroperitoneal mass and all pelvic masses had symmetrical midline position, but the clinical and ultrasonic examination of the testes were all negative for tumor. We speculate that this discrepancy may be related to small number of cases.

Primary extragonadal seminomas are pathologic malignancy, the tumors grow rapidly and irregularly, which develop into large, bulky, ill-circumscribed masses with lobulated shape [13-16]. In our study, 8 masses appeared lobulated and ill-circumscribed. On the other hand, the maximal diameter of all the 9 masses was 12.8 cm, with a mean diameter of 4.6 cm. Hence, a large, bulky mass with irregular margin and lobulated shape should be taken as characteristic CT findings of primary extragonadal seminomas.

In our study, 8 masses revealed heterogeneous attenuation on non-enhanced CT images with patchy low-attenuation foci and showed moderately heterogeneous enhancement after contrast administration. This result accords with previous reports [16-18]. Pathologic study confirmed that low-attenuation foci reflected necrosis within the lesions. One mass showed homogeneous attenuation on non-enhanced CT images and lightly homogeneous enhancement after contrast administration. Reports suggested that majority of seminomas presented heterogeneous attenuation with low-attenuation foci on non-enhanced CT images and heterogeneous enhancement on contrast-enhanced CT images [7]. In our cases, only one case revealed homogeneous on non-enhanced CT and homogeneous enhancement on contrast-enhanced CT images, which was in accord with reports.

Primary extragonadal seminomas present local invasion or distant metastasis, such as involvement of adjacent vascular structures and pul-

monary, hepatic, osseous metastasis [18]. In our study, mediastinal vessels were involved in 3 mediastinal masses (3 of 4). The abdominal aorta, renal artery and renal vein were circled in 2 retroperitoneal masses (2 of 2). One mass compressed the left ureter, causing severe hydronephrosis of the left kidney, and one mass had hepatic metastasis.

Conclusion

To sum up, the characteristic CT findings of primary extragonadal seminomas include bulky, ill-circumscribed masses with lobulated shape, heterogeneous attenuation with low-attenuation foci on non-enhanced CT and heterogeneous enhancement after contrast administration. The tumor is apt to involve adjacent structures. So primary extragonadal seminomas should be considered when masses detected with these CT findings, and located in midline structures, such as the mediastinum, retroperitoneum, pelvis or the pineal gland, especially in young and middle-aged males.

Disclosure of conflict of interest

None.

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