CASE REPORT
Oncology & Hematology

JKMS

http://dx.doi.org/10.3346/jkms.2013.28.3.476 • J Korean Med Sci 2013; 28: 476-479

Embryonal Rhabdomyosarcoma Arising from a Mediastinal Teratoma: An Unusual Case Report

Young-Joon Ryu,[1,2] Su Hyun Yoo,[1] Min Jung Jung,[1] Sejin Jang,[1] and Kyung-Ja Cho[1]

[1]Department of Pathology, University of Ulsan College of Medicine, Asan Medical Center, Seoul; [2]Laboratory of Veterinary Theriogenology, Seoul National University College of Veterinary Medicine, Seoul, Korea

Received: 10 July 2012
Accepted: 11 January 2013

Address for Correspondence:
Young-Joon Ryu, MD
Department of Pathology, University of Ulsan College of Medicine, Asan Medical Center, 88 Olympic-ro, Songpa-gu, Seoul 138-736, Korea
Tel: +82.2-3010-1764, Fax: +82.2-472-7898
E-mail: mindmader@gmail.com

We report an unusual case of 9.5-cm-sized embryonal rhabdomyosarcoma arose from a mediastinal mature teratoma in a 46-yr-old man. A man presented with chest trauma as a result of an accident at 10 September 2011. On chest X-ray, an anterior mediastinal mass was detected. To obtain further information, chest computed tomography (CT) with contrast enhancement was performed, revealing an anterior mediastinal mass. Complete surgical excision was performed and entire specimen was evaluated. Pathologic diagnosis was embryonal rhabdomyosarcoma arising in mature cystic teratoma. After surgical excision, two cycles of dactinomycin-based chemotherapy were performed. Lung metastasis was detected on follow up CT in September 2012, and wedge resection was performed. Pathological finding of the lung lesion showed same feature with that of primary rhabdomyosarcoma.

Key Words: Mediastinum; Rhabdomyosarcoma; Embryonal; Teratoma

INTRODUCTION

Teratoma is common neoplasm that is typically encountered in infants and children. The majority of cases is benign, and arises from the sacro-coccygeal region, ovary, mediastinum, or retroperitoneum. A small percentage of cases exhibit malignant components, present at unusual site, or present during adult middle age (1). Primary germ cell tumors of the mediastinum represent approximately 10% of all neoplasm in the mediastinal area. The histopathologic features are similar to those of germ cell tumors in the gonads (2, 3). Malignant transformation in mediastinal germ cell tumor, especially to embryonic rhabdomyosarcoma is very rare (4-7). Furthermore, mediastinal embryonal rhabdomyosarcoma that arose from benign mature teratoma without other germ cell tumor components has not been reported in English literature, to our knowledge. In this report, we present such a case of a primary embryonal rhabdomyosarcoma of the anterior mediastinum that arose from a tumor that was only associated with a mature teratoma in middle-aged adult.

CASE DESCRIPTION

A 46-yr-old man presented with chest trauma as a result of an accident on 10 September 2011. On chest X-ray, an anterior mediastinal mass was detected (Fig. 1A). To obtain further information, chest computed tomography (CT) with contrast enhancement was performed, revealing a 9.5-cm-sized anterior mediastinal mass (Fig. 1B). The initial radiological diagnosis was thymoma. The radiological differential diagnoses were germ cell tumor and mediastinal sarcoma. Clinical and ultrasonography examination of the testicles were normal. Complete surgical excision was performed and entire specimen was evaluated. Grossly, the specimen showed a well-circumscribed 9.5 × 8 × 6-cm-sized round firm mass that was covered with normal thymic and perithymic soft tissue (Fig. 1C). The cut surface was flesh-colored and pale tan appearances (Fig. 1D). Microscopically, tumor demonstrated two different areas, including mature teratoma and sarcomatous area. In the teratoma area, the tumor demonstrated mature squamous epithelium, hair, skin appendage, adipose tissue, and other mesenchymal tissue (Fig. 2A). Immature components were not identified. There is also intermingled areas compact cellular and loose and myxoid area (Fig. 2B). In the sarcomatous area, there is some histological architecture resembled embryonic muscle, which forms aggregates of myoblasts amid loose, myxoid mesodermal tissue. The myoblastic tumor showed sheets of rhabdoid cells, which were small and spindled in appearance, and demonstrated moderate to poor differentiation. The cell also demonstrated deeply eosinophilic cytoplasm, and small eccentric oval-shaped nuclei (Fig. 2C). Immunohistochemical (IHC) analysis revealed a strong positivity to anti-myogenin and anti-desmin antibodies in the rhabdomyosarcoma component (Fig. 2D). The surgical resec-

pISSN 1011-8934
eISSN 1598-6357

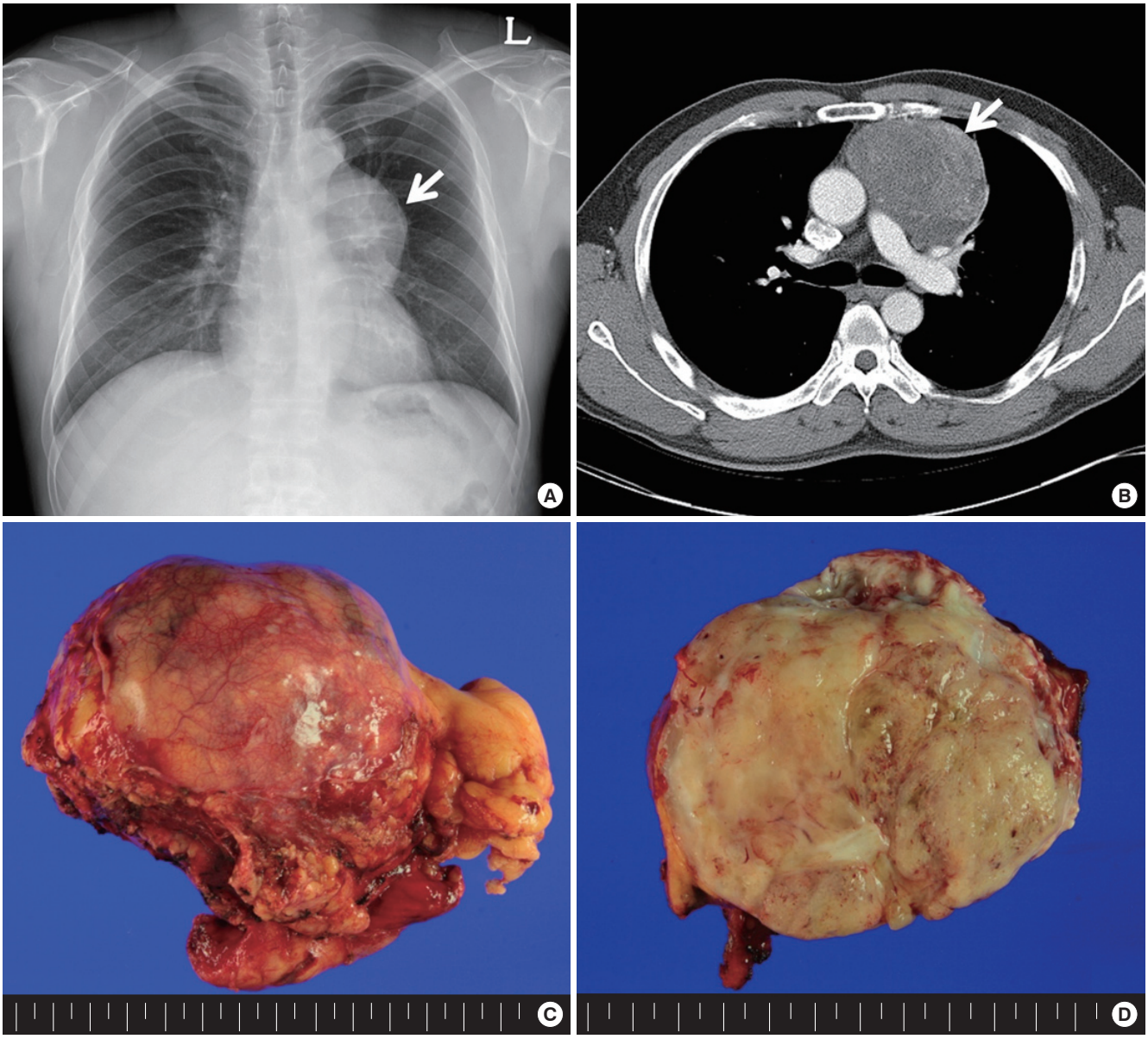

Fig. 1. Radiologic and gross findings of mediastinal rhabdomyosarcoma arising in mature teratoma. (**A**) An anterior mediastinal mass in chest X-ray. (**B**) A 9.5-cm-sized anterior mediastinal mass in Chest Computed tomography. (**C**) 9.5-cm-sized a well-circumscribed round and firm mass, which is covered as normal thymus and peri-thymic soft tissue. (**D**) The cut surface of mass show flesh meat like pale tan appearance.

tion margin was clear. After surgical excision, two cycles of dactinomycin-based chemotherapy were performed. Lung metastasis was detected on follow up CT on September 2012, and wedge resection was performed. Pathological finding of lung lesion showed same feature with that of primary rhabdomyosarcoma.

DISCUSSION

In our pathologic examination, sarcomatous area of tumor showed typical histological characteristics of embryonic rhabdomyosarcoma. It showed same typical cellular features with usual embryonal rhabdomyosarcoma. These tumor cells were confirmed by immunohistochemical marker, such as anti-myogenin and anti-desmin antibodies.

A review of literature revealed 32 previous reported patients with mediastinal rhabdomyosarcoma (including all subtypes) that arose from germ cell tumor in 12 separate studies (1-15). The average age of these patients (excluding 2 cases that lacked data) was 26.4 yr. The majority of patients were men (M:F ratio = 28:2). Among these 32 cases, embryonal rhabdomyosarcoma was only observed in 8 cases. Of particular note, there were no reported cases of embryonal rhabdomyosarcoma with only a mature teratoma, thus making our present case unique.

A non-germinal malignant tumor developing within a germ cell tumor is an extremely rare event in both gonadal and extragonadal loci. Moreover, embryonal rhabdomyosarcoma in adult

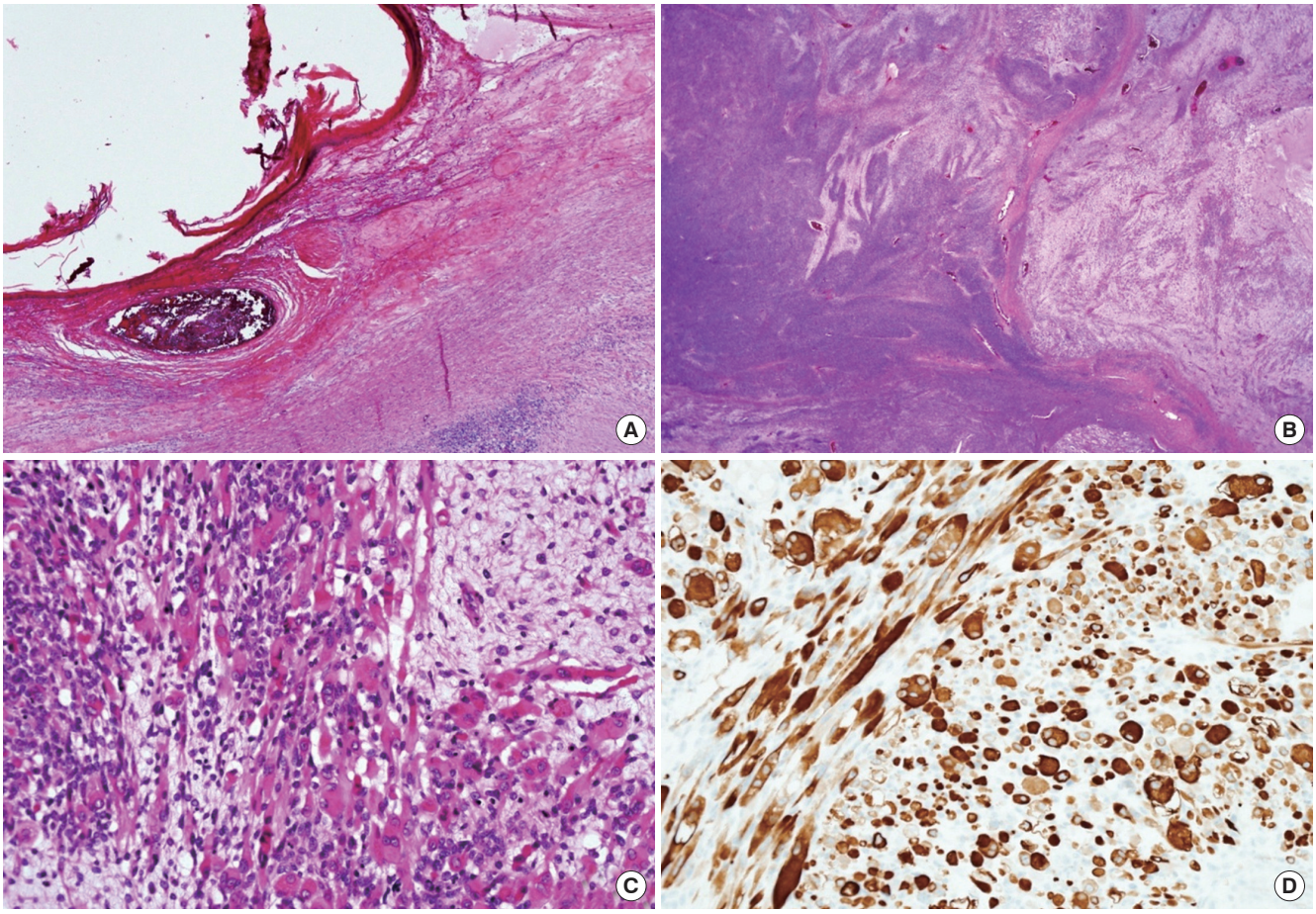

Fig. 2. Findings of microscopy and immunohistochemistry. (**A**) In teratoma area, mature squamous epithelium is seen. It is very close to embryonal rhabdomyosarcoma component (H&E, × 40). (**B**) Low power view of sarcoma area. Intermingled dense compact cellular area and loose myxoid area (H&E, × 12.5). (**C**) Numerous rhabdoid myoblasts. Epithelioid rhabdomyosarcoma cell showed deeply eosinophilic cytoplasm and small eccentric oval shaped nuclei (H&E, × 400). (**D**) Tumor cells show immunopositivity for Desmin (× 400).

patient's mediastinum is also very rare (4). In our review of the pathological findings reported in the literature, only three studies describe and undetermined type rhabdomyosarcoma with teratoma (1, 14, 15). In all reports, rhabdomyosarcoma was usually observed in combination with seminoma (6), myxoid liposarcoma (4), immature teratoma (4, 9, 13), embryonal carcinoma (4-6, 10), chondrosarcoma (4), choriocarcinoma (5), endodermal sinus tumor (6), angiosarcoma (6), and yolk sac tumor (7, 10, 13) (Table 1). This may represent important histological evidence that rhabdomyosarcoma arises from skeletal component of a mature teratoma. Consistently, previous immunohistochemical studies support this possibility.

In general, the prognosis for patients with mediastinal rhabdomyosarcoma is poor. Possible causes of this are the difficulty in obtaining early detection and the chemo-resistance of these tumors, and proper metastatic potential. The detection diagnosis of mediastinal rhabdomyosarcoma is usually fortuitous. Furthermore, because the common initial radiological diagnosis is benign thymoma, clinicians often pay little attention to these lesions (3). Unfortunately, these tumors exhibit chemo-resistance and metastatic potential (2, 14, 16), and presently, there is no standard schedule for performing chemotherapy to treat adult cases of embryonal rhabdomyosarcomas. Even existed pediatric protocols are often not effective in either the germinal and non-germinal components. Patients die mostly as a result of regional involvement and multiple metastases (4, 10). However, if complete excision is performed, the recurrence rate will decrease (10, 17).

In summary, primary rhabdomyosarcomas arising from mature teratoma in the mediastinum are very rare in adults. Because of the risk of recurrence in these cases, complete surgical resection should be considered both before and after chemotherapy.

REFERENCES

1. Cushing B, Bhanot PK, Watts FB Jr, Hertzler JH, Brough AJ. *Rhabdomyosarcoma and benign teratoma. Pediatr Pathol 1983; 1: 345-8.*
2. Dulmet EM, Macchiarini P, Suc B, Verley JM. *Germ cell tumors of the mediastinum: a 30-year experience. Cancer 1993; 72: 1894-901.*
3. Moran CA, Suster S. *Primary germ cell tumors of the mediastinum: I. analysis of 322 cases with special emphasis on teratomatous lesions and*

 http://dx.doi.org/10.3346/jkms.2013.28.3.476

Table 1. Clinical and pathologic details of primary mediastinal rhabdomyosarcoma

References	Case No.	Age	Sex	Subtype of rhabdomyosarcoma	Combined pathologic finding
Cushing et al. (1) 1983	1	14	Women	No data	Teratoma
Ulbringt et al. (4) 1984	2	21	No data	Embryonal	Teratoma/Seminoma/Myxoid liposarcoma
	3	32	No data	Embryonal	Immature teratoma/Embyonal carcinoma
	4	30	No data	Embryonal	Immature teratoma/Chondrosarcoma
Ahmed et al. (5) 1985	5	No data	No data	Embryonal	Endodermal sinus tumor/Embryonic carcinoma/Choriocarcinoma
Gonzalez-Vela et al. (6) 1990	6	16	No data	No data	Embryonal carcinoma/Endodermal sinus tumor
	7	23	No data	No data	Teratoma/Angiosarcoma
	8	22	No data	No data	Seminoma/Embryonal carcinoma/Endodermal sinus tumor/ Teratoma/Angiosarcoma
Caballero et al. (7) 1992	9	61	Man	No data	Teratoma/Yolk sac tumor
	10	29	Man	No data	Yolk sac tumor
Suster et al. (8) 1994	11	19	Woman	No data	No data
	12	20	Man	No data	No data
	13	26	Man	No data	No data
	14	27	Man	No data	No data
Corbett et al. (9) 1994	15	4	Man	Embryonal	Immature teratoma
Omezzine et al. (10) 2002	17	29	Man	Embryonal	Embryonal carcinoma/yolk sac tumor
Donadio et al. (11) 2003	18	No data	No data	Embryonal	No data
Sumerauer et al. (12) 2006	19	17	Man	Embryonal	No germ cell elements
Malagon et al. (13) 2007	20	22	Man	No data	Teratoma
	21	25	Man	No data	Teratoma
	22	27	Man	No data	Teratoma
	23	22	Man	No data	Teratoma
	24	74	Man	No data	Teratoma
	25	51	Man	No data	Yolk sac tumor
	26	24	Man	No data	Teratoma
	27	66	Man	No data	Teratoma
	28	34	Man	No data	Teratoma
	29	30	Man	No data	Teratoma/MPNST
	30	28	Man	No data	Immature teratoma
	31	23	Man	No data	Teratoma/yolk sac tumor
Vyas et al. (14) 2008	32	30	Man	Pleomorphic rhabdomyosarcoma	No evidence of germ cell tumor

a proposal for histopathologic classification and clinical staging. Cancer 1997; 80: 681-90.

4. Ulbright TM, Loehrer PJ, Roth LM, Einhorn LH, Williams SD, Clark SA. *The development of non-germ cell malignancies within germ cell tumors: a clinicopathologic study of 11 cases. Cancer 1984; 54: 1824-33.*
5. Ahmed T, Bosl GJ, Hajdu SI. *Teratoma with malignant transformation in germ cell tumors in men. Cancer 1985; 56: 860-3.*
6. Gonzalez-Vela JL, Savage PD, Manivel JC, Torkelson JL, Kennedy BJ. *Poor prognosis of mediastinal germ cell cancers containing sarcomatous components. Cancer 1990; 66: 1114-6.*
7. Caballero C, Gomez S, Matias-Guiu X, Prat J. *Rhabdomyosarcomas developing in association with mediastinal germ cell tumours. Virchows Arch A Pathol Anat Histopathol 1992; 420: 539-43.*
8. Suster S, Moran CA, Koss MN. *Rhabdomyosarcomas of the anterior mediastinum: report of four cases unassociated with germ cell, teratomatous, or thymic carcinomatous components. Hum Pathol 1994; 25: 349-56.*
9. Corbett R, Carter R, MacVicar D, Horwich A, Pinkerton R. *Embryonal rhabdomyosarcoma arising in a germ cell tumour. Med Pediatr Oncol 1994; 23: 497-502.*
10. Omezzine N, Khouatra C, Larivé S, Freyer G, Isaac-Pinet S, Gérinière L, Droz JP, Souquet PJ. *Rhabdomyosarcoma arising in mediastinal teratoma in an adult man: a case report. Ann Oncol 2002; 13: 323-6.*
11. Donadio AC, Motzer RJ, Bajorin DF, Kantoff PW, Sheinfeld J, Houldsworth J, Chaganti RS, Bosl GJ. *Chemotherapy for teratoma with malignant transformation. J Clin Oncol 2003; 21: 4285-91.*
12. Sumerauer D, Vicha A, Zuntova A, Stejskalova E, Krskova L, Kabickova E, Kodet R, Eckschlager T. *Teratoma in an adolescent with malignant transformation into embryonal rhabdomyosarcoma: case report. J Pediatr Hematol Oncol 2006; 28: 688-92.*
13. Malagón HD, Valdez AM, Moran CA, Suster S. *Germ cell tumors with sarcomatous components: a clinicopathologic and immunohistochemical study of 46 cases. Am J Surg Pathol 2007; 31: 1356-62.*
14. Vyas V, Al Awadi S, Nemec J, El Khodary A, Francis IM, Muralidharan KC, Delvadiya MD. *Primary mediastinal pleomorphic rhabdomyosarcoma: a case report. Med Princ Pract 2008; 17: 154-6.*
15. Motzer RJ, Amsterdam A, Prieto V, Sheinfeld J, Murty VV, Mazumdar M, Bosl GJ, Chaganti RS, Reuter VE. *Teratoma with malignant transformation: diverse malignant histologies arising in men with germ cell tumors. J Urol 1998; 159: 133-8.*
16. Comiter CV, Kibel AS, Richie JP, Nucci MR, Renshaw AA. *Prognostic features of teratomas with malignant transformation: a clinicopathological study of 21 cases. J Urol 1998; 159: 859-63.*
17. Kattan J, Culine S, Terrier-Lacombe MJ, Théodore C, Droz JP. *Paratesticular rhabdomyosarcoma in adult patients: 16-year experience at Institut Gustave-Roussy. Ann Oncol 1993; 4: 871-5.*