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

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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 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

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.
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**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 extra-gonadal loci. Moreover, embryonal rhabdomyosarcoma in adult
patient's mediastinum is also very rare (4). In our review of the pathological findings reported in the literature, only three studies describe 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), angioma (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, Macchiari 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
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 |
| Ulbright 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 sinu tumor/Embryonic carcinoma/Choriocarcinoma |
| Gonzalez-Vela et al. (6) 1990       | 6        | 16  | No data | No data | Embryonal carcinoma/Endodermal sinu tumor/ |
|                                    | 7        | 23  | No data | No data | Teratoma/Angiolsarcoma/ |
|                                    | 8        | 22  | No data | No data | Seminoma/Embryonal carcinoma/Endodermal sinu tumor/ |
|                                    |          |     |       |       | Teratoma/Angiolsarcoma/ |
| 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 | No evidence of germ cell tumor |
|                                    |          |     |     | rhabdomyosarcoma | |

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

4. Ulbright TM, Loehrer PJ, Roth LM, Einhorn LH, Williams SO, 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 1996; 66: 1461-46.

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: 234-96.

9. Corbett R, Carter R, MacVicar D, Horvich A, Pinkerton R. Embryonal rhabdomyosarcoma arising in a germ cell tumour. Med Pediatr Oncol 1994; 23: 497-502.

10. Omezzine N, Khouraou C, Larive S, Freyer G, Isaac-Pinet S, Geriniere L, Droz JP, Souquet PJ. Rhabdomyosarcoma arising in mediastinal teratoma in an adult: a case report. Ann Oncol 2002; 13: 323-6.

11. Donadio AC, Motzer RJ, Bajorin DF, Kantoff PW, Sheinfeld J, Houldsworth I, Chaganti RS, Bosl GJ. Chemotherapy for teratoma with malignant transformation. J Clin Oncol 2003; 21: 4285-91.

12. Sumerauer D, Vicha A, Zuntoha A, Stejskalova E, Krskova L, Kabickova E, Kodet R, Eckschlager T. Teratoma in an adolescent with malignant transformation into embryonal rhabdomyosarcoma: a 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: 659-63.

17. Kattan J, Cunliffe S, Terrier-Lacombe MJ, Théodore C, Droz JP. Paratesticular rhabdomyosarcoma in adult patients: 16-year experience at Institut Gustave-Roussy. Ann Oncol 1993; 4: 671-5.