Respiratory Medicine Case Reports 29 (2020) 100988

Contents lists available at ScienceDirect

Respiratory Medicine Case Reports

journal homepage: http://www.elsevier.com/locate/rmcr

Case report

A rapidly growing mature mediastinal teratoma with a testicular epidermoid cyst and familial Mediterranean fever

Kenshiro Omura [a], Masayuki Nakao [a,*], Hironori Ninomiya [b], Naoya Iwamoto [a], Hiroki Ozawa [a], Yohei Kawaguchi [a], Yasuto Kondo [a], Junji Ichinose [a], Yosuke Matsuura [a], Sakae Okumura [a], Mingyon Mun [a]

[a] Department of Thoracic Surgical Oncology, Cancer Institute Hospital of Japanese Foundation for Cancer Research, Tokyo, Japan
[b] Division of Pathology, Cancer Institute Hospital of Japanese Foundation for Cancer Research, Tokyo, Japan

ARTICLE INFO

Keywords:
Mediastinal tumor
Mature teratoma
Rapidly growing
Testicular epidermoid cyst
Familial Mediterranean fever

ABSTRACT

Anterior mediastinal teratomas are common and are generally characterized as slow growing tumors. Very few reports documenting rapidly growing tumors exist. Here, we describe a case of a mature teratoma showing rapid growth in 1 year treated with complete surgical resection.

A 25-year-old man who had a history of familial Mediterranean fever was referred to our hospital for further evaluation and treatment of an anterior mediastinal tumor with the largest diameter of 12 cm. A follow-up chest computed tomography of familial Mediterranean fever performed in the previous year showed no abnormal findings, therefore, he was suspected of having a malignant mediastinal tumor. During a systemic examination, a left testicular cyst was identified incidentally. We performed a complete resection of the mediastinal tumor and the left testicular cyst simultaneously. Following this, a benign mature teratoma in the anterior mediastinum and an epidermoid cyst in left testicle were pathologically diagnosed. The postoperative course was uneventful, and no evidence of recurrence was indicated 1 year after the surgery. We should be aware of the rapid growth potential in any mature teratoma and follow up accordingly.

1. Introduction

Teratomas in the anterior mediastinum are common tumors [1] that generally grow slowly; they are often asymptomatic and identified incidentally on chest X-rays [2]. However, there are some reported cases of benign mature teratomas in the anterior mediastinum that demonstrated rapid growth. The detailed mechanism of rapid growth is unclear in most of the previous cases [3–6]. Here, we describe a case of a mature teratoma showing rapid growth in one year treated effectively with complete surgical resection.

2. Case report

A 25-year-old man who had a history of familial Mediterranean fever (FMF) complained of sudden chest pain. His chest X-ray showed a large mass shadow in the left hilum, and subsequent chest computed tomography (CT) revealed an anterior mediastinal tumor, with the largest diameter of 12 cm, containing multiple cysts (Fig. 1A). However, there were no abnormal findings on the follow-up chest CT of FMF performed in the previous year (Fig. 1B). He was initially suspected of having a malignant mediastinal tumor and referred to our hospital for further examination and treatment. Our differential diagnoses included malignant mediastinal tumors, such as germ cell tumor, thymic epithelial tumor, or malignant lymphoma. However, serum tumor markers including β-human chorionic gonadotropin (HCG), alpha-fetoprotein (AFP), squamous cell carcinoma antigen (SCC) and soluble-interleukin 2 receptor (IL2R) were all within normal limits. Moreover, 18F-fluorodeoxyglucose-positron emission tomography/CT (PET/CT) showed low uptake of fluorodeoxyglucose ($SUV_{max} = 3.6$) in the mass, indicating a benign or low-grade malignant feature (Fig. 1C). Magnetic resonance imaging (MRI) demonstrated a huge mass with multiple cysts and solid components which suggested a mature teratoma (Fig. 1D). We then considered the possibility of a rare rapidly growing mature teratoma. We

* Corresponding author. 3-8-31 Ariake, Koto-ku, Tokyo, 135-8550, Japan.
E-mail address: masayuki.nakao@jfcr.or.jp (M. Nakao).

https://doi.org/10.1016/j.rmcr.2019.100988
Received 30 October 2019; Received in revised form 16 December 2019; Accepted 17 December 2019
Available online 17 December 2019

thought a percutaneous needle biopsy should be avoided considering the risk of tumor rupture or dissemination; therefore, surgery was planned for diagnosis and treatment. During a preoperative systemic examination, a left testicular cyst was identified incidentally (Fig. 2). Eventually, we performed a complete resection of the left testicular cyst and the mediastinal tumor, included a part of the left lung that was adhered to the mediastinal tumor (Fig. 3).

Microscopically, the tumor consisted of several cysts, which were lined by mature epithelium, containing sebaceous glands, nests of respiratory epithelium, cartilage, and parathyroid glands (Fig. 4A, B and C). There were no immature components in the specimen and therefore these findings confirmed the diagnosis of a mature teratoma. There were no characteristic findings to explain the rapid tumor growth. In addition, pathological findings of the testicular cyst, which was lined internally with squamous epithelium, revealed a testicular epidermoid cyst. The postoperative course was uneventful, and he was discharged without any complications. No disease recurrence was seen in the follow-up radiological findings 1.5 years after surgery.

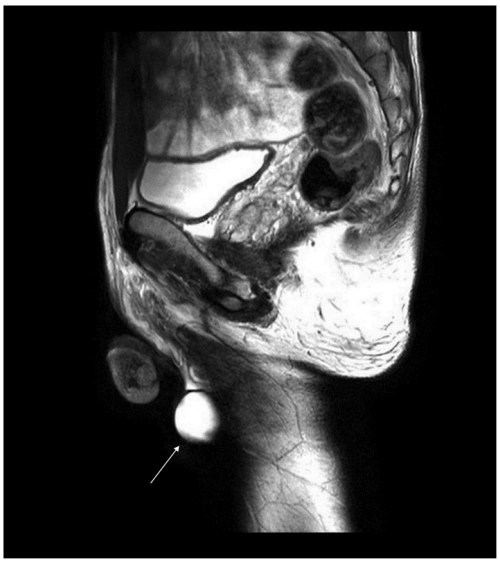

Fig. 2. T2-weighed MRI showing a homogenous high intensity mass (white arrow) in the patient's left testicle.

3. Discussion

We encountered a very rare case of a mature mediastinal teratoma showing rapid growth in 1 year accompanied by testicular epidermoid cyst, and FMF. A teratoma is a tumor that arises from germ cells and is usually found in the gonads; however, there are some extragonadal sites where teratomas can develop, including the anterior mediastinum which is the most common extragonadal site [1]. Mediastinal teratomas are generally asymptomatic and commonly found incidentally on chest radiography [2]. Most symptoms, including chest pain, dyspnea and cough, are related to compression of neighboring structures. When the tumor ruptures, these symptoms worsen, and some patients need to undergo an emergent operation [3]. In previous reports, most of the tumors had a slow-growing nature, while there were five reported cases associated with rapid growing tumors [3–7]. The cause of rapid growth among the tumors was unclear; however, bleeding, rupture, female hormones and pancreatic enzymes were thought to be associated factors [3–6]. In this case, we could not identify pathological findings of bleeding, rupture, and pancreatic tissue in the resected specimen. Also, as this patient is a male, female hormones were less likely to be the cause of rapid growth.

Testicular epidermoid cysts can be classified into a prepubertal type of teratoma according to the 2016 WHO classification [8]. From a clinical perspective, it seems that there is a relationship between mediastinal teratomas and testicular cysts. However, the histopathological findings of a testicular cyst show limited evidence of teratoma differentiation because mesodermal and endodermal components are absent [9]. Therefore, each disease is considered to be independent. Moreover, our patient had a history of FMF. Some authors have previously reported that FMF is associated with the pathogenesis of benign and malignant lesions such as multiple pelvic cysts [10] and pericardial cysts [11]. Therefore, the chronic inflammation associated with FMF may have some effect on the pathogenesis or rapid growth of a teratoma. In addition, tumor rupture, often accompanied by slight fever and chest pain, would be difficult to identify since these symptoms are also caused by FMF. Hence, a mediastinal teratoma may have been missed without the radiological findings.

This case is the first report of co-existence of a mature mediastinal teratoma, testicular epidermoid cyst, and FMF. Although each disease is considered to be independent, we suspect that each condition may have some relevance at the genetic level.

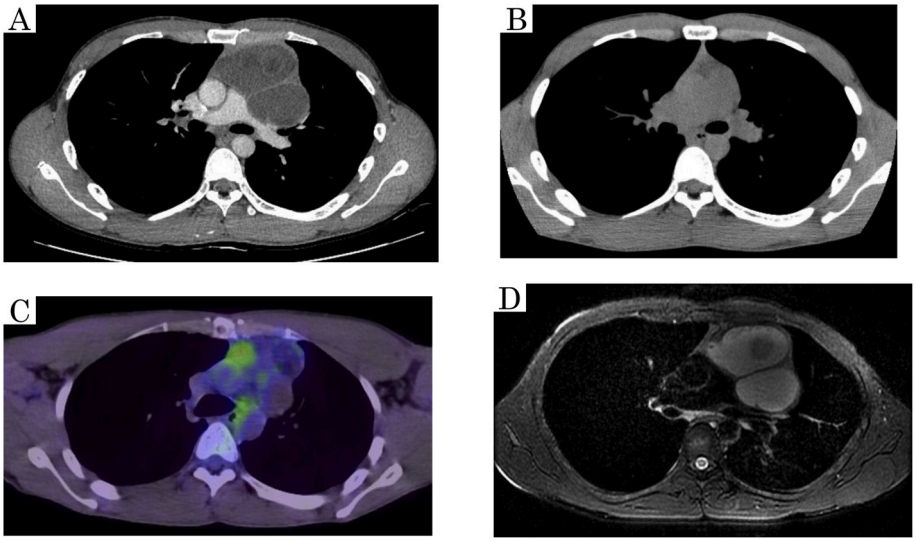

Fig. 1. Radiological examinations. A: Contrast-enhanced CT showing a large heterogenous mass shadow containing multiple cysts in the anterior mediastinum. B: There were no abnormal findings on the chest CT taken last year. C: PET-CT showing a low uptake only in the upper area of the mass. D: T2-weighed MRI showing high intensity areas predominantly consistent with multiple cysts.

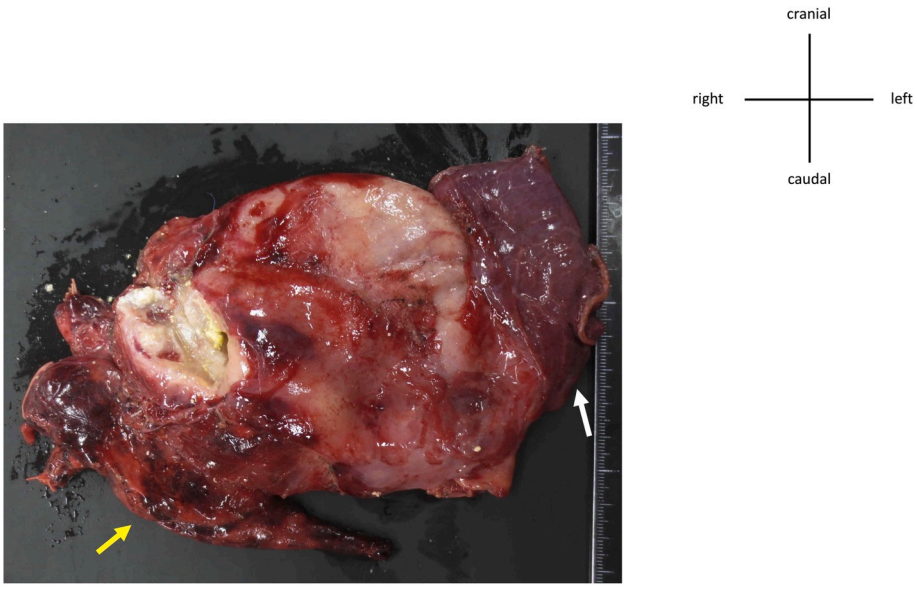

Fig. 3. Macroscopic findings. The mediastinal tumor, a solid and white colored mass, was resected completely with a part of the left lung (white arrow) and thymus (yellow arrow). (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

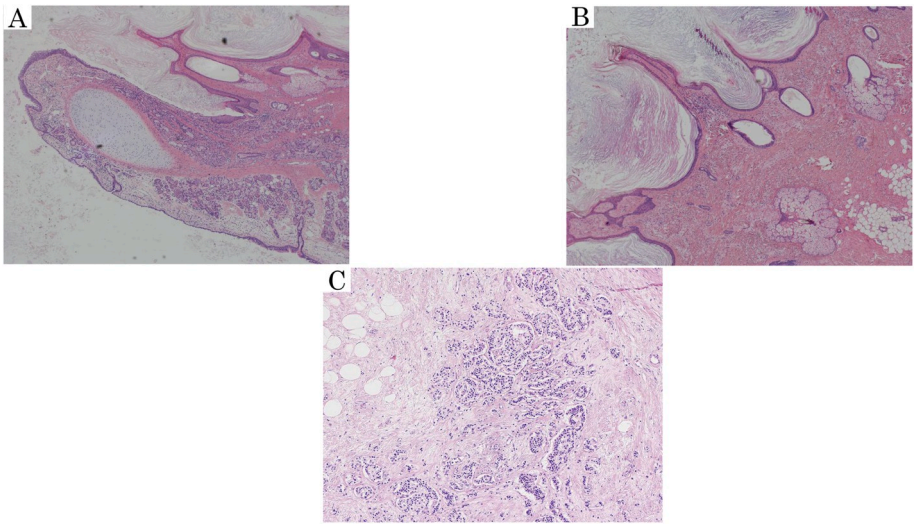

Fig. 4. Pathological findings. Histopathological analysis showing multiple areas of mature epidermis with respiratory epithelium (A), sebaceous glands (B), and parathyroid glands (C) in the mediastinal mass.

4. Conclusion

We encountered a very rare case of a mature mediastinal teratoma showing rapid growth. We should be aware of the rapid growth potential in any mature teratoma and follow up accordingly.

Declaration of competing interestCOI

The authors declare that they have no conflict of interest.

Funding acknowledgements

None to declare.

Informed consent statement

The patient provided written informed consent.

References

[1] C.R. Nichols, Mediastinal germ cell tumors. Clinical features and biologic correlates, Chest 99 (2) (1991) 472–479.
[2] B.D. Lewis, R.D. Hurt, W.S. Payne, G.M. Farrow, R.H. Knapp, J.R. Muhm, Benign teratomas of the mediastinum, J. Thorac. Cardiovasc. Surg. 86 (5) (1983) 727–731.
[3] N. Omachi, T. Kawaguchi, S. Shimizu, T. Okuma, M. Kitaichi, S. Atagi, et al., Life-threatening and rapidly growing teratoma in the anterior mediastinum, Intern. Med. 54 (19) (2015) 2487–2489.
[4] S.A. Hussain, S. Shenaq, L. Mendoza, J. Sundermeyer, S. Chakarvarty, Mediastinal teratoma presenting as a rapidly enlarging paracardial mass, Int. Surg. 68 (2) (1983) 179–180.
[5] T. Uyama, Y. Monden, K. Harada, S. Kimura, T. Morimoto, K. Miura, et al., Rapidly growing mature teratoma of the mediastinum: do sex hormones affect growth of the tumor? J. Surg. Oncol. 38 (4) (1988) 285–289.
[6] K. Fujita, K. Hayashi, M. Motoishi, S. Sawai, T. Terashima, T. Mio, Giant mature teratoma in the mediastinum presenting with rapid growth, Oxf. Med. Case Rep. 12 (2016) 309–312.
[7] D. Kim, S. Kim, J. Hong, Rapid growing huge teratoma: complete surgical resection, J. Thorac. Dis. 6 (10) (2014) 217–219.
[8] P. Anheuser, J. Kranz, E. Stolle, D. Höflmayer, F. Büscheck, S. Mühlstädt, et al., Testicular epidermoid cysts: a reevaluation, BMC Urol. 19 (1) (2019) 52.

[9] Z.V. Maizlin, A. Belenky, J. Baniel, P. Gottlieb, J. Sandbank, S. Strauss MB, Epidermoid cyst and teratoma of the testis: sonographic and histologic similarities, J. Ultrasound Med. 24 (10) (2005) 1403–1409.

[10] M.K. Eryilmaz, H. Mutlu, G. Tazegul, R. Eryılmaz, F.Y. Müsri, D.K. Salim, et al., Multiple pelvic cysts in a patient with familial Mediterranean fever: benign cystic mesothelioma, J. Cancer Res. Ther. 13 (6) (2017) 1047–1049.

[11] A. Çelik, B. Çalapkorur, I. Özdogru, Case images: a pericardial cyst due to familial Mediterranean fever, Turk Kardiyol. Dernegi Arsivi 38 (5) (2010) 379.