

Undifferentiated pleiomorphic sarcoma simultaneously occurring with thymoma

Christos Simoglou¹, Nikolaos Tsolakis²

¹Department of Cardiothoracic Surgery, University of Alexandroupolis, School of Medicine, Alexandroupolis, Greece; ²Cardiothoracic Surgery Department, St. Luke's Hospital, Panorama Thessaloniki, Greece

Correspondence to: Dr. Christos Simoglou. Afroditis 11, T.K.69100, Komotini, Greece. Email: simoglouchristosdoctor@yahoo.com.

Abstract: We report here a case of thymoma simultaneously associated with neuroendocrine tumor. A 65-year-old male, presented with cough. Radiographic studies showed a mediastinal mass. On fine needle aspiration cytology and histopathological examination, a diagnosis of thymoma with coexisting undifferentiated pleiomorphic sarcoma was made. Although thymoma is associated with many extrathymic malignancies, its association with neuroendocrine tumor is rare. This case is being reported on to reinforce that clinicians should bear in mind the possibility of extrathymic malignancies in patients with thymoma.

Keywords: Thymoma; cancer; neoplasm; diagnosis

Submitted Apr 11, 2014. Accepted for publication Aug 08, 2014.

doi: 10.3978/j.issn.2072-1439.2014.09.05

View this article at: <http://dx.doi.org/10.3978/j.issn.2072-1439.2014.09.05>

Introduction

Thymoma is a tumor of anterior mediastinum, however, thymomas may develop at various other sites like neck, trachea, thyroid, lung, and heart (1). It is the most common neoplasm of thymus, more common in men and in eighth decade of life (2). Thymoma is an uncommon neoplasm which arises from thymic epithelium. Thymomas are associated with paraneoplastic syndromes such as myasthenia gravis, hypogammaglobulinemia (3), pure red cell aplasia (4), and many kinds of immune mediated systemic disorders; however, few cases of extrathymic malignancies have been reported. Reported incidence of extrathymic malignancies is 2.6-27% (5-8). We report here a case of mediastinal thymoma with coexisting neuroendocrine tumor.

Case report

A 65-year-old male, presented with easy fatigability and a lump in the left side of his axilla for four months. On general examination mild pallor was noticed. Local examination of left axilla showed a lump of 8 cm × 8 cm, firm in consistency, fixed to underlying structures, and overlying skin was

normal. Investigations showed Hemoglobin is 7 g/dL and Erythrocyte sedimentation rate 25 mm in first hour. X-ray chest posteroanterior view revealed a mediastinal mass. CECT obtained at the level of arch of aorta and main pulmonary artery showed a well-defined enhancing mass of soft tissue attenuation in the left anterior mediastinum abutting arch of aorta, main and left pulmonary artery with maintained fat planes.

Adjacent mediastinal fat plane was partially obliterated. CT guided FNAC was done which showed moderately cellular smears consisting of a dual population of epithelial cells and mature appearing lymphocytes. The epithelial cells comprised of oval to elongated nuclei with dispersed chromatin and inconspicuous nucleoli.

Mitosis was not seen. Histopathology of mass revealed loose aggregates of lymphocytes admixed with neoplastic epithelial cells. A diagnosis of thymoma was rendered. Excisional biopsy of axillary mass was done which was composed of cells having pleomorphic, hyperchromatic spindle-shaped nuclei with clumped chromatin. Mitotic rate was high. Occasional multinucleated cell was also seen. Immunohistochemical analysis showed positivity for vimentin and focal positivity for CD 68 along with negative

smooth muscle actin, desmin, epithelial membrane antigen, CD 45, CD 3 and CD 30. On the basis of the above findings a diagnosis of thymoma with co-existent undifferentiated pleomorphic sarcoma was made. Postoperatively patient was discharged on 6th post-operative day. Follow up in our patient with invasive thymoma (surgical resection), treatment is based on induction chemotherapy and post-surgical radiation. 5-year survival for invasive thymoma is between 12-54% regardless of any myasthenia gravis symptoms.

Discussion

The coexisting thymomas with other malignancies is relatively rare, and the most common site for these extrathymic malignancies is colorectal (6,8) followed by lung, thyroid, prostate, female reproductive organs, breasts, kidney and skin melanoma. Only few cases of co-existent thymoma with coexisting undifferentiated pleomorphic sarcoma not otherwise specified (9,10) are reported in the literature. The pathogenesis of these associations is still unclear. In most of the cases associated tumors were diagnosed either before or at the same time of thymoma as in our case, which suggests that these patients may be genetically predisposed to develop tumors (11). Several different theories about the pathological basis have been proposed. According to Friedman (12), the onset of thymoma indicates defect within the thymus epithelium which is responsible for immune defect and increased incidence of neoplasia. Another theory suggests the potential ability of thymoma epithelial cells to stimulate T cells which predisposes to the onset of tumors (13,14). Undifferentiated pleomorphic sarcoma is the most common soft tissue sarcoma in adults, usually arising in extremities with a peak in seventh decade although cases in children have been reported (15,16). It can also develop at the site of previous radiation therapy (17). Enzinger and Weiss classified it histologically into storiform-pleomorphic, myxoid, giant cell, and inflammatory types (18). Storiform pleomorphic is the most common histologic type which consists of highly pleomorphic tumor cells arranged in storiform pattern. Immunohistochemistry is required for confirmation of diagnosis. Vimentin positivity and smooth muscle actin, desmin, epithelial membrane antigen, CD 45, CD 30, and CD 3 negativity as in our case made the final diagnosis. Local recurrence and metastasis to distant sites especially lungs and regional lymph nodes are common (19). Treatment is surgical resection and with or without adjuvant radiation and/or chemotherapy.

Acknowledgements

Disclosure: The authors declare no conflict of interest.

References

1. Myers PO, Kritikos N, Bongiovanni M, et al. Primary intrapulmonary thymoma: a systematic review. *Eur J Surg Oncol* 2007;33:1137-41.
2. Engels EA, Pfeiffer RM. Malignant thymoma in the United States: demographic patterns in incidence and associations with subsequent malignancies. *Int J Cancer* 2003;105:546-51.
3. Jacob S, Irani SR, Rajabally YA, et al. Hypothermia in VGKC antibody-associated limbic encephalitis. *J Neurol Neurosurg Psychiatry* 2008;79:202-4.
4. Rosenow EC 3rd, Hurley BT. Disorders of the thymus. A review. *Arch Intern Med* 1984;144:763-70.
5. Wilkins KB, Sheikh E, Green R, et al. Clinical and pathologic predictors of survival in patients with thymoma. *Ann Surg* 1999;230:562-72; discussion 572-4.
6. Masaoka A, Yamakawa Y, Niwa H, et al. Thymectomy and malignancy. *Eur J Cardiothorac Surg* 1994;8:251-3.
7. Hata M, Negishi N, Niino S, et al. A case of multiple colon cancer appeared after thymectomy for thymoma with myasthenia gravis. *Nihon Geka Gakkai Zasshi* 1993;94:1061-3.
8. Pan CC, Chen PC, Wang LS, et al. Thymoma is associated with an increased risk of second malignancy. *Cancer* 2001;92:2406-11.
9. Murphey MD. World Health Organization classification of bone and soft tissue tumors: modifications and implications for radiologists. *Semin Musculoskelet Radiol* 2007;11:201-14.
10. Nakagiri T, Okumura M, Inoue M, et al. Thymoma-associated graft-versus-host disease-like erythroderma. *J Thorac Oncol* 2007;2:1130-2.
11. Moran CA, Travis WD, Rosado-de-Christenson M, et al. Thymomas presenting as pleural tumors. Report of eight cases. *Am J Surg Pathol* 1992;16:138-44.
12. Honma K, Shimada K. Metastasizing ectopic thymoma arising in the right thoracic cavity and mimicking diffuse pleural mesothelioma--an autopsy study of a case with review of literature. *Wien Klin Wochenschr* 1986;98:14-20.
13. Payne CB Jr, Morningstar WA, Chester EH. Thymoma of the pleura masquerading as diffuse mesothelioma. *Am Rev Respir Dis* 1966;94:441-6.
14. Suster S, Rosai J. Histology of the normal thymus. *Am J Surg Pathol* 1990;14:284-303.

15. Maggi G, Casadio C, Cavallo A, et al. Thymoma: results of 241 operated cases. *Ann Thorac Surg* 1991;51:152-6.
16. Kondo K, Monden Y. Therapy for thymic epithelial tumors: a clinical study of 1,320 patients from Japan. *Ann Thorac Surg* 2003;76:878-84; discussion 884-5.
17. Regnard JF, Magdeleinat P, Dromer C, et al. Prognostic factors and long-term results after thymoma resection: a series of 307 patients. *J Thorac Cardiovasc Surg* 1996;112:376-84.
18. Okumura M, Shiono H, Inoue M, et al. Outcome of surgical treatment for recurrent thymic epithelial tumors with reference to world health organization histologic classification system. *J Surg Oncol* 2007;95:40-4.
19. Lucchi M, Davini F, Ricciardi R, et al. Management of pleural recurrence after curative resection of thymoma. *J Thorac Cardiovasc Surg* 2009;137:1185-9.

Cite this article as: Simoglou C, Tsolakis N. Undifferentiated pleiomorphic sarcoma simultaneously occurring with thymoma. *J Thorac Dis* 2014;6(9):E188-E190. doi: 10.3978/j.issn.2072-1439.2014.09.05