January 2006

5 year survival of a patient with malignant thymoma with superior vena caval syndrome

Saulat H. Fatimi
Aga Khan University, saulat.fatimi@aku.edu

Sadaf Sheikh
Aga Khan University

Saba Mansoor

5-YEAR SURVIVAL OF A PATIENT WITH MALIGNANT THYMOMA WITH SUPERIOR VENA CAVAL SYNDROME

Dear Sir,

Thymoma is an uncommon neoplasm that is derived from the epithelial cells of the thymus. It is well known for its association with myasthenia gravis (MG) or other autoimmune disease, histologic variability, and heterogeneity of malignant behaviour.[1] Surgery remains the mainstay of treatment while radiation and chemotherapy also have been applied widely as adjuvant and palliative procedures. Most malignant thymomas are slow-growing tumors which uncommonly metastasize hematogenously and have a tendency to recur locally.

A 41-year-old man presented to the Emergency Room with a 4-month history of cough, shortness of breath and low grade intermittent fever with no other past or present medical problems. On physical examination, he had extensive facial swelling and engorged neck veins. The rest of the physical examination was unremarkable. Chest X-ray showed widening of mediastinum. CT-scan of the chest was performed which showed a lobulated soft tissue mass (6x4cm) in the anterior mediastinum along with calcification in its peripheral margins. Superior Vena Caval (SVC) thrombosis was also identified with collateral development. VATS (video assisted thoracoscopic surgery) was performed to get the biopsy and the histopathology report showed changes consistent with malignant thymoma. Elective thymectomy was performed. The anterior mediastinal mass was found to be hard and encasing the SVC which was thrombosed. Further dissection revealed that the innominate artery, right carotid and right subclavian arteries were encased in the thymoma. The tumor was projecting deep posterior to the pulmonary artery and around the aorta and had completely invaded the left phrenic nerve and left innominate vein. Pulmonary extension of the tumor was evident by involvement of the right and left upper lobes. The entire tumor along with portions of both upper lobes was removed. Since there was involvement of the left phrenic nerve as well, pericardium and the left phrenic nerve were taken along with the tumor. The tumor was shaved off the thrombosed SVC.

The patient received postoperative radiotherapy and has been followed by the oncology service routinely since surgery. The facial swelling subsided completely 3 months after surgery. On a 5 years follow-up, the patient has shown no recurrence of his tumor on CAT scan and is totally asymptomatic.

Malignant thymomas are rare, slow-growing tumors and patients commonly present with cough, chest pain, phrenic nerve palsy or SVC syndrome, or less frequently with fever, night sweats and weight loss. CT scanning is usually accurate in predicting tumor size, location and invasion into vessels, pericardium and lung.

Even though invasiveness of the tumor remains the single most consistent factor in predicting outcome, several factors adversely affect the prognosis of malignant thymoma as well including intrathoracic or extrathoracic metastasis, tumor size > 10cm, extent of surgical resection, tracheal or vascular compromise, age < 30 years and epithelial/mixed histologies. Initially, the presence of an associated paraneoplastic syndrome or myasthenia gravis was regarded as a negative prognostic factor,[2,3,4] however, now these are not associated with inferior outcome any more, possibly because of earlier detection.[2]

Surgical resection remains the main mode of therapy, with an enbloc removal for invasive tumors in patients who can tolerate surgery and do not have extensive metastatic disease. The extent and completeness of surgical resection determines outcome.[4] In patients with extensive metastasis and unstable medical condition, debulking or biopsy may be the only option. In such cases, postoperative radiotherapy has been shown to increase survival rates.[2]

Thymomas appropriately treated by complete surgical resection, result in a 960% 10-year surviva.l The stage of the disease has an important impact upon survival outcomes. Malignant thymoma often involves the phrenic nerves and there is a probabililty of impaired respiratory function if the nerve is divided. Preoperative pulmonary function tests should be performed in all patients so that any respiratory compromise following surgical resection of the phrenic nerve can be estimated. In patients with completely resected stage II or III thymomas, adjuvant radiotherapy reduces the rate of local recurrence from approximately 28 to 5%.[4]

Thymomas are chemosensitive malignancies.[5] It is recommended that patients with unresectable disease should be given aggressive management with trimodality therapy. Cisplatin-containing regimens are by far the most active with overall response rates of 70-80%, of which half are complete. The most commonly used regimens are ADOC, PAC and Ep (etoposide and cisplatin). For recurrent disease, ADOC or PAC regimen can be used and in cases of patients not being able to tolerate cisplatin, carboplatin can be substituted. Currently, carboplatin and paclitaxel combination is being studied in clinical research.

REFERENCES

1. Kondo K, Yoshizawa K, Tsuyuguchi M, Kimura S, Sumitoma M, Morita J, *et al*. WHO histologic classification is a prognostic indicator in thymoma. *Ann Thorac Surg* 2004; **77**:1183-8.
2. Lardinois D, Rechsteiner R, Lang RH, Gugger M, Betticher D, von Briel C, *et al*. Prognostic relevance of Masaoka and Muller-Hermelink classification in patients with thymic tumors. *Ann Thorac Surg* 2000; **69**:1550-5.
3. Cameron RB, Loehrer PJ, Thomas CR Jr. Neoplasms of the mediastinum. In: Devita VJ, Hellman S, Rosenberg S, (edi). Cancer: principles and practice of oncology. 7th ed. Philadelphia: *Lippincott, Williams & Wilkins* 2005: 845-60.
4. Hernandez-Ilizaliturri FJ, Tan D, Cipolla D, Connolly G, Debb G. Rammathor multimodality therapy for thymic carcinoma (TCA): results of a 30-year single-institution experience. *Am J Clin Oncol* 2004; **27**: 68-72.

5. Venuta F, Rendina EA, Longo F, De Giacomo T, Anile M, Mercadant E, *et al.* Long-term outcome after multimodality treatment for stage III thymic tumors. *Ann Thorac Surg* 2003; **76**:1866-72.

DR. FATIMI SAULAT, DR. SHEIKH SADAF AND DR. MANSOOR SABA
Cardiothoracic Surgery, The Aga Khan University Hospital, Karachi.
E:mail: sadaf_sheik@gmail.com

.....☆.....

ERRATA

1. The name of co-author, Ali S.M. Akhtar has been misprinted as S.M. Akhtar in the article "NEONATAL COMPLICATIONS IN INFANTS BORN TO DIABETIC MOTHERS" published in JCPSP 2006, Vol. 16 (3): 212-15. The name may be corrected and read as A.S.M. Akhtar.

2. A discrepancy has occurred in the authorship of the article: "TRACHEAL INJURY DUE TO BLUNT CHEST TRAUMA: A RARE SURGICAL EMERGENCY" by Usman Ahmad, Muhammad A. Javed and Saulat H. Fatimi, published in JCPSP 2006,Vol. 16 (6): 422-23. The name of fourth co-author, Fahad Shuja, was inadvertently omitted and could not be printed. His name may be included in the above article and the authors' list should read as: Usman Ahmad, Muhammad A. Javed, Saulat H. Fatimi and Fahad Shuja.

3. Due to a typographic error the 'u' of edu was omitted from the E-mail address of correspondence author of the article titled "CORD BLOOD LEPTIN LEVELS IN PAKISTANI NEWBORNS: RELATIONSHIP WITH BIRTH WEIGHT, LENGTH AND OCCIPITOFRONTAL CIRCUMFERENCE" published in JCPSP 2006, Vol. 16 (6): 393-95. The E-mail address should read as ghulam.lakho@aku.edu.

Editor