32-Year Survival with Metastatic Adrenal Cortical Carcinoma – Update of a Case Report

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Abstract. Adrenal cortical carcinoma (ACC) is a rare and highly malignant tumour with up to 70% of the patients diagnosed at an advanced clinical stage and up to 40% presenting with metastasis. In 2004, we reported a case of a 62-year-old woman with a non-functional ACC of the left adrenal gland (T2 N0 M0, classified as stage II) who survived the disease for 28 years with 3 operations for metastases of the contralateral adrenal gland and 4 operations for metastases of the lung. We can now give an update of a further four years of survival and one additional operation. The case again emphasises the need for thorough radiographic examination and follow-up for the possibility of a repeated re-operation whenever possible to provide long-term survival over decades.

Adrenal cortical carcinoma (ACC) is a rare and highly malignant tumour that is often diagnosed in an advanced stage of disease. Despite curative surgical resection of the primary tumour, in up to 80% of the patients a local recurrent tumour or metastases develop that leads to a poor prognosis. In these patients, only re-operation of the recurrent tumour or resection of solitary metastases can enhance the survival. We present an update of a patient who is still alive, 32 years after the first tumour resection for non-functional left-sided adrenocortical carcinoma and repeated resections of lung metastases and contralateral adrenal gland metastases (1).

Case Report

We report an update of a case of a 66-year-old woman with a non-functional ACC of the left adrenal gland (T2 N0 M0, classified as stage II) who was treated by total left adrenalectomy and splenectomy 32 years ago. Histopathological examination revealed non-functional adrenal cortical carcinoma with a maximal diameter of 17 cm. Four years later resection of contralateral right-sided adrenal gland metastasis was carried out by subtotal adrenalectomy, nine years later two metastases of the left lung were resected and chemotherapy with mitotane was started then stopped after 15 months due to adrenal insufficiency. Seven years later, local recurrent disease of the right adrenal gland was resected; three years later metastases of the left lung were resected. One year later, metastases of the left and right lung were resected. Two years later again local recurrent disease in the region of the former right adrenal gland was resected with simultaneous cholecystectomy. Two years later new pulmonary metastasis of the left lung was successfully resected. The patient showed no evidence of disease for the next 3 years, but computed tomography of the abdomen again showed a newly developed formation in the region of the right adrenal gland with a diameter of 1.5 cm (Figure 1). A magnetic resonance image of the abdomen verified the diagnosis of recurrent metastases in the region of the right adrenal gland with a diameter of 1.5 cm (Figure 2). Therefore an extirpation of this tumour was carried out with simultaneous adhesiolysis of the bowel and partial resection of liver segment VI. Histopathological examination verified the diagnosis of metastatic disease of the known adrenocortical cancer; the hepatic lesion was inconspicuous. After this last operation, the patient is still alive 32 years after the first operation without any sign of metastatic disease.

Discussion

ACC is a highly aggressive and therefore lethal malignancy. Despite surgical excision as the mainstay of
primary therapy, locoregional relapse or distant metastases are common, occurring in approximately 23% to 80% of the patients with a mean disease-free interval of only approximately 12-22 months (2). In these patients, the use of chemotherapy, including mitotane, has only palliative character and eventually has to be stopped due to toxicity, providing only a limited benefit regarding survival with a mean survival of only 16-19 months and a 5-year survival rate of 0-8% (3). Only re-operation can prolong the survival and ameliorate symptoms in patients suffering from functioning tumours (4). By the use of re-operation in a selected group of patients, the mean disease-free survival time can be enhanced to 42-56 months and a 5-year survival rate of 27-57% (2). In these cases even long-term survival over decades can be achieved if the recurrences are detected early and if they are amenable to surgical excision, as can be shown in other case reports (5-8). The survival reached by other treatment modalities is in comparison rather limited (9). The update of our case report presents the longest survival of a patient with metastasised adrenal cortical carcinoma treated by multiple re-operations for metastases of the lung and of the contralateral adrenal gland, with a survival of 32 years calculated from the first operation. In conclusion, due to the high risk of recurrent or metastatic disease occurring during the further course of the disease, careful follow-up is essential for early detection of recurrent disease that may be amenable to re-operation.

References


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