Giant adrenal germ cell tumour in a 59-year-old woman

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Abstract

Adrenal germ cell tumour is very rare. We report a case of a 59-year-old woman who presented with right flank discomfort. The laboratory examinations were normal and the chest computed tomography (CT) showed right pleural effusion. The abdominal CT scan revealed a large mass on the right adrenal gland. The patient underwent an adrenalectomy. Histopathologic examination and immunohistochemical findings were consistent with mixed germ cell tumour. Three months later following the operation, the patient was admitted to our hospital again with chest tightness and shortness of breath. The chest CT showed right pleural effusion recurrence and enlargement of mediastinal lymph nodes and right hilar lymph nodes. The patient had right supraclavicular lymphadenectasis on physical examination. Fine needle aspiration cytology from the supraclavicular lymph nodes showed groups of malignant tumour cells. The patient died within 6 months postoperatively. In this case, the lymph node pathway played an important role in the metastatic procedure.

Introduction

Extragonadal germ cell tumours (GCTs) are uncommon and represent 2% to 5% of adult germ cell malignancies. The histogenesis of extragonadal GCT is unknown. We present an extremely rare case of an adrenal mixed GCT composed of dysgerminoma and embryonal carcinoma. To our knowledge, this is the first report of GCT in the adrenal gland.

Case report

A 59-year-old woman was admitted to hospital with a four-month history of right flank discomfort. Physical examinations and laboratory findings were normal. An abdominal computed tomography (CT) scan revealed a large, heterogeneous, poorly defined solid mass measuring 7.6 × 6.7 × 9.0 cm on the right adrenal gland (Fig. 1A). The chest CT showed right pleural effusion. Moreover, the thoracic drainage device was closed. There were no tumour cells in pleural effusion. A successful right adrenalectomy was thus performed on the patient. Observed by the naked eye, we partially circumscribed and encapsulated the excised tumour with maximum measurement of 9.0 cm. On the cut section, it had a uniform, yellowish, solid and partially nodular appearance within the cystic necrosis area.

Histologically, the tumour consisted of diffuse sheets, alveolar aggregates, and cords of incohesive round monotonous cells. These cells contained moderate amounts of eosinophilic granular cytoplasm and eccentrically located large round nuclei with prominent nucleoli. Cords of tumour cells were separated by fine connective tissue septa containing many lymphocytes (Fig. 2A). On immunohistochemical staining, the tumour cells were positive for human placental lactogen (Fig. 2B), c-kit, Vim, CEA, CgA, S-100, CD117 and alpha-fetoprotein (weakly positive), but negative for EMA, Syn, CD20, CD30, placental alkaline phosphatase, CK7, CK2, and a-inhibin. These results confirmed a final diagnosis of mixed GCT with dysgerminoma and embryonal carcinoma.

Initially, the tumour was considered metastatic. Postoperative ultrasonography, pelvic CT scan, however, showed no abnormal findings in the bilateral ovaries. The patient’s serum concentrations of the tumour markers, such as alpha-fetoprotein (16 ug/L), CA-125 and carcinoembryonic antigen, were within normal ranges. Chemotherapy was recommended, but the patient refused it.

Three months after the operation, the patient presented to the respiratory department with a three-day history of chest tightness and shortness of breath. The computed tomography scan revealed a large amount of pleural effusion on the right side and enlargement of mediastinal lymph nodes and right hilar lymph nodes (Fig. 1B). On physical examination she had right supraclavicular lymphadenectasis. Subsequently,
we assessed the closed thoracic drainage and performed a superficial fine needle aspiration biopsy. Tumour cells still had not been found in pleural effusion. However, fine needle aspiration cytology from the supraclavicular lymph nodes showed groups of malignant tumour cells (Fig. 2C). Adjuvant chemotherapy and fibreoptic bronchoscopy were recommended, both of which the patient had declined. Ultimately, the patient died within six months postoperatively.

**Discussion**

Extragonadal GCT was first described in 1939. Its etiology is unknown and whether extragonadal GCTs are metastatic remains controversial. There are two theories to explain this phenomenon. The first is spontaneous regression of the primary GCT after its metastasis. Possible mechanisms are an immune response or ischemia caused by the disseminated neoplasm due to its high metabolic rate. The second is the de novo development of a primary GCT in extragonadal tissues. Extragonadal GCTs share the gain of isochromosome 12p with gonadal GCTs. In rare cases, extragonadal GCTs have been associated with Klinefelter syndrome. Their anatomic distribution varies widely and includes the mediastinum, sacrococygeal region, neck, retrobulbar, retroperitoneum, and other rare sites. GCT rarely involves the adrenal gland. To our knowledge, this is the first report of adrenal mixed GCT composed of dysgerminoma and embryonal carcinoma.

In contrast to mixed GCT composed of dysgerminoma and embryonal carcinoma, primary teratomas of the adrenal gland have been reported. Our unique case differs from most reports of reported adrenal teratomas. On CT scans, teratoma is frequently shown as a heterogeneous fat dense mass with calcifications. Magnetic resonance T2 weighed images demonstrate teratoma as a highlighted intensity around the tumour components. In our patients’ case, the diagnosis of adrenal GCT was difficult to make based on the imaging studies. There were no definite radiologic features...
to distinguish primary adrenal GCT from cortisol-producing adenoma, aldosterone-producing adenoma, adrenal cortical carcinoma, and non-functioning adenoma.

Lymphatic vessels and lymph nodes may not only act as a route of spread, but also sanctuaries of metastatic deposit. Mixed GCTs are very aggressive and have early metastatic potential to the lymph nodes. It had been reported that lymph nodes were the most common site of metastasis in malignant mixed GCTs. In this case, the lymph node pathway plays an important role in the metastatic procedure. The patient had right supraclavicular lymphadenectomy three months after the operation, a chest CT revealed pleural effusion on the right side and enlarged mediastinal lymph nodes and right hilar lymph nodes. Ultimately, she died within six months postoperatively.

Although there is no standard treatment for adrenal mixed GCT, complete surgical excision of the lesions may be the only potentially curative treatment. There is little data with long-term followup; moreover, the therapy and prognosis of adrenal mixed GCTs have not been properly clarified. It has been previously reported that patients with primary extragonadal GCTs respond well to cisplatin-based combination chemotherapy, regardless of the site and presentation. For this reason, the combination of chemotherapy and surgery is the most appropriate treatment strategy.

**Conclusion**

Adrenal GCT is rare. Because of the limited number of cases, the treatment of adrenal germinoma has not been established yet. The combination of chemotherapy and surgery is the most appropriate treatment strategy.

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**References**


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