Case Report

Malignant Cervical Teratoma in an Adult Presenting with Impending Airway Obstruction

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Extragonadal teratomas and germ cell tumours are uncommon. Most teratomas of the head and neck present in the paediatric age group. Occurrence of such tumours in an adult is extremely rare and, to date, less than 40 cases have been reported in the literature. We report a case of a young man presenting with impending airway obstruction secondary to a malignant teratoma of the neck. [*Asian J Surg* 2005;28(3):227–9]

Key Words: adult, airway obstruction, cervical teratoma, rare

Introduction

Extragonadal teratomas and germ cell tumours are uncommon. Most teratomas of the head and neck present in children and such tumours are extremely rare in adults. To date, less than 40 cases have been reported in the literature. We report a case of a young man presenting with impending airway obstruction secondary to a malignant teratoma of the neck.

Case report

A 22-year-old Chinese man presented with a left anterior neck swelling of 1 year's duration. The swelling was initially asymptomatic and had progressively increased in size over the last few months. There was no hoarseness of voice but he experienced dysphagia as the swelling grew. He had some difficulty breathing, mainly on exertion, which prompted him to seek treatment. There was no previous history of surgery or irradiation to his neck.

On clinical examination, he appeared well built and not in obvious distress. There was a firm oval-shaped mass over the anterior border of his left sternocleidomastoid muscle, which measured 6 × 7 cm and extended down retrosternally. The mass moved with swallowing and it was fixed to the underlying structures. There were no palpable cervical lymph nodes. The rest of the physical examination and his genitalia were normal.

A chest radiograph showed a soft-tissue mass over the thoracic inlet compressing the trachea to the right. The lung fields were normal. Computed tomography (CT) of the neck showed a large inhomogeneous mass closely related to the left lobe of the thyroid. The mass extended inferiorly to the manubrium sterni and the trachea was displaced and severely compressed (Figure 1). The thyroid gland and the rest of the neck were normal. Fine needle aspiration cytology showed that the mass was a malignant undifferentiated spindle-cell tumour.

The patient underwent excision of the tumour under general anaesthesia. An encapsulated tumour weighing 95 g was found deep to the left sternocleidomastoid muscle. The thyroid gland was normal. However, the trachea was compressed and tracheomalacia was noted. The tumour was removed and a temporary tracheostomy was performed.

The patient recovered well. Histopathology of the tumour revealed areas of mature teratoma alternating with embryonal carcinomatous areas (Figure 2). The tumour cells were positive for cytokeratin and α-fetoprotein and weakly positive for

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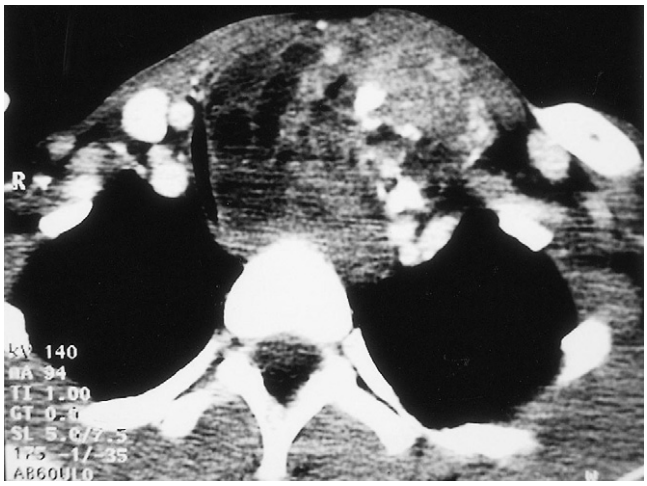

Figure 1. Contrast-enhanced computed tomography scan showing a large inhomogeneous mass below the left sternocleidomastoid muscle at the thoracic inlet compressing the trachea into a narrow slit.

β-human chorionic gonadotrophin. The patient subsequently underwent whole body CT that showed two nodules in the right lung suggestive of metastasis. In view of this, he was given bleomycin, etoposide and cisplatin (BEP) chemotherapy. He responded well with regression of the lung nodules. He was reviewed regularly and showed no evidence of tumour recurrence 2 years after his initial presentation.

Discussion

Extragonadal teratomas are intriguing as their aetiology, biological behaviour and management are uncertain. Opinions concerning their histogenesis range from a gonadal origin, where they represent distant spread, to a true extragonadal origin with separate and distinct behaviour. Generally, it has been accepted that most extragonadal teratomas arise from germinal elements distributed to sites without a primary gonadal tumour. The common sites of occurrence are along the midline or in a para-axial location from the brain to the sacral region.[1]

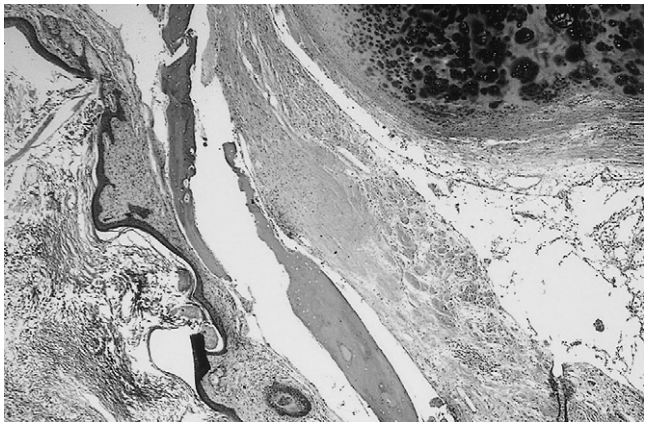

Figure 2. Section shows that the teratomatous part of the tumour is composed of mature bone fragments, epidermal cyst and hair follicles.

A great majority of teratomas of the head and neck present during infancy or childhood and are biologically benign but have a high mortality rate due to their space-occupying and obstructive character. Teratomas involving the neck account for about 3% of all teratomas in children. Teratomas in the neck of an adult is much rarer.

The clinicopathological aspects of adult neck teratoma are distinctly different from those of childhood ones. Histologically and biologically, teratomas in adults are usually malignant and their response to therapy is unpredictable. The median age at diagnosis is 32 years and ranges from 17 to 85 years.[2] It has been reported that the prognosis worsens with increasing age at time of diagnosis. Some authors have suggested that cervical teratoma in adults undergoes a rapid change from benign to malignant once they are above 20 years of age as the youngest reported malignant case was in a 19-year-old female.[3]

Classification of the teratoma based on its relationship to the thyroid gland has been attempted but is of no or little clinical significance because there are no differences in prognosis or treatment. The thyroid parenchyma within the tumour may be the remaining gland from where the tumour arose or differentiated elements arising in the teratoma itself.[4] CT of benign teratomas usually shows focal areas of fat and fluid content with areas of calcification. Malignant teratomas tend to be more lobular and visible calcification is rare. Fat density is not usually seen and it mostly appears as an inhomogeneous mass, as in our patient.[5]

The mainstay of treatment for this condition is complete surgical removal followed by adjuvant therapy. The outcome and prognosis have been dismal, with an average survival of about 8 months despite various treatment protocols. The longest survival, reported by Als et al, is more than 5 years from diagnosis.[6] They attributed their good result to giving adjuvant BEP chemotherapy immediately after surgery, in contrast to other reported cases where the adjuvant therapy was always applied late in the course of the disease when there was a large tumour burden. This case was well tabulated and compared with 23 previously reported cases of adult cervical malignant teratoma in the literature.[6]

In our case, the patient was treated with complete excision and adjuvant chemotherapy without delay. The surgery was

performed as for a thyroidectomy. We used the lateral approach to access the tumour and did not resort to a median sternotomy as the inferior edge was just at the level of the manubrium sterni. The patient responded well and we believe that the outcome for this condition would improve with better understanding of the disease and the development of better cytotoxic drugs.

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