

## Embryonal carcinoma presenting as unilateral leg swelling and weakness

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### Introduction

Primary germ cell tumours originate at various sites including the central nervous system, mediastinum, thymus, lung, kidney, testis, ovary, broad ligament, fallopian tube, vulva and retroperitoneum. Germ cell tumours of the testis account for 94% to 96% of testicular tumours, whereas female germ cell tumours constitute 4% to 6% of ovarian tumours [1]. Primary retroperitoneal germ cell tumours are uncommon but a silent testicular primary should always be excluded [2]. Mediastinal and retroperitoneal germ cell tumours occur almost exclusively in males although teratomas are seen in both sexes [1]. The patients are 15 to 35 years old and present with non-specific symptoms due to the mass or inferior vena cava (IVC) obstruction. Some are asymptomatic.

### Case history

A 19-year old male presented with swelling and weakness of the left leg of one week duration. His mother had developed fits at 36 weeks of gestation and died of disseminated malignancy with brain deposits confirmed at autopsy when the child was 1 month. Unfortunately, histology was not performed at that time. On examination he had weakness of the left leg (grade III motor loss). The sensory loss could not be assessed due to the oedema. A central abdominal mass was present. Investigations revealed an elevated ESR, normal blood counts, renal functions and coagulation profile. Ultrasound scan of the abdomen showed enlarged para-aortic lymph nodes encasing the IVC and probably compressing the left iliac vein. Liver, spleen, gall bladder, kidney and bladder were normal. Both testes were scrotal and normal. A venous doppler study of the left leg showed popliteal vein thrombosis. The IVC appeared normal with good flow. Computed tomography (CT) scan of the abdomen revealed enlarged aortic lymph nodes extending from the level of the renal hilum to the pelvis. Some nodes showed central necrosis. There was extradural extension and direct compression by tumour at the exit foramina near nerve roots, which was the reason for his leg weakness. The upper IVC and ureter were displaced laterally and the lower IVC and iliac veins were not visualised. The liver, spleen, gall bladder, kidneys, bladder and both testes were normal. The conclusion was enlarged para-aortic nodes with no evidence of a primary lesion in the abdomen or

pelvis. An exploratory laparotomy revealed a large mass of matted lymph nodes encircling the left common iliac vein and numerous discrete nodes in the retroperitoneal area. As the mass was not respectable biopsies were taken for histology.

Biopsy of a para-aortic lymph node showed an outer fibrous wall with an extensive central necrosis and nests of pleomorphic tumour cells with vesicular nuclei and prominent nucleoli with high mitotic rate. Immunohistochemical staining was strongly positive for CD 30 (ki-1) and CAM 5.2 (cytokeratin). There was no positive staining for leukocyte common antigen (LCA) in the tumour cells although LCA positive small lymphocytes were seen in the stroma. Epithelial membrane antigen (EMA), cytokeratin (AE 1/AE 3), carcino-embryonic antigen (CEA), placental alkaline phosphates (PLAP), alpha fetoprotein (AFP) and human chorionic gonadotrophin (HCG) was not positive in the tumour cells. The morphological and immunohistochemical staining pattern were in keeping with a malignant teratoma undifferentiated (MTU) according to the British testicular tumour panel classification [3] with the features of an embryonal carcinoma according to the modified WHO classification [4]. Other germ cell elements or even mature elements could be present elsewhere. Serum AFP and HCG were >5000 IU/dL. Smears for acid fast bacilli and mycobacterial cultures were negative.

Six cycles of combination chemotherapy consisting of cisplatin, ectoposide and bleomycin (PEB) were given at 3-weekly intervals. During the first cycle he developed radiculopathy and myelopathy. Magnetic resonance imaging of the spine showed direct invasion by tumour and extradural deposits and in vertebral appendages of vertebrae L2–L5. His motor and sensory loss could clearly be assessed after the fourth cycle since there was a remarkable regression of the tumour. The patient had radiotherapy to the paravertebral residual disease after completing chemotherapy. A dose of 40 Gy was delivered in 25 fractions over 5 weeks using a Co 60–Teletherapy machine.

### Discussion

The diverse views on the histogenesis of germ cell neoplasms and the wide range of their histological appearances are reflected in various classifications that have been proposed [5]. For the purposes of treatment,

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germ cell tumours are divided into two major categories: seminoma and non-seminomatous germ cell tumours (NSGCT) [6]. The NSGCT must be further classified as pure or a mixed tumour and all the components present should be reported with the approximate volumes of each component. The NSGCT include embryonal carcinoma, immature or mature teratomas, choriocarcinoma and other rare trophoblastic tumours, endodermal sinus tumour, diffuse embryomas and polyembryomas; these may be found alone or in various combinations as mixed tumours. Tumours containing both seminomatous and NSGCT components and tumours that are histologically pure seminomas but with a significantly elevated serum AFP level are regarded as NSGCTs or mixed tumours for treatment purposes.

Embryonal carcinoma "pure" constitutes 1% to 3% of germ cell tumours when yolk sac differentiation was based on AFP immunoreactivity [7]. Presence of AFP in the cell or serum is an evidence of yolk sac differentiation [8]. Microscopically embryonal carcinoma exhibits an acinar, tubular, papillary or solid pattern with areas of necrosis, haemorrhage and fibrosis. Cells are markedly pleomorphic with eosinophilic cytoplasm. Immunohistochemically cytokeratin cocktail, PLAP and ki-1 (CD30) are positive. AFP may be positive in tumour cells. HCG, CEA and Leu-M1 are negative. Cytogenetic analysis has confirmed the presence of isochromosome 12p in many cases.

There is a high cure rate obtained with modern chemotherapy even if metastases have developed. The most frequently used chemotherapy protocol for advanced NSGCTs is the combination of bleomycin, cisplatin and either vinblastine or ectoposide. After treatment a residual mass may require surgery or more aggressive chemotherapy. Adverse prognostic factors of a NSGCT include presence of an embryonal carcinoma of more than 80% of the total volume in the primary tumour, presence of vascular or lymphatic invasion, presence of a teratoma in less than 50% of the tumour [1].

Our patient had an embryonal carcinoma with an elevated serum AFP and HCG. After chemotherapy his

tumour volume subsided. The well designed PEB chemotherapy is equally effective for primary and secondary germ cell tumours. The tumour mass and the vertebral deposits were well controlled by chemotherapy, and the residual disease was helped by radiation therapy.

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