CASE REPORTS

Male choriocarcinoma with choroidal metastases

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A young man presented with bilateral solid haemorrhagic retinal detachments associated with choroidal metastases of pure choriocarcinoma, thought to be of testicular origin. The histopathological appearance and immunohistochemistry of this rare tumour are described and illustrated.

Case report

A 26-year-old man presented with a 2 day history of sudden loss of left vision, the sight in that eye having been blurred for the past 11 weeks. On examination his visual acuities were 6/6 right and hand movements left. Left subconjunctival haemorrhages were present, and ophthalmoscopy showed bilateral solid haemorrhagic retinal detachments, involving the right nasal quadrant, while the left was totally detached apart from the superior peripheral retina.

His medical history included a left orchidectomy for cryptorchidism at age 18; and torsion of the right testis, at the age of 19, which had been surgically released. Since then he had been maintained on 250 mg depot testosterone intramuscularly every 6 weeks.

Seven weeks before his ophthalmic admission he had severe chest pain and dyspnoea, when bilateral pneumothoraces were found. Following their drainage localising signs developed in the left lung. He was referred to the chest surgeons who, at left thoracotomy, found a few small clear cysts on the pleura. He was discharged home after 12 days.

One month later, on ophthalmic emergency admission, he had a normochromic anaemia with a haemoglobin of 10.7 g/dl, and an erythrocyte sedimentation rate of 45 mm/h. Chest x ray showed a large right pneumothorax with collapse, and a number of cyst-like radiolucencies near the edge of the right lower lobe. Abnormal lesions in the left lung were suggestive of metastases.

A diagnosis of choriocarcinoma involving the choroid and lung was suggested and confirmed by a positive urine pregnancy test, while human chorionic gonadotrophin (HCG) was estimated at about 20 000 IU/litre, and serum oestradiol was 354 pmol/l (normal range in men 90–220 pmol/l).

The patient was treated with radiotherapy to both orbits, and chemotherapy, but the right retinal detachment extended. Lymphangiography was performed, and blood transfusion given. Death occurred 23 days after his ophthalmic admission.

At autopsy the body was that of a tall young adult male with poorly developed secondary sexual characteristics. Both lungs had widely disseminated haemorrhagic tumour deposits, and multiple subpleural cysts. A lobulated mass (9×6 cm) was present in the anterior mediastinum at the site of the thymus. This was almost completely necrotic and haemorrhagic, with survival of tumour cells around vessels, but no

Figure 1 Portal area of the liver almost wholly replaced by cytotrophoblastic (Cy) and syncytiotrophoblastic tumour cells (Sy); a small bile duct persists (B) (haematoxylin and eosin, ×300).

Figure 2 Section of left eye: haemorrhagic detachment of retina, and tumour foci (arrowed) in anterior and posterior choroid (haematoxylin and eosin, ×3).

Figure 3 Higher power photomicrograph of tumour cells in choroidal deposit, right eye (immunoperoxidase stain with antibody to human chorionic gonadotrophin, $\times 900$).

residual thymic tissue could be found. Multiple small haemorrhagic metastatic foci were found throughout the liver, spleen, pancreas, kidneys, adrenals, thyroid, and lumbar vertebral bodies. Lymph nodes, in particular para-aortic and mediastinal, were largely replaced by necrotic haemorrhagic tumour. There was no primary teratomatous mass over the posterior abdominal wall and pelvis, or along the germinal ridge line. The left inguinal canal contained only scar tissue, and the right canal contained 7 cm of spermatic cord with fibrous coverings. No residual normal or neoplastic testicular tissue could be identified.

Histological examination showed that the tumours were all composed of pure choriocarcinoma, showing both cytotrophoblastic and syncytiotrophoblastic cells, and no other teratomatous elements were found in any of the metastases throughout the body. Both types of cell are shown in a liver metastasis (Fig 1).

The ocular pathology confirmed the clinically observed retinal detachments (Fig 2). Subretinal yellowish, lipoidal material suggested successive episodes of bleeding. The thickened choroid contained many haemorrhagic, necrotic foci, of variable size, with thin areas of surviving neoplasm. The equatorial choroid had a number of isolated tumour cells, as did the right ciliary body, but the iris seemed free from invasion. A few tumour cells were found in a scleral channel in the left eye, close to a choroidal deposit. Microscopy showed bizarre, large, irregular cells, some multinucleated, with hyperchromatic large nuclei in an eosinophilic cytoplasm. No villus formation was observed, but the two main cell types corresponded to cytotrophoblast and syncytiotrophoblast. Mitoses were rarely encountered.

HCG was demonstrable using immunohistochemistry, but α fetoprotein for yolk sac elements was negative. HCG is the product of the

multinucleated syncytiotrophoblastic cells, which are shown stained positively (Fig 3), while the cuboidal cells of cytotrophoblast are not stained. Placental alkaline phosphatase and human placental lactogen both showed occasional positive cells, reinforcing the germ cell nature of the tumour. A CAM 5.2 (cytokeratin) was strongly positive and the S 100 negative.

Comment

Pure choriocarcinoma of the testis is the most malignant of germ cell tumours. The criteria laid down in authoritative works for the diagnosis of choriocarcinoma¹ seem to have been satisfied here. Choriocarcinoma may be gestational, or, as in this case, associated with a strongly suggestive history of testicular disease, and other primary sites such as the mediastinum are considered exceptional. The mediastinal mass found at autopsy may have been a metastasis, but an extragenital teratoma of the mediastinum cannot entirely be excluded. Most cases of extragenital choriocarcinoma cannot be substantiated unless normal, scar-free testes have been demonstrated.² With no recognisable testicular tissue having been identified at autopsy, and in the light of the previous left cryptorchidism, and right testicular torsion, an undisclosed testicular tumour at that time seems the most likely primary site. Review of the histology of the orchidectomy specimen showed a small fibrosed atrophic testis with no evidence of any tumour. Other tumours on occasion may produce HCG,³ including some lung tumours.

On reviewing published reports between 1936 and 1953, six cases of choroidal metastases in choriocarcinoma of the testis were documented and summarised.⁴ Since then three more cases have been reported.⁵⁻⁷ In 1978 a clinically undiagnosed choroidal metastasis in one eye was found on histopathological examination of a case of primary mediastinal choriocarcinoma in a male.⁸

Although this patient's lung cysts were photographed at thoracotomy, the chest x ray at that time did not suggest metastatic disease. Thus the diagnosis of secondary choriocarcinoma was made ophthalmically after considering the differential diagnosis of haemorrhagic choroidal metastases, and confirmed with the positive pregnancy test.

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