CASE REPORT

Metastatic Testicular Carcinoid Tumour

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ABSTRACT

Carcinoid tumours occur predominantly in the gastrointestinal tract and bronchial system of the lungs, but they can originate in less common anatomical sites such as the testes, which accounts for <1% of all testicular neoplasms. Carcinoid tumour is a rare and indolent neoplasm with the potential for distant metastasis. We report on a man with metastatic right testicular carcinoid tumour who had multiple liver and bone metastases. The radiological and clinicopathological features of carcinoid tumours are also reviewed.

Key Words: Carcinoid tumor; Neoplasm metastasis; Testicular neoplasms

中文摘要

轉移性睾丸類癌

劉顯宇、黎永信

類癌主要發生在胃腸道及肺部支氣管，但也可原發於罕見部位，如睾丸；佔所有睾丸腫瘤的1%。類癌是一種罕見、無痛的腫瘤，可以遠處轉移。本文報告一名右側轉移性睾丸類癌病人，他同時有多發性肝和骨轉移癌。本文續討論類癌的放射學和臨床病理特徵。

INTRODUCTION

Carcinoid tumours most commonly occur in the gastrointestinal tract and bronchopulmonary system. Carcinoid tumours are uncommon tumours that are derived from neuroendocrine cells. These tumours can release serotonin and other vasoactive substances into the circulation resulting in carcinoid syndrome, especially in patients with complicated metastases. However they can also occur at rarer anatomical sites such as the testes, ovaries, breasts, and pancreas, with behaviours ranging from indolent unrecognised entities to highly active metastatic secretory tumours causing a potential diagnostic dilemma. Testicular carcinoid tumours are extremely rare, constituting <1% of all testicular neoplasms, and they rarely present as carcinoid syndrome. This report is of a man with metastatic testicular carcinoid tumour presenting with liver and bone involvement. The radiological and clinicopathological features of testicular carcinoid tumour are reviewed.

CASE REPORT

A 60-year-old man with good past health presented to Tuen Mun Hospital, Hong Kong, in 2008 with right
upper quadrant pain lasting several months. Physical examination showed hepatomegaly and a right testicular hard mass. Blood test revealed impaired liver function: bilirubin 58 μmol/L (reference range, 5-20 μmol/L), alkaline phosphatase 2691 U/L (reference range, 56-119 U/L), and alanine aminotransferase 210 U/L (reference range, 10-57 U/L). Serum α-fetoprotein, human chorionic gonadotropin-β, and lactate dehydrogenase levels were all normal.

Ultrasoundography of the hepatobiliary system showed multiple target-like lesions of 1 to 2 cm scattered throughout both hepatic lobes (Figure 1). Ultrasonography of the scrotum revealed a 7-cm well-defined isoechoic lesion at the right testis with an internal cystic component (Figure 2) and calcifications (Figure 3). Computed tomography (CT) of the abdomen and pelvis showed multiple (>10) arterial-rim enhancing hepatic lesions with capsular retraction (Figure 4). An enhancing solid lesion with a cystic component and tiny calcific foci was noted to be occupying the right testis (Figure 5). Multiple osteoblastic osseous lesions involving the lumbar vertebral bodies and sacrum and iliac bones were observed (Figure 6). No abdominal or pelvic lymphadenopathy, or any focal wall thickening was discerned along the small bowel loop, in particular at the ileocaecal region. The overall features were compatible with metastatic testicular carcinoid tumour with multiple liver and bone metastases.

Right inguinal orchidectomy was performed, with total excision of the tumour. Histological examination...
HY Lau and V Lai

confirmed the diagnosis of carcinoid tumour, showing the presence of tumour cells containing hyperchromatic nuclei with eosinophilic cytoplasm, and demonstrating strong staining with cytokeratin and synaptophysin. No teratomatous or germ cell component was discerned.

In view of the diagnosis of metastatic carcinoid tumour, the urinary level of 5-hydroxyindoleacetic acid (5-HIAA) was also measured in the postoperative period and was within normal limits. Chest radiograph showed no focal lung mass. The patient was then referred to the Department of Oncology at Tuen Mun Hospital for further treatment.

DISCUSSION

Carcinoid tumour was originally described by Oberndorfer in 1907 as a group of tumours that behaved less aggressively than conventional carcinomas. Carcinoid tumour is a type of neuroendocrine tumour that occurs predominantly in the gastrointestinal tract (74-85%), especially the appendix, and the bronchial system of the lungs (15-25%). Misdiagnosis or delay in diagnosis may occur if the carcinoid tumour arises from atypical sites or with atypical presentation.

Of all carcinoid tumours, less than 1% have been reported in the genitourinary system. Carcinoid tumour is even rarer in the testis, accounting for <1% of all testicular neoplasms. Less than 100 cases have been reported to date. Testicular carcinoid tumours occur in an age range of 10 to 84 years, with a peak in the fifth to seventh decades. The presentation is usually either a discrete testicular mass or diffuse testicular enlargement, with the left testis being more commonly involved. Testicular carcinoid tumours can be encountered in three clinical settings: as a component of a teratomatous tumour, as a metastatic lesion, or as a de-novo neuroendocrine neoplasm. Most testicular carcinoid tumours are pure primary carcinoid tumours but the other two types are more unusual. In a large serial analysis of testicular carcinoid tumours by Zavala-Pompa et al., 90% were primary tumours (either pure carcinoid tumours or carcinoid tumours associated with mature teratoma) and were rarely associated with carcinoid syndrome, which had a rate of 5.6% only. This study also showed that large tumour size (>7.3 cm), poor cellular differentiation, and presence of carcinoid syndrome were independent predictive factors for metastasis or indicators of a malignant course.

In general, carcinoid syndrome occurs in 1% of all patients with primary carcinoid tumours and in 20% of those with widespread metastasis. Vasoactive substances produced by gastrointestinal tract carcinoid tumours can enter the portal circulation and undergo metabolic degradation by the liver, thus explaining the presence of liver metastases with gastrointestinal carcinoid tumours. Other systemic carcinoid tumours are associated with a high incidence of carcinoid syndrome because the vasoactive substance can enter the systemic circulation without being degraded by the liver. It has been postulated that the low incidence of carcinoid syndrome in association with testicular carcinoid tumours is because the hormones could be secreted in an inactive form, that they are secreted at insufficient levels to cause a clinical syndrome, or that they are rapidly inactivated in the circulation. Associated testosterone deficiency and subsequent testicular atrophy had also been reported.
Macroscopically, carcinoid tumours are usually solid and yellow-brown in colour, with some masses showing cystic degeneration or calcifications, and they are typically well-circumscribed, but not encapsulated. Microscopically, the tumour cells are arranged predominantly in a nesting pattern separated by thin fibroconnective tissue. Electron microscopy commonly shows an eosinophilic granular cytoplasm, with round-to-oval inconspicuous nucleoli. No teratomatous or germ cell tumour component was identified in this patient. Immunohistochemical analysis demonstrated reactivity to chromogranin A, neuron-specific enolase, synaptophysin, and cytokeratin. The ultrasonographic features of testicular carcinoid tumour are non-specific, with most showing a well-defined solid hypoechoic mass with calcification. Although the tumour of our patient was manifested as an isoechoic mass which might be atypical, calcification was detected. This appearance simulates testicular germ cell tumour, especially teratoma, seminoma if large, embryonal cell carcinoma, and benign testicular epidermoid tumour, which are the major differential diagnoses. Carcinoid hepatic metastases are typically hypervascular in nature, as demonstrated in this patient by rim-enhancing lesions. Central umbilical depression is another unique feature indicating capsular retraction, and is due to necrosis and desmoplastic reaction within the tumour, which distorts the tumour margin and adjacent liver capsule. The prevalence of capsular retraction has been reported as 2.0% to 2.8% and is thought to occur as a result of the mechanism of local release of serotonin and other substances produced by the carcinoid tumour leading to formation of dense fibrosis and desmoplasia. However, this mechanism is not pathognomonic and has also been reported in other malignant tumours such as those of the colon, breast, lung, and gallbladder. Osteoblastic osseous metastasis is another typical feature of carcinoid tumour, as shown by this patient.

The diagnosis of metastatic testicular carcinoid tumour in this patient was based on the findings of a solitary right testicular lesion, multiplicity of lesions in the liver and bone, and absence of any discrete bowel wall thickening or mass, in particular at the ileocaecal region. A point to note is that it is mandatory to exclude the presence of a primary tumour in another organ or any other site of involvement before confirming the diagnosis of primary carcinoid tumour, due to the lack of morphological differences between primary and metastatic carcinoid tumours. There are survival implications in that metastatic carcinoid tumours have a poor survival rate. Therefore, complete staging examination with CT scan of the chest, abdomen, and pelvis, barium or capsule endoscopy study of the gastrointestinal tract, and / or octreotide scintigraphy is essential. A 24-hour urinary 5-HIAA level should also be obtained as a high level has a positive correlation with widespread disease.

The treatment of choice for testicular carcinoid tumour is radical orchiectomy, while efficacious chemotherapeutic regimens (including platinum derivatives and anthracyclines) are still lacking to date and only achieve partial responses in <10% of patients. Radical orchiectomy is usually curative for localised disease without evidence of metastases, while metastatic disease has a more unfavourable clinical course. Regular long-term follow-up at every 3 months for the first year and then yearly thereafter is also necessary because of the capacity of carcinoid tumour to metastasise. Serial estimations of serum serotonin and its degradation products in the urine, mainly 5-HIAA, are considered to be useful follow-up markers.

In summary, this report is of a patient with metastatic testicular carcinoid tumour, probably originating from the right testis given the distribution of the lesions. When multiple liver lesions with desmoplastic reaction are identified in addition to osteoblastic bone lesions, clinicians should be alert to a diagnosis of metastatic carcinoid tumour and search for the primary tumour. Apart from the commonly involved gastrointestinal and bronchopulmonary systems, other rare sites such as the ovaries, testes, and pancreas should not be ignored.

REFERENCES


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