Primary pure carcinoid tumour of the testis: A case report and review of the literature

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DISCUSSION

Primary TCTs are rare. Carcinoid tumours usually occur in the gastrointestinal tract, pancreas, lungs, or ovaries. TCTs can originate either primarily from the testis or from a mature teratoma, or metastasise from an external source (6). It is important to thoroughly investigate patients with TCTs to find or exclude an extra-testicular primary organ. NETs, which are epithelial neoplasms with predominant neuroendocrine differentiation, arise in most organs of the body (7, 8). However, neuroendocrine cells have not been described in the testes, meaning the origin of primary TCTs is debatable (1). According to the literature, primary NETs presumably originate from the testicular germ cells in the testicles (1), whereas the majority of TCTs originate from the Kulchitsky cells in the embryonic, primitive intestinal mucosa (5).

In general, the prognosis of primary TCT patients can be quite favourable. However, there are significant implications for survival as metastatic carcinoid tumours usually form part of a widely disseminated disease with a poorer clinical course (9). Histologically, it is difficult to differentiate between benign and malignant carcinoid tumours. According to the *World Health Organization's* classification of gastrointestinal neuroendocrine tumours (fourth edition) and the *European Neuroendocrine Tumor Society* (10), NETs can be divided into three groups based on Ki-67 immunoreactivity or mitotic count: Grade 1 (Ki-67, < 2%); Grade 2 (Ki-67, 3-20%); and Grade 3 (Ki-67, > 20%) (11). Grade 1 NETs have also been termed carcinoid tumours. Immunolabelling for general neuroendocrine markers, such as chromogranin A and synaptophysin, may not be necessary in histologically typical resected primary tumours (12), and immunolabelling for specific peptide hormones is only useful in highly defined circumstances. Adverse prognostic factors not included in grading and staging (e.g. vascular/perineural invasion) should be documented (13).

Primary TCTs have not yet been established. In the literature, the prognosis of TCTs and tumours associated with teratomas is better than that of pure TCTs (3). *Reyes et al.* (14) reported that when TCTs are graded using a three-tiered system and the criteria for lung NETs, patients with intermediate grade testicular NETs have the poorest prognosis. Conversely, a review of the literature revealed differing opinions on the prognostic significance of primary tumour necrosis, mitotic activity, and vascular or tunica albuginea invasion; overall, these parameters appeared to have no effect on the behaviour of TCTs, whereas a large tumour size and the presence of carcinoid syndrome were risk factors for metastatic disease (3).

Surgical management via high inguinal orchidectomy has proven to be curative for tumours confined to the testes. Metastasising TCTs that cannot be curatively resected are treated with medication. However, a standardised treatment for metastatic TCTs has not been established. In general, carcinoid tumours do not respond well to chemotherapy (15) or radiotherapy (5). With respect to carcinoid tumours at different sites, somatostatin analogues (octreotide and lanreotide) have an anti-proliferative effect on the primary tumour cells and the metastasising cells (16). Administration of somatostatin analogues may potentially control tumour growth in patients with metastatic TCTs. Metastatic TCTs have a poor prognosis, and a report of a carcinoid tumour with metastases occurring 17 years after resection of the primary tumour highlights the need for long-term follow-up (17).

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