CASE REPORT

Precocious Paratesticular Metastasis From Well-Differentiated Neuroendocrine Tumour of Ileocaecal Junction: A Case Report and Review

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Received: 26 November 2015 / Accepted: 31 January 2017 / Published online: 9 February 2017
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Introduction

Paratesticular tumours can be either benign or malignant. Malignant tumours include predominantly sarcomas like liposarcomas, rhabdomyosarcomas and leiomyosarcomas. Metastatic deposits to paratesticular region are relatively rare and account for 8.1% of all malignant tumours [1], representing a part of generalised systemic metastases. Solitary metastases are rare; even more uncommon is the precocious presentation of metastases at this site as a presenting symptom [1].

Case Details

A 45-year-old male presented with history of testicular swelling to a general practitioner. He was suspected to have a malignant germ cell tumour and underwent right orchiectomy. Pathological examination showed neuroendocrine tumour of paratesticular region and was referred to our institute for further evaluation. On histopathology review, it was a neuroendocrine tumour (grade 1) involving the spermatic cord and epididymis with the testis free of tumour; the proliferative (MIB) index was <1% with the possible source being the gastrointestinal tract. On evaluation by computerised

tomography (CT), growth was noted in the ascending colon and caecum with mesocolic lymphadenopathy. Colonoscopy revealed a polypoidal growth in the terminal ileum extending to involve the ileocaecal junction, biopsy of which was positive for neuroendocrine tumour with MIB index of 1%. Subsequent DOTATOC scan showed an increased somatostatin receptor expression in the soft tissue mass involving the ileocaecal junction representing site of the primary neuroendocrine tumour. The serum chromogranin and carcinoembryonic antigen (CEA) levels were 148.5 and 1.97 units, respectively.

The patient underwent radical right hemicolectomy. Histopathological report revealed a well-differentiated neuroendocrine tumour (grade 2) of the terminal ileum and ileocaecal junction (Fig. 1) reaching till the sub-serosa with an MIB index 8–10% of the highest proliferative areas (Fig. 2). Margins were free of tumour. There was presence of lymphovascular invasion with 5/17 nodes positive for tumour with perinodal extension. Immunohistochemical studies revealed positivity to synaptophysin and chromogranin (Fig. 3).

Discussion

The first description of spermatic cord tumour was enunciated by Lesauvage. Metastases to the spermatic cord-epididymis complex occur commonly from gastrointestinal tract (GIT), pancreas, prostate and kidneys in that order of frequency [2]. In one study [3], the source of metastases were from prostate (37%), stomach (18%), kidneys (16%), colon (13%), ileum (8%) and pancreas (5%). The most common source from the GIT is colon. Age distribution can be typically bimodal [1].

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Fig. 1 Resected right hemicolectomy specimen showing a grey white desmoplastic tumour at the ileocaecal junction (red circle)

The exact route of spread of cancer cells to the paratesticular region has not been fully elucidated. Several theories have been propounded including haematogenous route, lymphatic, retrograde vascular spread: either luminal or along the vessel wall and transperitoneal seeding via the tunica vaginalis [2]. Certain features indicate that the deposit is of metastatic origin which includes age beyond 50 years, prior history of malignancy, histology that is uncommon that of a primary paratesticular tumour, presence of lymphovascular invasion and an interstitial growth pattern [4]. Occasionally, it may be difficult to identify the exact site of spread as the epididymis, spermatic cord and tunica are contiguous structures [5].

The ileocaecal junction is the second most common site of neuroendocrine tumours affecting the large bowel, the most common being the caecum. In one study [6], 363 cases of ileocaecal and colonic carcinoids were analysed. Ileocaecal carcinoids were more common in females. They were generally large tumours at presentation, majority being greater than 2 cm in size with a high incidence of metastases (61.3%): both locoregional and distant. These tumours had a lower positivity for serotonin (on immunohistochemistry) and a low 5-year survival of around 40%.

Fig. 2 H and E stain showing nested arrangement of tumour cells

Fig. 3 IHC positivity to chromogranin

The prognosis of these tumours is uniformly poor as it heralds the presence of widespread metastases. Besides, in patients with history of gastrointestinal cancer, one of the differential diagnoses of paratesticular or inguinal swellings must be metastatic deposits. Furthermore, this can be an important and the only sign of tumour recurrence following cancer treatment, thereby underscoring the importance of thoroughly examining all male patients for any testicular or scrotal swellings during follow-up.

Although a handful of studies [7–9] have reported metachronous or synchronous metastases from carcinoma of colon to paratesticular region, a case report by Meacham et al. [10] showed a precocious presentation. Another report by O'Brien [11] also showed similar findings in the setting of an ileal carcinoid. Subsequent report by Lodato et al. [12] showed similar presentation to the scrotum.

Adjuvant chemotherapy is not indicated for well differentiated tumours and in patients undergoing R0 resection. It may be indicated for poorly differentiated tumours (neuroendocrine carcinomas) or in patients with unresectable metastatic disease. Liver involvement may warrant liver directed therapies. Follow-up of patients following complete clearance of disease includes 3–6 monthly clinical evaluation and tumour markers. Imaging is done yearly or earlier if patient is symptomatic or in the presence of elevated tumour markers.

The term 'precocious' in this report aptly describes the fact that the metastatic focus was clinically apparent prior to the manifestation of the primary tumour which was located in the ileocaecal junction. As the metastatic signature of the tumour was extra-abdominal, the paratesticular swelling became the first presentation of the disease.

Any paratesticular swelling must be thoroughly investigated for the possible site of its origin before excision. As a caveat, if the histology of any paratesticular swelling shows a neuroendocrine tumour, then the source from an intra-abdominal pathology must be investigated for before excision, as neuroendocrine tumours can rarely show extra-abdominal metastases.

Conclusions

Paratesticular neoplasms are rare with benign tumours outnumbering their malignant counterparts. Metastasis of neuroendocrine tumours to the paratesticular area is more common, often as a part of generalised metastatic disease rather than an isolated entity. If a patient is detected with a neuroendocrine tumour of paratesticular region, he should be investigated for the primary site of origin especially arising in the gastrointestinal tract. Octreotide scans are ideal to evaluate for such metastases. Surgery forms the primary modality of treatment. A long-term follow-up is essential to diagnose recurrences in these patients.

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