CASE REPORT

Asymptomatic primary rectal neuroendocrine carcinoma presented as a large pelvic mass

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SUMMARY

A 55-year-old healthy Thai man presented with incidental finding of a well-circumscribed, 5.8×5.4 cm mass in the right side of the pelvic cavity with heterogeneous density by a CT scan performed for trauma. No other distant lesion was detected. The pathology from pelvic mass showed poorly differentiated neuroendocrine carcinoma (NEC). A submucosal mass of 10 cm from the anal verge was found at colonoscopy, and a biopsy showed the same histopathology as in the pelvic mass. Final diagnosis was poorly differentiated primary rectal NEC with metastasis to the right iliac node (by CT scan). Carboplatin and etoposide were given for six cycles and tumour size was decreased.

BACKGROUND

Rectal neuroendocrine carcinoma (NEC) is typically diagnosed by colonoscopy for colon cancer screening. CT usually reveals small regional lymphadenopathy and evidence of distant metastasis. In general, lymph node metastasis is usually smaller than the primary rectal tumour. We presented the first reported case in the literature that nodal metastasis is much larger than the primary rectal NEC. From this point of view, if abdominal imaging reveals isolated pelvic mass, rectal NEC should be considered as one of the differential diagnoses and colonoscopy should be performed.

CASE PRESENTATION

A 55-year-old previously healthy Thai man underwent CT scan for evaluating blunt trauma of lower abdomen. He had mild pelvic pain but denied any gastrointestinal symptoms. A CT scan found a well-defined 5.8×5.4 cm heterogeneous density mass in the right side of pelvic cavity, near the right external iliac vessels, showing minimal enhancement (figure 1). Neither liver nor lung mass was detected by the CT scan.

INVESTIGATIONS

Enblock removal of the large pelvic mass, a 6.0×5.3×4 cm tan—brown and pale—yellow cystic mass, along the right iliac vessels was performed. Histopathology revealed a tumour encircled by fibrous tissue. The tumour exhibited nests of trabeculation of round cells with pleomorphic nuclei associated with salt and pepper chromatin, and scant eosinophilic cytoplasm, which was compatible with NET (figure 2). The tumour expresses immunoreactive for AE1/AE3, CD56, chromogranin and synaptophysin. The S-100

Figure 1 A 5.8×5.4 cm well-defined, heterogeneous density mass in the right pelvic cavity near the right external iliac vessels with mild enhancement (arrow).

protein and melan A were negative. These immunostainings were supportive for neuroendocrine in origin. Ki-67 revealed nuclear staining more than 20/10 HPFs, representing poor differentiation (WHO classification).¹ Colonoscopy showed a 2 cm submucosal mass with dilated superficial vessels at 10 cm from the anal verge (figure 3). Biopsy from this submucosal rectal mass was performed and showed the same histopathology as in the pelvic mass. Final diagnosis was primary rectal NEC with metastasis to the right iliac node by CT scan.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis includes lymphoma and primary neuroendocrine tumour of rectum with lymphnode metastasis.

Figure 2 Nests of trabeculation of round cells with pleomorphic nuclei, salt and pepper chromatin and scant eosinophilic cytoplasm.

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Figure 3 A 2 cm submucosal mass at posterior wall of rectum with dilated superficial vessels.

TREATMENT

The standard treatment in poorly differentiated NEC with metastasis is not well established. In this case, carboplatin and etoposide were given for six cycles.

OUTCOME AND FOLLOW-UP

Follow-up sigmoidoscopy and CT scan were performed 3 and 6 months after chemotherapy. No new lesion was seen from the CT scan and the tumour size was decreased.

DISCUSSION

The poorly differentiated NEC is clinically aggressive and capable of rapid distant spreading.¹ Our patient exhibits only regional lymph node metastasis. In general, lymph node metastasis is usually smaller than primary rectal tumour. To our knowledge, this is the first reported case in the literature that nodal metastasis is much larger than the primary rectal NEC.

Several factors have been reported to be associated with metastasis, including size of tumour (especially more than 2 cm), depth of invasion, evidence of lymphovascular invasion, presence of central depression and high Ki-67 ratio.² However, evidence of lymphovascular invasion and high Ki-67-ratio seem to be the most important risk factors.³ In this case, there is only one important risk factor, that is, high Ki-67-ratio. However, this rectal NEC is located in the second layer of the rectum by an endoscopic ultrasound (EUS). CT scan is able to detect only a large metastatic right iliac node, but not a primary tumour. As gastrointestinal tract is the common site for NEC, if the abdominal imaging reveals isolated pelvic mass, further investigation by colonoscopy or EUS is warranted and rectal NEC should be considered as one of the differential diagnoses. Moreover, abdominal imaging (CT or MRI) is needed to determine evidence of metastasis in rectal NEC even though the primary

lesion is limited to the rectal wall or has only one risk factor. The standard treatment in poorly differentiated NEC with metastasis is not well established.⁴ Etoposide plus carboplatin seems to be effective with the response rate around 40%, and the median survival varying from 10 to 15 months.⁵ However, the renal function and performance status of this patient were not good for cisplatin. Although carboplatin is not commonly used in poorly differentiated NEC, one other study had shown some benefit,⁶ similar to that found in our patient.

Learning points

- ▶ Typically, poorly differentiated neuroendocrine carcinoma (NEC) usually presents with distant metastasis.
- ▶ If isolated large pelvic mass is detected, primary rectal NEC should also be considered and searched for by either sigmoidoscopy, colonoscopy or endoscopic ultrasound.
- ▶ Tissue diagnosis for NEC is necessary, and palliative chemotherapy is the treatment of choice.

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Competing interests

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