Rare disease

Squamous cell carcinoma in the small intestine

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Summary

Primary carcinomas of small intestine are rare, and most are adenocarcinomas. Duodenum is the commonest site. Primary squamous cell carcinomas are exceptional. Small intestine, however, is the commonest site for metastatic spread in the gastrointestinal tract. Both adenocarcinoma and squamous cell carcinoma from a wide variety of sites can metastasise to the small intestine. The authors report a case of a squamous cell carcinoma arising in the ileum of a 65-year-old woman.

BACKGROUND

Primary carcinomas of the small intestine are rare neoplasms comprising only 0.1–1.3% of all gastrointestinal tract neoplasms.^{1–2} Most of these are adenocarcinomas. US data show an age adjusted incidence rate of 0.4 per 100 000 per year.³ The duodenum is the commonest site, accounting for more than half of all small intestinal carcinomas,^{3–4} while jejunum is a more common site than ileum.^{5–6} Adenocarcinomas comprise just over 33% of all small intestinal malignancies.⁷ However, lymphomas and endocrine tumours are almost as frequent as carcinomas.⁸ Adenocarcinomas are still sometimes described as case reports.⁹ Primary squamous cell carcinomas of small intestine are exceptionally rare with only occasional case reports in literature.^{10–11} Despite being more common, adenosquamous carcinomas are also extremely rare.^{12–13} Squamous cell carcinomas in the small intestine are more likely to be metastatic from other sites.¹⁴ We report a case of a pure squamous cell carcinoma arising in the ileum of a 65-year-old woman from Afghanistan.

CASE PRESENTATION

A 65-year-old woman from Afghanistan presented with a 1-week history of abdominal pain and cramps, abdominal distension and blood in stools. She underwent a hysterectomy with bilateral salpingo oophorectomy for carcinoma of cervix about 5 years back. On examination, she was anaemic. Tenderness was present in right lower quadrant of the abdomen, however no mass was palpable. A CT scan of the abdomen showed a soft tissue density, contrast enhancing mass, with minimal surrounding fat, in the distal ileum. It measured 3.0×2.5 cm and was causing proximal high grade small bowel obstruction. The terminal ileum and large bowel distal to the mass were collapsed. A few small subcentimetre lymph nodes were noted in the right side of the mesentery. No significant lymphadenopathy was present in the retroperitoneum, and there was no ascites or omental caking. Liver was normal, with no focal lesion. Multiple large calculi were seen in the lumen of the gall bladder, however there was no evidence of cholecystitis. Pancreas, spleen, kidneys and

adrenals were unremarkable. Uterus and ovaries were not visualised due to previous surgery. CT findings were suggestive of a primary neoplastic lesion such as a carcinoid tumour. However, the possibility of metastases could not be excluded.

Peroperatively, a hard, circumferential, obstructing mass was seen 30 cm from the ileocecal junction.

INVESTIGATIONS

The specimen was received in the Section of Histopathology, Aga Khan University, Karachi, coded as 'ileal mass'. It comprised of a segment of small intestine measuring 8×4 cm. Serosal surface was smooth, tan and glistening. On opening, a polypoid, firm to hard mass was seen arising from the mucosal aspect, causing almost total obstruction of the lumen. It measured 2.5×2.3 cm in dimension (figure 1).

The mass was 3 cm from one of the peripheral excision margins, and 5 cm from the other. No lymph nodes were recovered. Representative sections were submitted from the full thickness of the lesion with painted serosal surface and from both peripheral resection margins. Random sections were also submitted from grossly normal appearing intestinal mucosa. Histological examination showed a well-differentiated squamous cell carcinoma arising from the small intestinal epithelium and forming nests and sheets of squamous cells. (figures 2 and 3) Multiple foci of squamous metaplasia were seen in the glandular epithelium adjacent to the tumour. The tumour was infiltrating the full thickness of the wall and extending into the serosal fat. It was 0.1 cm from the painted serosal surface. However, no tumour was present at the painted serosal surface in any of the sections.

A diagnosis of infiltrating squamous cell carcinoma was made. In view of presence of foci of squamous metaplasia adjacent to the tumour, and also owing to the fact that the serosa was smooth on gross examination, and no tumour was present microscopically at the serosal surface, a primary ileal origin was favoured. However, due to the history of cervical cancer, the possibility of metastasis could not be excluded.

Figure 1 Gross photograph of segment of small intestine showing smooth, glistening serosal surface.

Figure 2 Squamous cell carcinoma arising from small intestinal epithelium. H&E×10.

TREATMENT

The patient responded well to the surgery and postoperative recovery was quick and uneventful.

OUTCOME AND FOLLOW-UP

The patient responded well to the surgery and postoperative recovery was quick and uneventful.

DISCUSSION

Primary squamous cell carcinomas of the small intestine are extremely rare, and only occasional case reports are seen in the literature.^{10 11} They may arise in congenital anomalies such as small bowel duplications and Meckel's diverticula.¹⁵ More commonly, squamous cell carcinomas may represent metastases.¹⁴ The small intestine is the commonest site for metastatic spread in the entire

Figure 3 Squamous cell carcinoma arising from small intestinal epithelium, forming nests and sheets. H&E×20.

gastrointestinal tract and secondary carcinomas are as frequent as primary carcinomas.¹⁶ The most frequent tumours which metastasise to the small intestine include carcinomas of lung, breast, kidneys and colon; malignant melanoma also commonly metastasises to the small intestine.^{14 17–19}

Other cancers which have been known to metastasise to small intestine include testes, ovaries, stomach, adrenals, uterus, cervix and liver.¹⁴

Learning points

- ▶ It is often difficult to differentiate metastatic from primary carcinomas of the small intestine, as the metastatic carcinomas closely mimic primary tumours both grossly and microscopically (in case of adenocarcinomas).
- ▶ Grossly, they are mostly submucosal or subserosal, but may form polypoid masses, and patients may present with obstruction. If the majority of the cancer cells are deep in the wall of the small intestine, and there is little involvement of the mucosa, the lesion is most likely to be metastatic.¹⁴
- ▶ In our case, the lesion was seen to arise from the epithelium with evidence of squamous metaplasia of the glandular epithelium. Also, the serosal surface was smooth on gross examination.

Competing interests None.

Patient consent Obtained.

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