CASE REPORT

Follicular dendritic cell sarcoma of the duodenum: an extremely rare entity

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SUMMARY

Extranodal follicular dendritic cell sarcoma (FDCS) is a very rare tumour, only reported in case reports and case series. It poses diagnostic and management challenge both to the clinician and pathologist. We present such a rare case of duodenal FDCS in a 56-year-old woman who was recently managed in our institution. Repeated pre surgical biopsies were non-diagnostic and the final diagnosis was made only after surgical excision of the tumour and with the help of histopathological and immunohistochemical studies. The patient had a complete en block resection of the tumour and was discharged home well 5 days postsurgery. To the best of our knowledge, this is the first case of FDCS reported arising from the duodenum.

BACKGROUND

Follicular dendritic cell sarcoma (FDCS) is a rare neoplasm that arises from follicular dendritic cells of primary and secondary lymphoid follicles. Follicular dendritic cells along with Langerhans cells and interdigitating dendritic cells are non-lymphoid, non-phagocytic cells that participate in the immune system. They are considered generally as accessory cells of the lymphoid system and primarily function as antigen presentation to B cells as well as stimulate B-cell proliferation and maturation.¹ FDCS is a relatively recently described pathology and was first reported in 1986 by Monda *et al.*² The majority of FDCS are of nodal origin and more frequent in the head and neck regions.³ Extranodal FDCS are extremely rare and can occur in head and neck, thorax and abdomen. Although there have been case reports of extranodal FDCS of stomach, spleen, appendix and pancreas,^{4–7} to the best of our knowledge, our case is the first to be described arising in the duodenum.

CASE PRESENTATION

In October 2016, a 62-year-old woman was referred to our centre by her primary care physician with a short history of an increasing upper abdominal mass, unintentional weight loss and anaemia. Apart from a previous abdominal hysterectomy for fibroid disease, the patient did not have any other comorbidities. Primary investigation with an ultrasound (US) scan of the abdomen showed an 8 cm circumscribed solid mass in the right mid-abdomen. A CT scan was recommended to further evaluate the mass. The abdominal CT axial and coronal images confirmed the large solid mass in the right upper quadrant,

measuring 9.1 cm in the longest diameter ([figure 1](#)). The epicentre of the mass was the right side of the mesenteric root. It was reported as the mass could be arising exophytically from the duodenum or alternatively might be a very large mesenteric mass. There were smaller adjacent mesenteric masses measuring 1.6 and 2.4 cm ([figure 1](#)), respectively. There was no evidence of distant metastases. Upper gastrointestinal (GI) endoscopy was performed and no mucosal lesion was noted. Colonoscopy was not performed as was deemed unnecessary in the absence of lower GI symptoms and negative CT finding for colonic pathology. Given the wide differential diagnoses and its treatment implication, attempt at tissue diagnosis were made. Unfortunately both US-guided percutaneous needle biopsy and endoscopic ultrasound (EUS)-guided needle cytology were non-diagnostic although they were suspicious for malignancy. The first US-guided percutaneous needle biopsy was performed using a 17/18 gauge coaxial system. A total of three needle passes were made with good tissue specimens obtained. The procedure required particular care due to significant transgressing intervening vascular structures. On a pathological review, however the specimen only produced predominantly necrotic tissue with scanty atypical cells. Although the scanty atypical cells were suspicious for malignancy, they were not diagnostic with certainty. It was then suggested that a fine-needle aspiration (FNA) be attempted under EUS guidance. EUS confirmed the bulky 9×8 cm well-defined and hypoechoic mass that appears to arise from the gastric wall (exact origin of mass was hard to define due to numerous necrotic spaces). Targeted FNA cytology using 22G needle were obtained in two passes. Again a lot of the specimen was necrotic material with some apparent cellular material retrieved. The cytological assessment revealed extensively necrotic specimen with only one smear slide containing occasional irregular overlapping groups of epithelioid cells (C4). This finding was suspicious but not diagnostic for GI stromal tumour (GIST).

Pathology report

A distal gastrectomy and right hemicolectomy specimen was received. There was a large 90 mm well-circumscribed tumour beneath and attached to the duodenum (D1) segment. Peripherally the tumour was white and firm, with central extensive necrosis ([figure 2](#)). The outer surface was smooth. The tumour was attached to but did not infiltrate the transverse colon.

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Figure 1 CT scan showed large solid mass in right upper quadrant (A) with two smaller adjacent mesenteric masses (B).

The tumour was composed of fascicles and nests of spindle/ovoid cells containing a moderate amount of eosinophilic cytoplasm and with nuclei demonstrating finely granular and occasionally vesicular chromatin with prominent nucleoli. Occasional multinucleated cells were noted and there was a prominent admixed inflammatory infiltrate present composed of plasma cells and lymphocytes. Numerous mitoses (average 8 per 10 high-power field) and extensive tumour-related and ischaemic-type necrosis was present. The tumour extensively replaced the muscularis propria of the duodenum with no extension into overlying mucosa and no ulceration. The visceral peritoneum and adjacent adherent colon were uninvolved. One of 25 lymph nodes were involved by a spindle cell tumour morphologically similar to the primary duodenal neoplasm, whereas a second node contained numerous dispersed multinucleated cells similar to those seen admixed within the spindle cells of the primary tumour. The surgical margins were uninvolved.

Immunohistochemistry performed revealed the tumour spindle cells to be positive for CD21, CD23, CD68, CD45, EMA and vimentin. The tumour cells were negative for cytokeratin (AE1/AE3), CD34, CD117, DOG-1, S100, CD1a, CD20, CD3, CD 15, CD30, desmin, smooth muscle actin, myeloperoxidase, lysozyme and HMB45. Epstein-Barr (EBV) (EBER-ISH) was negative. Background lymphocytes stained positive for CD3 and CD20. Ki67 proliferative index within the tumour cell nuclei

was estimated at 30%. There was no activating mutation of the BRAF V600E codon detected.

The histological and immunohistochemical features were consistent with a diagnosis of FDCS (figure 3).

DIFFERENTIAL DIAGNOSIS

GIST arising from the duodenum.

Primary GI lymphoma.

Mesenteric lymphoma.

Desmoid tumour.

Mesenteric sarcoma.

TREATMENT

The patient underwent open radical surgical excision of the entire mass in December 2016. Intraoperatively, the mass appeared to be arising from the pylorus or first part of the duodenum. It was close to the head of pancreas, superior mesenteric vein and transverse mesocolon. The operation involved en block distal gastrectomy with Billroth I reconstruction, and extended right hemicolectomy with ileocolonic anastomosis.

OUTCOME AND FOLLOW-UP

The patient made a rapid recovery and was discharged home 5 days postoperatively. She completed six-cycles of gemcitabine

Figure 2 Gross pathology (A) and cut surface (B) showing a large 90 mm well-circumscribed solid mass attached to the first part of duodenum.

Figure 3 Histological (A) and immunohistochemical (B) features, which are consistent with follicular dendritic cell sarcoma.

and cisplatin adjuvant chemotherapy successfully in June 2017. Currently she has no clinical or radiological evidence of disease recurrence.

DISCUSSION

FDCS is a rare tumour, accounting for only 0.4% of soft tissue sarcoma. FDCS often arises in lymph nodes, most commonly in the cervical, mediastinal and axillary areas.^{3 8} It is a relatively recently described entity, first reported by Monda *et al* on a series of four cases, all of which were lymph nodes origins.² Extranodal FDCS is extremely rare and has been reported in case reports in locations throughout the body including the tonsil, palate, pharynx, thyroid, peritoneum, pancreas, stomach, small intestine, colon, mesentery, mesocolon, spleen and appendix.^{4-7 9} The causes of FDCS are largely unknown. Association with Castleman disease¹⁰ and EBV infection¹¹ have been reported but have not been substantiated.

GI FDCS tumours often present as slow-growing and painless masses. Their location is variable and primary differential diagnoses should include GIST, primary GI lymphoma, desmoid tumour and mesenteric sarcoma.

Histologically, FDCS are characterised by a proliferation of oval-shaped to spindle-shaped cells with fine chromatin and ill-defined cell membranes organised in fascicles, whorls, diffuse sheets, vague nodules and occasionally a storiform pattern. It demonstrates an expression of CD21, CD23, CD35 and D2-40.⁴ Establishing preoperative diagnosis is challenging and up to one-third of FDCS tumours are initially misdiagnosed as routine immunohistochemistry panel will often not test for the diagnostic markers.¹²

Due to a lack of prospective studies, there is no established treatment protocol and therefore variable approaches have been described in the literature. However Soriano *et al* noted certain treatment response patterns in a review of 14 patients with FDCS.¹³ Eight patients who were initially treated with surgery alone or a combination of surgery and radiation relapsed shortly after treatment. Three patients received chemotherapy alone and only one had partial response. The remaining three patients treated with combination of surgery, chemotherapy and radiation as the initial treatment had the longest disease-free survival of 2 years and 5 months, 3 years and 8 years, respectively.

Although surgical resection is preferred, there is no consensus on the optimal chemotherapy regime.¹⁴ The

cyclophosphamide–hydroxydaunorubicin–vincristine–prednisolone chemotherapy regimen is perhaps the most widely used, producing some degree of response.¹⁵ However, other centres use sarcoma regime chemotherapy that includes doxorubicin, ifosfamide, gemcitabine and vinorelbine.¹⁶ The role of neoadjuvant treatment for extranodal FDCS has not been described in the literature.

Learning points

- ▶ Extranodal follicular dendritic cell sarcoma (FDCS) is rare pathological entity posing diagnostic challenge both to the clinician and pathologist.
- ▶ Diagnosis of FDCS is primarily dependent on special pathological characteristics and special immunological staining of a surgical specimen.
- ▶ Surgical excision is generally accepted as a primary treatment and the longest survivors in case series also received adjuvant chemotherapy. However there is no consensus on adjuvant therapy or type of chemotherapy regime. More research is required to better understand the pathogenesis of FDCS and develop appropriate treatment strategies.

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