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Rare Case of Reactive Nodular Fibrous Pseudotumor Causing Small Bowel Volvulus and Obstruction

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Abstract

This is a rare case of a reactive fibrosing pseudotumor that caused small bowel volvulus in a 55-year-old gentleman that resulted in small bowel resection. This case report will focus on the diagnosis of this rare cause of small bowel obstruction and compare the histological and radiological findings with that of the other commonly mistaken differential diagnoses such as GIST. Although this is a rare cause of small bowel volvulus this article provides comprehensive information across the subspecialties to aid in the diagnosis of this tumour.

Keywords: Gastrointestinal surgery; Clinical Histopathology; Clinical Radiology and imaging

Background

This is a case report of a rare clinical finding of Reactive Nodular Fibrous Pseudotumor (RNFP) that caused small bowel obstruction, and according to our literature review has only been documented 22 times worldwide. This case report will look at the histological findings of the tumour and compare these to the previous cases reported in the literature. We will also look at the radiology and see how this compares to documented and expected radiological findings outlined in other cases and highlight how PET CT benefits clinical practice when comparing differential diagnoses i.e. GIST.

Methods

This tumour presented in a 55-year-old gentleman with no predisposing factors to developing malignancy and had no other comorbidities. He had been suffering for around 4 months with obstructive type symptoms particularly after meals. A CT colon showed a 42x90mm lesion in the mid-illeal region which at first was thought to represent a Gastrointestinal Stromal Tumour (GIST). He went on to have further positron emission tomography (PET) which confirmed no distant spread of the tumour and underwent a small bowel resection with an unremarkable recovery.

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Discussion & literature review

When comparing the tumour found in our patient to that of the cases in the literature, the tumour was large, measuring approximately 120mm in maximum dimensions. The tumour was found to centre in the mesentery, extending towards both serosal and luminal surfaces. Microscopically, findings were large, plump, fibroblast-like cells set within collagenous/kelloidal stroma lacking obvious architectural pattern. There was no evidence of necrosis or vascular invasion. This tumour appeared histologically similar to the original description of the tumour by Yantiss et al in 2003, in particular, the hyalinised/kelloidal collagenous matrix. Although this case has similarities in macro/microscopic findings, it does, however, appear to match in terms of immunohistochemistry (see Table 1). There was no CD117 staining which matched the majority of the previous case reports (the main difference being the original description in 2003). Other markers stained for were all negative in our tumour. These can be seen in Table 1.

The original report by Yantiss et al (and other cases in the literature including our case report) demonstrated 100% positive results for vimentin staining in their series of 5 patients, leading diagnosis away from neoplastic diagnosis. Multiple other reports since have demonstrated similar results. Significantly, our staining was positively for SMA, both of which having reasonably high reporting in previous results and so confirmed the diagnosis. SMA, for example, was found in 72.7% of Daum et al.In this case, we did, however find differences between RNFP and GIST immunohistochemistry, which was the main differential diagnosis.With it being suggested that GISTs arise from Cajal cells, GISTs have been shown to have a high positive correlation to CD117 staining[7,8]. GISTs also have a high percentage staining for CD34 [7-9]. This is quite different from our tumour which had strongly negative CD117 and CD34 within the lesional cells.

Another condition that is important to consider when diagnosing RNFP is inflammatory myofibroblastic tumour (IMT), also known as inflammatory pseudotumors and inflammatory fibrosarcomas. They most commonly appear in the lungs, and there are well documented cases appearing in the mesentery and omentum. Macroscopic features are similar, with white/yellow firm masses, and some seconday changes of ulceration[10]. It does show a high positive immunostaining for SMA (86%), desmin (41%), CK (26%) and was found to stain for vimentin and CD117 in the submesothelial areas of the tumour 10. The tumour in this case, was not positive for either Desmin or CK.PET-CT was performed in our case (see Figure 1), as from the CT findings, there was concern regarding a possible diagnosis of gastrointestinal stromal tumour (GIST) and plans were made for surgical excision. We know that GISTs typically demonstrate CT findings that are suspicious of malignancy, it is known that GIST demonstrates high metabolic activity and thereby high fluorodeoxyglucose (FDG) uptake therefore showing “hot spots” on PET-CT. Our case did not demonstrate high FDG uptake on PET (see Figure 1) which suggests that PET-CT may be a useful imaging method for differentiating RNFP and GIST before biopsy. As this is a one-off case, further studies would need to be performed to look at the relationship between RNFP and PET-CT scanning, however this may be difficult due to the low number of cases of RNFP.

Learning points

This is a case report of a rare clinical finding of reactive nodular fibrous pseudotumour (RNFP) that caused small bowel obstruction, and according to our literature review has only been documented 22 times worldwide.

This case report will look at the histological findings of the tumour and compare these to the previous cases reported in the literature.We will also look at the radiology and see how this compares to documented and expected radiological findings outlined in other cases and highlight how PET CT benefits clinical practice when comparing differential diagnoses i.e. GIST.

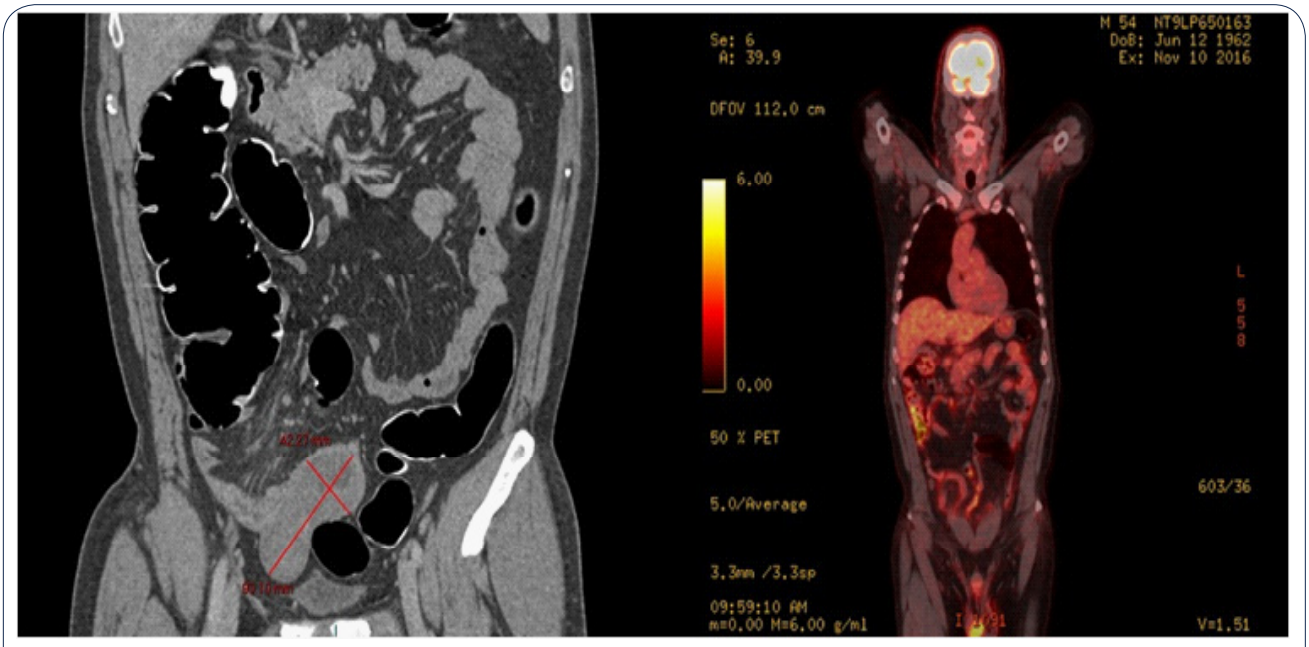

Figure 1: A comparision of the radiological finding of this case. Left is CT colon clearly showing the tumour and volvulous and to the right the PET CT that showed no “hot spots”.

Table 1: A review of all of the histological findings of the other reported cases of RNFP [1-6].

Author	Year	Case no.	Vim	CD117	CD34	CK	SMA	Desmin
Yantiss	2003	1	+	+	-	-	+	+
		2	+	+	-	-	+	+
		3	+	+	-	-	+	+
		4	+	+	-	-	-	-
		5	+	-	-	-	-	-
Daum	2004	1	+	-	-	+	+	
		2	+	-	-	+	+	
		3	+	-	-	+	+	
		4	+	-	-	+	+	
		5	+	-	-	+	+	
		6	+	-	-	+	+	
		7	+	-	-	-	+	
		8	-	-	-	-	+	
Saglam	2005	1	+	ND	+	+	+	-
Gauchotte	2009	1	+	-	-	+	+	-
McAteer	2012	1	+	-	-	+	+	
Yi	2014	1	ND	-	+	ND	+	-
Salihi	2014	1	ND	ND	ND	ND	ND	ND
Ciftci	2015	1	+	-	-	-	+	-
Yan	2015	1	+	+	-	+	ND	+
This Case	2017	1	+	-	-	-	+	-
		Total	77.30%	22.70%	9.10%	45.50%	72.70%	

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