Granular cell tumour of the appendix: an unexpected location

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DESCRIPTION

Granular cell tumour (GCT) is an uncommon but usually benign lesion of neural/Schwann origin. This tumour type is most frequently found in skin, subcutaneous tissue and oral cavity, mostly in black middle-aged women.[1] This lesion type can involve any organ, including the gastrointestinal (GI) tract. The appendicular involvement though is extremely rare.

We report a case of a pregnant female patient in her early 30s who went to the emergency department after 2 days of enduring persistent pelvic pain. The blood tests indicated leucocytosis, neutrophilia and elevated C reactive protein concentration.

Abdominal ultrasound examination revealed an outer appendiceal diameter of 8 mm, with a markedly thickened and stratified wall. The increased density of the surrounding fat tissue was consistent with phlegmon.

A laparoscopic appendectomy was performed.

Careful gross examination showed that the appendix was increased in size, the appendiceal wall was markedly thickened and the lumen filled with faecal material.

The complete histological analysis of the specimen revealed a well-circumscribed and unencapsulated nodule with 0.5 cm in diameter confined to the submucosa. There was also a florid granulomatous chronic inflammation measuring 1.2 cm extending to the tip of the appendix (as shown in figure 1A). This granulomatous inflammation was located in the wall and involved its entire thickness, except the overlying mucosa. The multiple, coalescent microgranulomas (non-necrosing granulomas) were composed only of epithelioid histiocytes. The authors want to emphasise that there was no clinical or imaging procedure of the thorax, which may constitute evidence for tuberculosis diagnosis. The surrounding tissues showed reactive and hyperplastic mucosa, with no atrophy. There were no acute appendicitis lesions.

The nodule was composed of nests of epithelioid cells with abundant granular eosinophilic cytoplasm and central small round nuclei (as shown in figure 1C). The tumour was devoid of cytological atypia and necrosis.

Immunohistochemistry revealed S-100 protein strong and diffuse positivity, suggestive of Schwann cell origin, and it was not expressed by the granulomatous epithelioid cells. The GCT cells also displayed SOX-10 nuclear expression (as shown in figure 1D) and CD68 membrane expression. CD68

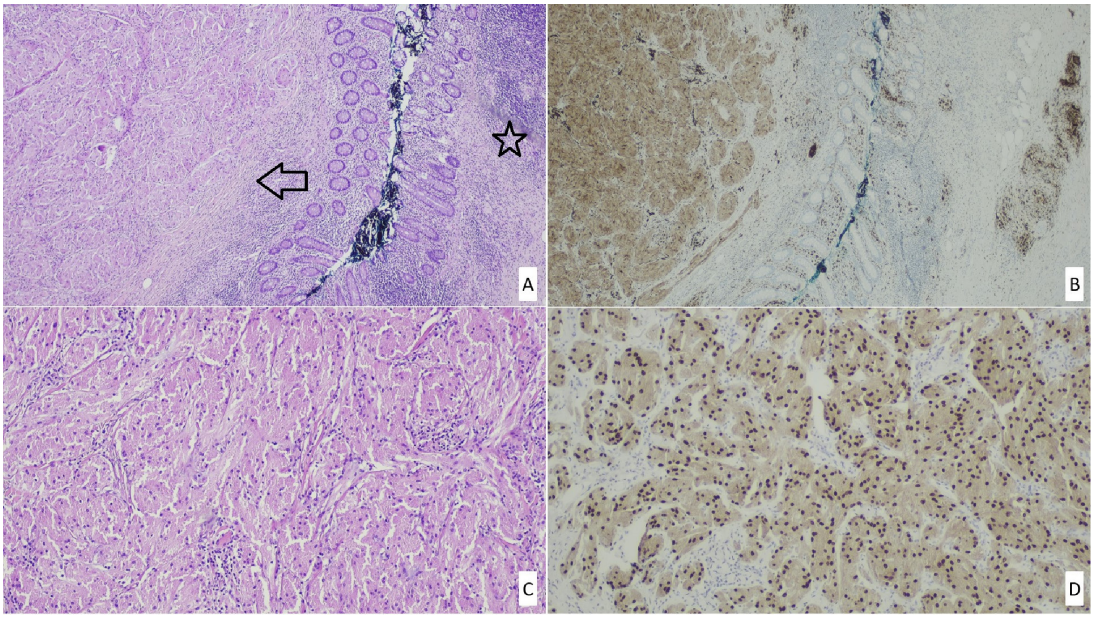

Figure 1 (A) Submucosal GCT (arrow) was contiguous with a chronic granulomatous inflammation zone (star), (H&E, ×20). (B) The tumour cells and granulomas without necrosis found in the appendix wall were immunoreactive for CD68. (C) The round to oval tumour cells displayed a granular cytoplasm (H&E, ×100). (D) The neoplastic cells typically expressed nuclear SOX-10 (×100). GCT, granular cell tumour.

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Table 1 Clinical characteristics of patients reported in the literature with GCT of the appendix

Case number	Author(s), year	Age, gender	Clinical symptoms	Radiology assumption	Size of tumour (cm)
1	Wanick, 1956[4]	30s, female	Acute pain in right iliac fossa, vomiting and diarrhoea	Acute appendicitis	0.5 and 1 (double nodules)
2	Hausman, 1963[5]	40s, male	NP	NP	0.8
3	Apisarnthanarax, 1981[6]	30s, female	NP	NP	5
4	Sarma *et al*, 1984[7]	50s, male	Asymptomatic	NP	0.5
5	Fried *et al*, 1984[8]	30s, male	NP	NP	0.8
6	Pipeleers-Marichal *et al*, 1990[9]	40s, male	Asymptomatic	NP	4
7	Kaltschmidt *et al*, 1992[10]	30s, male	NP	Acute appendicitis	NP
8	Gavelli *et al*, 2005[11]	40s, male	Acute pain in right iliac fossa and fever	Acute appendicitis	0.5
9	Moreno Gijon *et al*, 2009[12]	30s, female	Acute pain in right iliac fossa	Endometrioma	NP
10	Saleh *et al*, 2009[13]	60s, female	Asymptomatic	NP	0.5
11	Singhi and Montgomery 2010[14]	40s, female	NP	Acute appendicitis	0.6
12	Zoccali, 2011[15]	late adolescence, male	Acute pain in right lower quadrant	Abscess	3.5
13	Roncati *et al*, 2013[1]	40s, female	Acute pain in right lower quadrant, pain at defecation, tenesmus, rectal bleeding and haematuria	Phlegmon	0.2
14	Roy and Goswami, 2015[16]	20s, male	Acute pain in right iliac fossa and vomiting	NP	NP
15	Allison and Rao, 2020[2]	50s, female	NP	NP	NP
16	Current case	30s, female	Acute pelvic pain	Phlegmon	0.5

Source: PubMed.
GCT, granular cell tumour; NP, not provided.

pointed out to the granulomas found in the appendix wall (as shown in figure 1B).

The postoperative period was uneventful, and the patient was discharged after 1 week.

Less than 15% of GCTs occur in the GI tract, with only 15 cases being reported in the appendix so far.[1 2]

A detailed analysis (table 1) of the features of GCTs in the appendix reveals that the male/female ratio was 1:1, the mean age was that of 41 years old and the average size of the lesions was 1.4 cm. The patients' clinical characteristics, including from the patient reported in this article, can be consulted in table 1.

The pathogenesis of GCTs remains unclear; however, the medical literature data suggest that a chronic inflammation, surrounding the GCT, is an antecedent condition that may favour its appearance.

In fact, chronic and unresolved inflammation promotes immunosuppression and so it has been associated with an increased risk of developing both benign and malignant tumours.

Furthermore, published reports indicate that GCT in the appendix is a lesion which reflects local reactive changes of the neural/Schwann cells rather than a true neoplasm.[3]

This case documents a GCT probably arising from a granulomatous appendicitis background, which is a rare entity. This chronic inflammation corresponds to less than 2% of appendectomies as a cause of appendicitis.

Surgical intervention with wide local excision and complete resection is still the best treatment for GCTs in the GI tract, with excellent outcomes for benign lesions and with a low rate of recurrence or malignant transformation.

Learning points

- Granular cell tumour (GCT) is an uncommon benign lesion of neural/Schwann origin.
- The appendicular involvement is extremely rare.
- We report a case of granulomatous appendicitis and a GCT as an incidental finding, whose symptoms and analytical parameters mimic an appendicitis.

There are no data in the literature that correlate with the emergence of GCT in the lower GI tract and the risk of developing colorectal cancer.

Previous reports suggest that regardless of the initial nature of the lesion, all patients should be followed to prevent recurrence and distant metastasis.

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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