Journal of Surgical Sciences (2021) Vol. 25(2) : 52-55

JOURNAL OF SURGICAL SCIENCES

Case Report

A Retroperitoneal Granular Cell Tumour Mimicking Gastric Outlet Obstruction

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Abstract:

Granular cell tumour (GCT) is a rare entity which is thought to be of neural origin. It is usually benign in nature with characteristic histological features. The tumour can occur anywhere in the body with common sites being the tongue, skin, subcutaneous tissue, breast, etc. But a retroperitoneal GCT is extremely uncommon. We report the case of a 50 years old male with a retroperitoneal lump with features of gastric outlet obstruction. Ultrasonography and CT scan report were inconclusive. The tumour was completely resected and histologically confirmed as GCT. The immunohistochemistry came positive for S100. Though rare, retroperitoneal GCT can be encountered in surgical practice posing diagnostic difficulty. This case report may guide its effective management.

Keywords: Granular cell tumour, Retroperitoneal, Gastric outlet obstruction, Schwann cell, Immunohistochemistry

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Receided on: 28.08.2021 **Accepted on:** 30.08.2021

Introduction:

Granular cell tumour (GCT), also known as granular cell myoblastoma is a rare variety of mesenchymal neoplasm thought to be originating from Schwann cell.[1] The tumour is usually benign except for some rare malignant cases.[2] The women are more commonly affected.[3] Histopathology is the gold standard for diagnosis of GCT which is characterized by large neoplastic mass with unclear boundaries, neoplastic cells distributed

between collagen bundles, and granular eosinophilic cytoplasm.[4] The tumours often show positivity for S-100, CD-57, Neuron-specific enolase (NSE), etc supporting its Schwann cell origin.[5] Because of the slow-growing nature of the tumour, it produces few clinical symptoms and presentation mainly depends on the location of the tumour.[4] GCT commonly occurs in the head and neck region with the tongue being the most common area, but it can also occur in the skin, subcutaneous tissue, and breast, gastrointestinal, biliary, and respiratory tract.[6] Multiple tumours have been reported in 5% of cases.[7] However the GCT originating at a retroperitoneal site is extremely rare.[8]

Here, we present a case of benign retroperitoneal gastrointestinal tumour where the patient presents with the clinical features of gastric outlet obstruction and underwent complete surgical excision with a favourable outcome.

Case Report:

Mr. Abu Bakar Siddique, 50 years old got admitted to a local hospital with the complaints of early satiety, nausea, and epigastric fullness following meal for 6 months, occasional vomiting for the same duration and unintentional weight loss for the last 3 months. On clinical examination, the patient was found undernourished with normal vital signs. The abdomen was normal in size and shape with no visible lump or peristalsis. On palpation, the abdomen was soft, non-tender with no palpable masses and there was no palpable lymph node. His other systemic examination was unremarkable. Ultrasonography showed a thick walled SOL containing thick fluid in the right hypochondriac region. CT scan report revealed a large mixed density mass in the right hypochondrium with central necrosis and multiple peripheral calcific foci. His laboratory findings and other reports were within the normal limit.

Laparotomy was done by midline incision. On opening of the peritoneal cavity, there was no ascites or other abnormality. A solid retroperitoneal mass was found measuring about 7cm in length and 5 cm in breadth. The mass was well-capsulated and it compressed the 2nd part of the duodenum medially and inferior vena cava posteromedially. The tumour was resected completely with no injury to the surrounding structures. His postoperative period was uneventful and the patient was discharged on 3rd POD.

On gross examination, the specimen was a globular solid mass of about 7x5x4.5 cm in size with smooth surface. Cut section revealed an

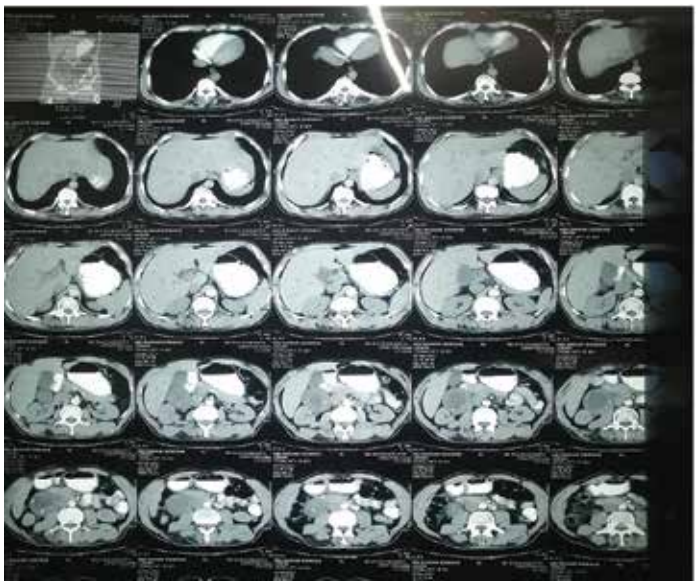

Figure 1: CT scan showing the retroperitoneal mass

Figure 2: The resected tumour

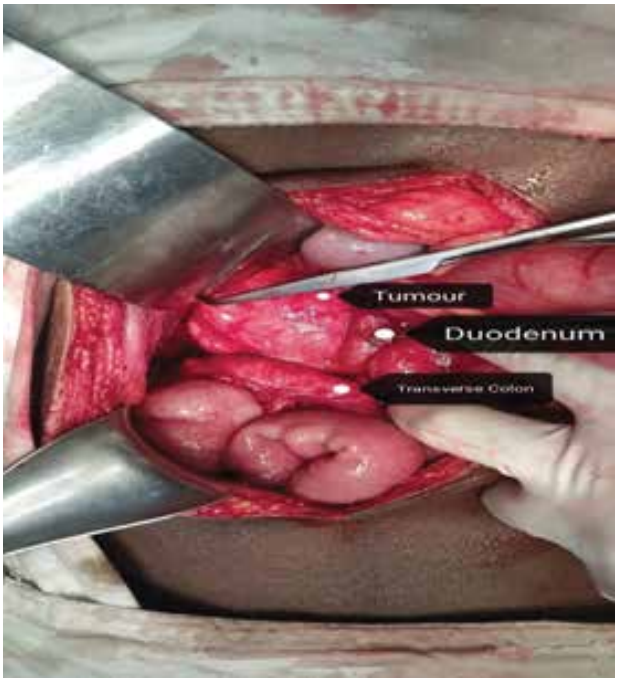

Figure 3: Per operative view of the tumour

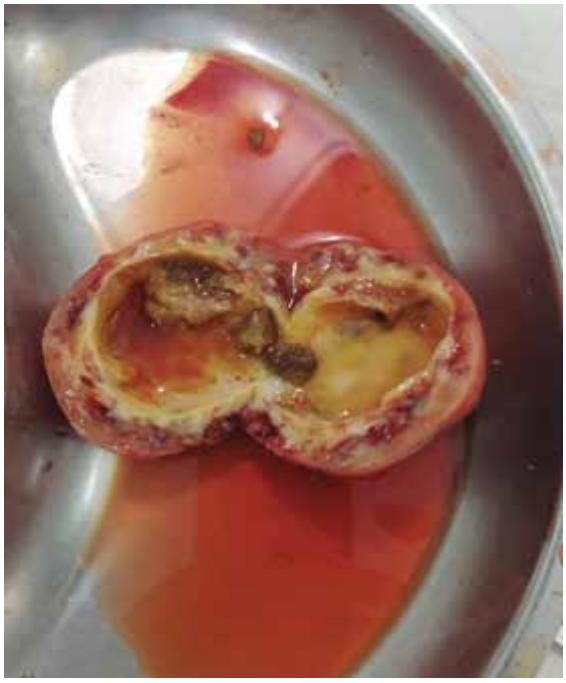

Figure 4: The cut section of the tumour

encapsulated greyish yellow cyst with central necrosis.

Microscopic examination revealed fibro-collagenous tissue with cystic degeneration lined by large cells with abundant granular eosinophilic cytoplasm and centrally placed nuclei. The cyst wall contains areas of haemorrhage, necrosis, and dystrophic calcification.

Immunohistochemical stain of tumour cells was positive for S100 but negative for CD68.

The patient was followed up for a period of 2 years and no recurrence or malignant conversion was detected.

Discussion:

Granular Cell tumour was first described by a Russian pathologist in 1926 named Alexei I Abrikossoff.[9] GCT is prevalent between 40 to 50 years of age with the male to female ratio being 1:3.[10] The GCT is known to be derived from the neural cell having the histological composition of granular eosinophilic cytoplasm.[11] Due to its slow-growing nature it hardly produces any clinical symptoms unless too large.[4] Till to date very few cases of GCT have been reported and among them, cases of retroperitoneal GCT are very unusual.[8]

In 1996, Varma DGK et al.[6] first reported a benign retroperitoneal granular cell tumour in a 38-year-old female diagnosed by CT scan which was inoperable and chemotherapy was ineffective. A 5-year follow-up of the patient revealed no untoward outcome.

Lee EJ et al[8] also reported a retroperitoneal tumour in a 46-year-old male mimicking pancreatic cancer in 2008 which was diagnosed incidentally by CT scan and MRI. The patient was treated surgically and confirmed histologically to be GCT with S100 being positive.

Our patient is a 50 years old male in whom the compression effect of the retroperitoneal mass produced clinical symptoms of gastric outlet obstruction for which he sought surgical consultation.

The USG report and CT scan findings of our case were non-specific making it difficult to confirm the diagnosis preoperatively. The same dilemma was reported by the study of Varma DGK et al[6] who found a homogenous mass in the CT scan. But Lee EJ et al[8] reported a heterogenous mass with central low attenuation in CT scan which was further evaluated by MRI. However, in all cases, imaging could not guide a specific diagnosis.

GCT is predominantly benign with only 2% being malignant.[12] Fanburg Smith et al[13] proposed the histologic criteria of malignancy where at least three of the six criteria needed among (i) necrosis, (ii) spindling of tumor cells, (iii) vesicular nuclei with large nucleoli, (iv) increased mitotic rate (more than 2 mitoses/10 HPF), (v) high nuclear to cytoplasmic ratio, and (vi) pleomorphism and those with less than three criteria were termed atypical. Our case lacks most of the criteria marking it to be benign. However, it has been reported that histologically benign-appearing tumors can undergo malignant transformation and metastasize.[14] That is why follow-up for recurrence or metastasis is crucial for effective management.

The pathogenesis of GCT has been controversial for long until recent immunohistochemical studies suggest its origin to be from the Schwann cell.[15] The positive immunohistochemical stain for S-100, NSE and CD57 confirms the theory. But S-100 negativityy doesn't exclude it.[16] Again expression of vimentin and CD68 is caused by aggregation of the lysosomes.[17] In our case, the immunohistochemical stain came positive for S100 confirming it to be GCT of Scwann cell origin.

Ours is one of the few reported cases of benign retroperitoneal granular cell tumours that despite its preoperative diagnostic uncertainty was managed effectively.

Conclusion:

The granular cell tumour is a rare lesion with its retroperitoneal location being uncommon. Moreover, its epidemiology, clinical symptoms and radiological features are variable. But still, surgeons can encounter them during their clinical practice which may be difficult to diagnose. Whatever might be the symptoms, granular cell tumour should be considered in the differential diagnoses of retroperitoneal masses and treated surgically and malignancy should be ruled out with adequate follow-up. This case report may be a valuable guide in dealing with such cases.

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