Case Report

Inter-Sphincteric Resection of Rectum for Ulcerated S-100 Negative Large Granular Cell Tumour

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Abstract

Non-neural granular cell tumor was first described in 1991 as an unusual primitive, polypoid variant of the conventional GCT (granular cell tumor). To date, this neoplasm remains as a rare entity and the cell of origin is uncertain. While the histological features are similar to the conventional (traditional) GCTs, it represents a distinct entity, as it is negative for S-100 and lacks true nerve sheath differentiation. We report here a case of a 49-year-old male patient with S-100 negative, vimentin positive large benign, ulcerated, bleeding non-neural GCT in the lower rectum. We confirmed the diagnosis histopathologically and immunohistochemically. We treated it by ISRR (Inter-Sphincteric Resection of Rectum) and primary colo-anal anastomosis. We find no other report of non-neural GCT located in the rectum in surgical literature. This is probably the first case of non-neural GCT identified in the rectum. This case report is presented here to document a non-neural GCT located in the rectum for the first time ever reported, and also to highlight the optimum best surgical approach, as we deemed here for this large ulcerated bleeding tumor in the rectum. This rectal non-neural GCT posed here as an unusual differential diagnosis from such more common neoplastic bleeding lesions in the rectum as carcinoma rectum, hemorrhoid, rectal polyp, etc. Here we like to discuss our experience, reviewing in brief the available literature implicating the biological behavior, histopathological and immunohistochemical features, differential diagnosis and therapeutic considerations of neural and non-neural GCTs.

Key words: *Abrikossoff tumour, S-100 negative granular cell tumor, Vimentin positive granular cell tumor.*

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Introduction

Apart from circulating granulocytes (having primary azurophilic granules found in young cells and secondary specific granules found in more mature cells), definite pathological granular cells have been described since 1926 as causation of GCTs (the granularity of the latter being due to accumulation of secondary lysosomes). This pathological nonspecific granularity can be seen in many neural and non-neural tumors, including those arising from smooth muscle, connective tissue, neuroglia, endothelial, and epithelial cells, etc. GCTs are very uncommon STTS (Soft Tissue Tumors). They (also known as Abrikossoff tumors) are rare, mostly benign tumors, now thought to be originated from Schwann cells. A conventional GCT was fist described by the Russian pathologist Alexei Ivanovich Abrikossoff in 1926.[1,2] The GCTs are commonly found in the oral mucosa, specifically in the tongue, whence the first case was also named as granular cell myoblastoma. Later on, their origin from neural crest ectodermal Schwann cell was confirmed by immunohistochemical staining, and thence they are sometimes called granular cell Schwannoma, or occasionally granular cell neurofibroma. Thereafter, non-neural GCTs have also been identified in different locations of the human body. The first

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case of this later variant was discovered by LeBoit et al in 1991.[3] Because of primitive polypoid phenotype, the first case had been given the term PPGCT (Primitive Polypoid Granular Cell Tumor). After that the term PNNGCT (Primitive Non-Neural Granular Cell Tumor) was universally accepted, that also includes the non-polypoid non-neural GCTs, which are collectively quite different form conventional GCTs of neural differentiation.[4,5] Many forms of PNNGCT have been detected that include plaque-like or polypoid cutaneous variant, an intra-oral variant, a congenital variant and a variant developing in the uterus and a pulmonary variant, etc. The PNNGCTs are less common than conventional GCTs.[6,7] These are described as low-grade tumors, probably of mesenchymal origin of unknown family with promiscuous lysosomal granules. Until now, multiple case reports have been published, describing the neural granular cell tumors located in the rectum, but no case report has yet been found for any non-neural granular cell tumor located in the rectum. Our case report appears to be the first one ever reporting non-neural granular cell tumor located in the rectum.

Case Presentation

A 59-yrs old male married, non-alcoholic, non-smoker, non-chewer of betel-nuts, non-asthmatic, ill-looking anxious farmer of average body-build with no such co-morbidity as hypertension, diabetes mellitus, heart or cerebro-vascular disease, no significant weight loss, hailing from a lower middle class Muslim family of Khilkhet, Tangail was admitted on the 10th December, 2021, with a provisional diagnosis of carcinoma rectum, having complaints of intermittent per-rectal bleeding and lower abdominal colicky pain, weakness, and and no constipation, but with feeling of incomplete defecation for about two years. The pain had been more severe during defecation, and not significantly affected by traditional OTC analgesic medications and any dietary habits. None of his family members suffered from this type of illness. He was not allergic to any known diet, drugs, pollens or any other known allergens and food items. He had mild anemia, no jaundice, no cyanosis, no clubbing, no koilonychia, no leuconychia. His neck and other superficial veins were not engorged. His accessible lymph nodes, heart, lungs, testes, thyroid glands, breasts seemed to be normal. His hernia orifices were intact. His temperature, respiration rate and blood pressure were normal, with tachycardia (pulse rate 104 per minute) at admission. His abdomen was normal in size and shape with no distension, no visible peristalsis, no abnormal bowel sound. His liver, spleen and kidneys were not palpable. On digital per rectal examination, his anal tone was found intact with a large ulcerated lesion in the left postero-lateral wall of rectum within five cm of the anal verge. The ulcer appeared was not circumferential, with no finger-like or cauliflower-like projection& not fixed to any surrounding structures. No peri-rectal lymph-node was palpable. Finger was moderately blood-tinged with no active bleeding. Provisionally it appeared as a case low grade ulcerated carcinoma rectum involving its mostly lower third. Total colonoscopy showed the mucosa and vascular pattern all through normal seen up to caecum with a large ulcer in the rectum. Colonoscopically, the ulcer seemed to be non-specific with no raised and no irregular edge. Colonoscopic biopsy from the rectal ulcer showed fibro-purulent exudates mixed fragments of rectal mucosa with underlying acute on chronic non-specific inflamed granulation tissue, no epithelioid granuloma, no malignancy, suggestive of Chronic non-specific ulcer. CT Scan of whole abdomen with screening of Chest showed Irregular calcification at irregularity – suggestive of inflammatory changes, and Cystic bronchiectactic changes, fibrosis with somewhere bronchial wall thickening, associated with patchy ground glass opacity noted at apical-posterior segment and middle lobe of right lung and posterior segment of left lung. A thick-walled tiny cavity lesion (13 mmx10mm) having outer irregular margin noted at posterior segment of right lung--chronic inflammatory change - simulating tuberculosis, subcentimetric mediastinal lymph-nodes, largest one measuring about 8.9 mm at subcardinal region. Chest X-ray P/A view showed normal bony thorax, mild cardiomegaly with LV preponderance, lung fields with chronic inflammatory lesions were suggestive of bilateral bronchiactasis and bilateral upper lobar opacity simulating tuberculosis or lung metastases, central trachea, normal diaphragm, ill-defined left dome with obscured left costophrenic angle probably due to fibrosis and bronchiectatic lesions. Sputum for AFB were negative in all samples. ECG and echocardiographic findings were within normal limit. USG of whole abdomen showed mildly thick rectal wall indicative of a suspicious Lesion and mildly enlarged prostate.

His other laboratory results were as follows: total red cell count : 3.32x1012/L, Hb: 9.5gm/dl , Platelet count: 307x109/L, total leucocyte count: 8.32x109/L, ESR:31mm in 1st hour; PBF (Peripheral Blood Film) : RBC-Normocytic normochromic, WBC: Mature with normal count and distribution with relative neutrophilia; Platelets: Adequate; Parasite: Not found; Random Blood Sugar: 5.52 mmol/L; HbA1C: 5.5; S creatinine: 85.5 micromol/L; S Albumin: 40.93 gm/L; S Alkaline phosphatase: 55 U/L; S Calcium: 2.29 mmol/L; S ADA (Adenosine deaminase): 21 U/L; ICT for tuberculosis: Negative; Total Bilirubin: 10.35 micromol/L; SGPT/ALT: 12 U/L; Alkaline Phosphatase: 56 U/L; S Electrolytes:- Na+: 138.18 mmol/L, K+: 3.71 mmol/L, Cl-: 100.52 mmol/L, HCO3-: 22.30mmol/L; Bleeding time: 02min 04sec; Clotting Time: 13 min 14 sec; S CEA 3ng/ml; S TSH: 0.52 IU/ml; HBsAg(C): Negative; Anti-HCV: Negative; Anti-HIV (l & ll): Negative; VDRL/RPR: Non-reactive; S Magnesium: 0.62 mmol/L ; RT-PCR for covid-19: Negative.

After adequate counseling and SDM (Shared Decision Making), ISSRR (inter-sphincteric resection of rectum (Fig. 1 showing specimen of resected rectum) and primary coloanal anastomosis were done under general anesthesia on the 13th December, 2021.

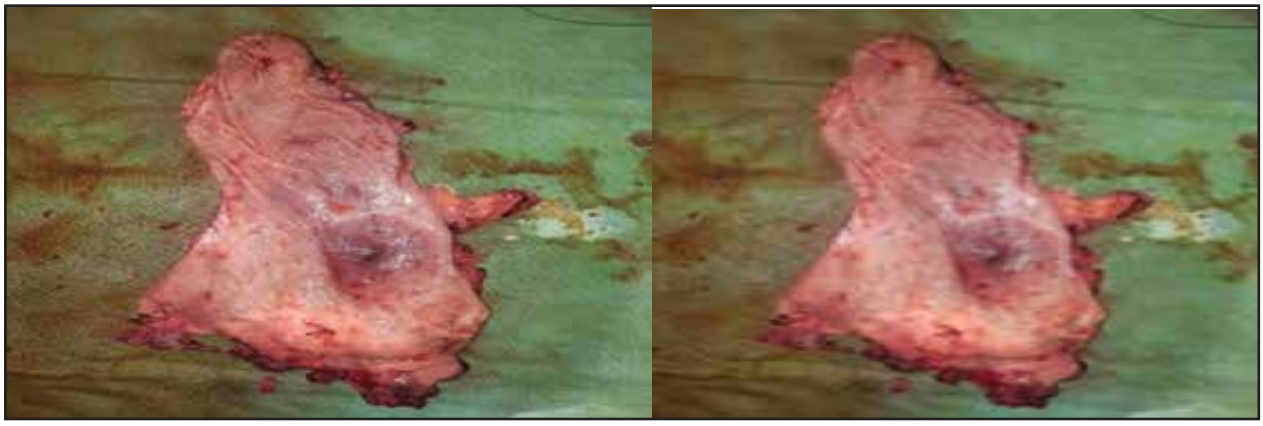

Figure 1: Specimen of ISRR (Inter-sphinteric resection of rectum) for a large rectal ulcer of granular cell tumor.

Grossly,¬¬ 16 cm long piece of resected gut (rectum and adjoining parts) showed the rectal mucosa with a 5x5 cm sized ulcer and the peri-ulcer adjoining thick areas. Histopathology examination of rectal wall showed proliferation of monomorphic granular cells having abundant granular eosinophilic cytoplasm with homogeneous nuclei. These cells were arranged in sheets & lobules with some multinucleated forms. No pleomorphism, mitosis or necrosis were seen (Figs. 2 & 3).

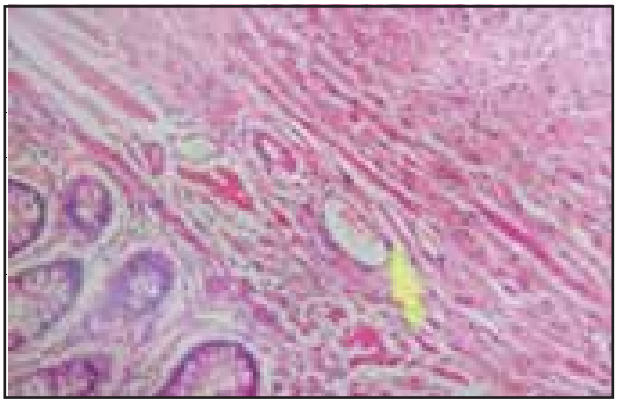

Figure 2: H & E stain of non-neural GCT of rectum under low power.

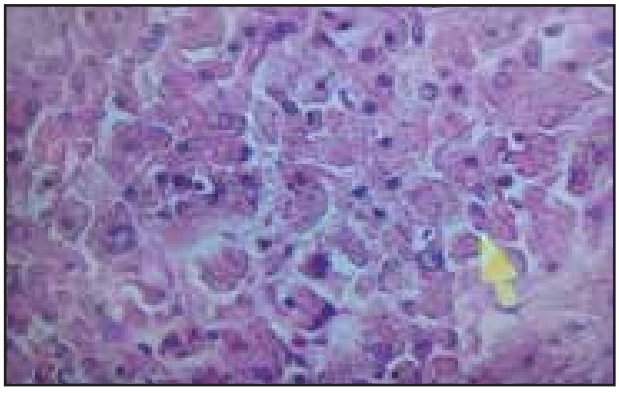

Figure 3: H & E stain of non-neural GCT of rectum under high power.

Immuno-histochemical staining showed that the neoplastic cells have immunophenotype S-100: Negative (Fig. 4), & Vimentin: Strongly positive (Figs. 5a and 5b). Thus immune-morphologically, our tumor appeared to be consistent with a non-neural GCT.

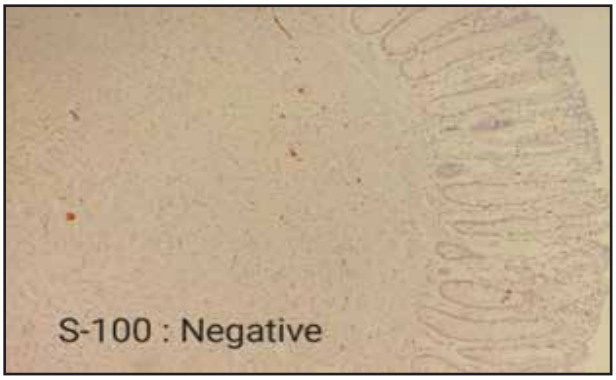

Figure 4: Immunohistochemistry (IHC) where the neoplastic cells showed immunophenotype: S-100: Negative, with non-neural granular cell tumor.

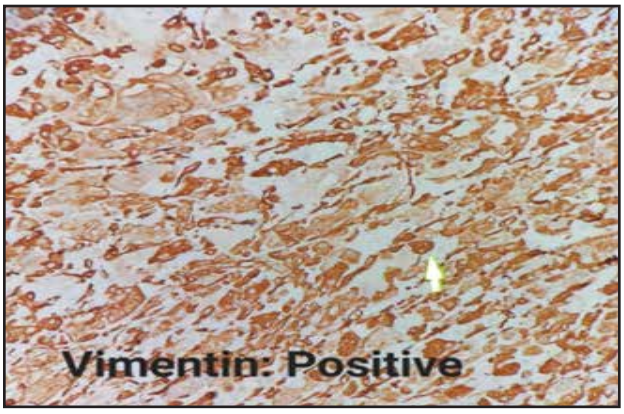

Figure 5a: IHC (Immunohistochemistry) where the neoplastic cells showed Vimentin: Strongly positive (+++), thus immune-morphologically, it is consistent with non-neural granular cell tumor

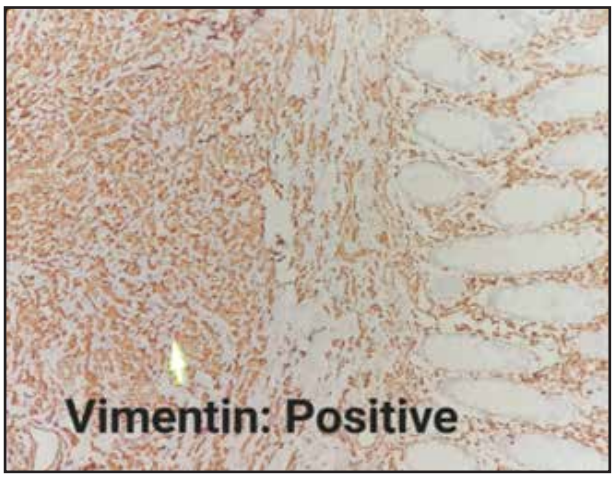

Fig 5b: IHC (Immunohistochemistry) where the neoplastic cells showed Vimentin: Strongly positive (+++), thus immune-morphologically is consistent with non-neural granular cell tumor. (source: Author)

Based on the above findings, a diagnosis of non-neural granular cell tumor was made. After 21 month follow-up, no evidence of recurrence or metastasis was seen.

Discussion

Granular cell tumor is a descriptive term characterized by eosinophilic PAS positive finely granular cytoplasm, which may have neural or non-neural origin. Immunohistochemically, both benign and malignant granular tumor cells typically stain positively with S-100, CD68, neuron-specific enolase (NSE), CD57, inhibin, calretinin, TFE3, SOX10, CD56, PGP95, and vimentin. Vimentin (57-kD protein, also known as fibroblast intermediate filament) is expressed virtually in all mesenchymal cells, including endothelial cells of blood vessels, renal tubular cells, macrophages, neutrophils, fibroblasts, leukocytes, follicular lymphomas, often Reed-Sternberg giant cells (R-S cells) of Hodgkin's disease and many other mesenchymal tumors, that asserts its significant value in detecting mesenchymal tumors. CEA positivity in some cases of GCTs is nothing but false positive reaction, most likely may be due to non-specific cross-reacting antigen/antigens in GCTs. There is still a rarer non-neural variant which stains negatively with S-100, but positively with CD68, CD10, and occasionally with NSE. In some studies, proliferation markers ki-67 and PHH3 were shown to be good predictors of atypical histology.[8-11]

Conventional (traditional) GCTs have a prevalence of about 0.019% to 0.03% of all human tumors. They can occur anywhere in the human body. They account for an incidence of 0.5% of all STTs. It is reported to be most common in the dermis of skin, or less commonly in the subcutaneous tissue of head and neck region, still less often in the smooth muscle, or the striated muscle. They may be found in the internal organs, particularly in the upper ADT (Aero-Digestive tract). In about or over 50% cases of all the conventional GCTs, they may be found in the submucosa of the oral cavity including the tongue. About 6.6% of GCTs are accounted in the breasts by many investigators. In some other study, it was found that they are commonly detected in the mouth at about 40% cases, the skin and subcutaneous tissue at about 30% cases, the respiratory tract at about 15% cases, and the breast at about 15% cases. About 8 to 10% of GCTs occur in the GIT (Gastro-Intestinal Tract), where the most usual location is the esophagus, accounting up to 65% of all gastrointestinal tract GCTs. Still esophageal GCTs are unusual; their incidence has been calculated to be about 0.033%, accounting about 1% of all benign esophageal neoplasms. Occurrence in other such GIT locations as the duodenum, anus, biliary tree, stomach and colon are much less reported, with the rectum being the most unusual site.[12-15]

GCTs are more common in females and black people. Mostly they are detected between the third and fifth decades of life. A granular cell tumor is very often a single, small (commonly 1–3 cm), and painless. Exceptionally mild itching or tenderness may occur. They are commonly slowly growing skin-colored nodules having a smooth or mild rough surface. It is occasional for the clinicians to find an ulcerated GCT. Our reported case here presented with a large bleeding ulcerated benign rectal lesion simulating carcinoma rectum. Most granular cell tumors are benign, although some benign GCTs may be locally aggressive. Malignant GCTs are detected in about 1 to 2% cases of all GCTs. These malignant GCTs are frankly aggressive, with a very poor prognosis. A GCT, 3 cm or less in size, at presentation, should be considered as benign. But if it grows rapidly and forms an ulcer, one should suspect it as a malignant variant. About 10% of GCTs may be multiple, i.e., about two to four lesions, appearing as synchronous lesions.

Conventional neural GCTs are derived from Schwann cells, as confirmed by immunohistocheistry. They are characterized by infiltrative, non-encapsulated nests, cords, or sheets of polygonal and occasionally spindled cells with abundant eosinophilic, finely granular cytoplasm. Many of the tumors have Pustulo-ovoid bodies of Milian, which are large granules with clear halos.[1,16,17]

GCTs of the neurohypophysis are rare tumors seen in the sellar and supra-sellar region, that are poorly understood because of the confusion regarding their nomenclature and indeterminate imaging features. Certain genetic mutations have been linked to GCTs, although the exact mechanisms still remain as very poorly understood. In some case reports, multiple GCTs have been found associated with certain rare syndromes, including Leopard syndrome, Noonan syndrome, and neurofibromatosis type 1.[2,18]

Since the first one reported by LeBoit in 1991, more and more non-neural GCTs have been reported in patients between 5 and 83 years of age in either sex. But none were found in the rectum,

(before this case report). Here, our reported patient with non-neural GCT in the rectum is a middle-aged male person, unlike more of the conventional GCTs occurring in females. Until now, there are very few reported cases of neural GCTs in the rectum, particularly in males. And we failed to detect any case report or case series or article describing non-neural GCT in the rectum.[4,19]

Non-neural GCTs are very unusual uncommon rarer STTs of unknown, uncertain and indefinite histogenesis. At the molecular level, a basic ALK gene re-assortment has been indicated in a little subset of these neoplastic diseases. The abbreviation ALK stands for Anaplastic Lymphoma Kinase. ALK was first reported in anaplastic large-cell lymphoma (ALCL) cell lineage in 1994, as a tyrosine kinase. Most ALK-positive malignant tumors are found as non-small cell lung cancer. The ALK gene is said to persist in human tissues since embryogenesis. It is usually meant for differentiation and normal development of the tissue of the nervous and digestive systems. Some research scientists reported loss-of-function mutations in multiple genes encoding vacuolar H+ -ATPase (V-ATPase) components in the pathogenesis of GCTs.[1,2,4]

Unlike conventional GCTs, our non-neural GCT in the rectum showed a moderate rate of growth of about two years since the first development of symptom, in contrast to classical PNNGCTs that usually show a more rapid rate growth. The PNNGCTs are classically small, about 0.2 to 2.8 cm in size (the median being: 0.5–0.8 cm), in sharp contrast to our case that presented in the form of a large bleeding ulcer. The PNNGCTs are often detected in the trunk. But, Rawal et al. described two cases in the oral mucosa and hard palate. The oral lesions of PNNGCTs are described in a wide age range, like other classical GCTs. Other anatomical sites of most common involvement in cases of PNNGCTs include the tongue, alveolar ridge over tooth/teeth extraction sites, vermilion of the lower lip, etc. These neoplasms typically develop in the sub-epithelial connective tissue. And despite their cellular atypia and often frequent mitotic figures, most authors confirmed excision as curative. Moreover, such complications as lymphogenous/lymhatic metastasis have also been described. Surprisingly, intensive follow-up of these patients showed no remnant and no recurrent (locoregional or distant metastatic) disease. Though the neoplasm is commonly superficially sited, extension beyond the immediate sub-epithelial zone with infiltration into adjacent tissue has also been described. Despite the invasiveness and lymphogenous spread, lymph node dissection or sentinel lymph node biopsy in PNNGCTs is not yet clearly suggested. In our case, we found some invasiveness beyond immediate sub-epithelial tissue and no lymphogenous or no hematogenous metastasis. Histopathologically, we found no necrosis, no mitosis, no cellular pleomorphism and no evidence of malignancy or epithelioid granuloma. Like classical PNNGCTS and unlike classical GCTs, our case showed S-100 negativity and vimentin positivity, that are suggestive of non-neural origin of uncertain mesenchymal lineage. Follow-up of our patient after surgery at 21 months reported no complaint suggesting recurrence or metastasis. Cellular atypia is often a classical feature of PNNGCTs, often without any pseudoepitheliomatous hyperplasia, unlike that seen commonly in cases of conventional GCTs. Our case had neither cellular atypia nor pseudoepitheliomatous hyperplasia. Classical non-neural GCTs are commonly well-circumscribed neoplasms having pleomorphism, and vesicular nuclei, and mitotic figures about 5-6/10 HPF (high-power field), in contrast to conventional benign GCTs that classically have poorly circumscribed margins and zero mitotic figures. The sites of the different GCTs are variable. The conventional GCTs are most often detected in the tongue, mouth, palate, head, neck, limbs, trunks and GITs (GastroIntestinal Tracts), in contrast to the PNNGCTs that are rarer, classically developing in the dermis of the trunk. But our case was a non-neural GCT in the rectum. Under electron microscope, the PNNGCTs, like conventional GCTs, show primitive immature neoplastic cells with variably sized secondary lysosomes. Ultrastructurally, these lysosomal granules are indifferent from those seen in the conventional GCTs. The PNNGCTs do not exhibit clear proof of neural crest ectodermal Schwann cell differentiation, and immunohistochemically negative staining for S-100 proteins and NSE. Congenital gingival GCTs commonly occur only in the alveolar ridge of the jaws of infants with negative staining for S-100 protein. No details are known about these rare oral neoplasms in infants and their pathological similarities and dissimilarities to PNNGCTs. There is no known clearly recognized origin of PNNGCT, but some authors point that these neoplasms may originate from the hair follicle, representing a GCDRSF (Granular Cell Dermal Root Sheath Fibroma). The granular cells are found commonly positive for NKI-C3 and CD68, both pointing to a non-specific reactivity to cytoplasmic lysosomes.[1,2,20,21] Some other neoplasms have granular changes owing to aggregation of lysosomes, like PNNGCT. These neoplasms include conventional GCTs, smooth muscle tumors, papules, atypical fibroxanthomas, dermatofibromas, dermatofibrosarcoma protuberans, fibrous epithelioid cell histiocytomas, perineuromas. PNNGCTs are always negative for cytokeratin, desmin, and CD34, and Smooth muscle actin (SMA) etc., strongly excluding the most of the probable differential diagnoses. Another differential diagnosis contemplates melanoma or a non-malignant melanocytic lesion that exhibits granular cell changes. Here, immunohistochemistry also distinguishes these lesions from PNNGCTs, because melanocytic lesions are found to be positively reacting with S-100 proteins and more melanocytic such diagnostic markers as MITF, HMB-45, or MART-1.[5-8]

The following table (Table 1) shows Differences between granular cell tumors and non-neural granular cell tumors in general, as described by Hernan Mejía et al in 2019.

Table I: Differences between granular cell tumor and non-neural granular cell tumor (Courtesy: Hernan Mejía et al, 2019).

Characteristic	GCT	NN-GCT
Clinical examination		
Location	Head and Neck	Entire body
Oral cavity	Common	Uncommon
Number	Solitary	Solitary
Histology		
Architecture	Polypoid	Polypoid
Level of involvement	Dermis, adipose tissue, muscle	Papillary dermis
Pseudoepitheliomatous hyperplasia	+++	+
Delimitation	+/-	+
Cytologic atypia	+/-	+/++
Mitotic activity	-	++
Pustulo-ovoid bodies of Milian	+	?
IHC reactivity		
S-100	+	-
Vimentin	+	+
CD68	- (71%)	+
Calretinin	+	-*

*may be positive in some cases.

For appropriate management strategy, the non-neural granular cell tumor located in the rectum needs to be rightly differentiated from such other bleeding lesions of rectum as solitary rectal ulcer syndrome, carcinoma rectum, hemorrhoid, rectal polyp, ulcerative proctitis, Crohn's proctitis, tubercular proctitis, lymphomatous proctitis, gonococcal proctitis, syphilitic proctitis, eosinophilic proctitis, etc.[1,2,4-6,8]

Once the diagnosis of any GCT is confirmed, the treatment strategy depends principally on the number of lesions, their sizes and sites, considering other such features as the age and presence of comorbidities, etc. The spectrum of treatment for benign GCTs includes expectant and observation, excision or wide local resection. Resection of all lesions, by either endoscopy or surgery, because of the risk of the malignancy is commonly advised. But less aggressive strategy may be followed with surveillance by endoscopy for asymptomatic small lesions if resection-related risks and morbidity outweigh the potential benefits. GCTs less than 1 cm, as commonly limited to mucosa, can often be successfully removed with biopsy forceps or by standard snare polypectomy. Thermal ablation of GCTs by laser may also be performed. Larger GCTs may be treated by either endoscopic mucosal resection, like other GI (Gastro-Intestinal) sub-mucosal tumors, or by endoscopic submucosal dissection. Despite the advent of advanced endoscopic techniques, it may need laparoscopic surgery or minimally invasive interventions (i.e., laparoscopy-assisted resection with or without colonoscopic guidance or trans-anal resection for rectal lesions) as an effective alternative in selected cases. The choice is to be individualized on basis experience and feasibility.[2,6,7,9]

Recurrent GCTs may occasionally be sensitive to adjuvant radiotherapy. Metastatic lesions from malignant GCTs are to be managed by Chemotherapy. The patients of GCTs are to be kept on follow-up schedule at least annually. Benign GCTs are cured by specific surgical treatment (complete excision). Recurrence can occur even after excision with clear margins. Wide en bloc resection/excision is warranted for malignant GCTs.[1,2,21]

As our case is reported as the first one ever reported non-neural GCT located in the rectum, the above discussion in brief, though doesn't directly and truly reflect our case report, is mentioned here to understand the clinic-pathological features, similarities, dissimilarities, and reasonably explain the management strategy for our ever newly detected tumor in the rectum. That is until now, our case report appears to be the only reported one so far for non-neural GCT located in the rectum. Macroscopically, it was found in the form of a large (5x5 cm) nonspecific ulcer, with no feature of malignancy both macroscopically and microscopically. It was symptomatic in the form of per rectal fresh bleeding for two years. As preoperative biopsy revealed no malignancy and no other specific lesion, we had no other option than to undertake conservative resection in the form of inter-sphincteric resection of rectum and primary colo-anal anastomosis. Grossly and traditionally, indications of ISRR (Inter-Sphincteric Resection of Rectum) includes presence of tumors very close to the ano-rectal ring (roughly within 1 cm or so), when endoscopically, no curative resection of the tumor is possible, like our case. Contraindications of ISRR include tumors invading the levator ani, puborectalis and external anal sphincter (when if operable, abdominoperineal excision of rectum is advised). Other contraindications of ISRR include (in case of malignant disease) poorly differentiated tumors and metastatic diseases. So, in our setting, we think we did the right operation. Our postoperative histopathological and immunohistochemical findings confirmed its benign character and non-neural origin. Our 21 month follow-up findings were also satisfactory in the form of no recurrence, no morbidity and preservation of anal sphincteric function. We think that postoperative minor incontinence (that our patient complained initially) is negligible, and expectantly, his anal continence functions improved quite satisfactorily. Thus, our case report seems to be quite exceptional in multiple dimensions.

Conclusion

The non-neural GCT of rectum we described here, like all other PNNGCTs, is assumed here as an unusually uncommon

neoplasm with benign and innocent behavior. We don't know whether non-neural GCTs have the potentiality of undergoing metastatic spread to lymph-nodes or beyond that. Recognition of these neoplasms is essential to evade mismanagement. The value of sentinel lymph node biopsy has not yet determined. A non-neural GCT at any site is to be surgically treated depending on its own merit, location, size, number, behavior, function of the affected part, age and comorbidity of the patient, etc. This unique non-neural GCT of rhe rectum requires additional research to explain all of its clinical implications. Simple curative excisional surgery can be correctly advised if biopsy fails to reveal any malignancy.

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