J Ped Surg Case Reports 10 (2016) 23–25

Contents lists available at ScienceDirect

Journal of Pediatric Surgery CASE REPORTS

journal homepage: www.jpscasereports.com

The first endoscopic resection of a colonic granular cell tumor in a child

Basak Erginel [a,*], Melih Akin [a], Abdullah Yildiz [a], Canan Alatas Alkim [b], Canan Tanik [c], Cetin Ali Karadag [a], Nihat Sever [a], Meltem Tokel [a], Ali İhsan Dokucu [a]

[a] Sisli Etfal Training and Research Hospital, Department of Pediatric Surgery, Istanbul, Turkey
[b] Sisli Etfal Training and Research Hospital, Department of Pediatric Gastroenterology, Istanbul, Turkey
[c] Sisli Etfal Training and Research Hospital, Department of Pathology, Istanbul, Turkey

ARTICLE INFO

Article history:
Received 28 February 2016
Received in revised form
17 April 2016
Accepted 23 April 2016

Keywords:
Granular cell tumor
Endoscopic resection
Pediatric

ABSTRACT

Granular cell tumors (GCTs) are rare, generally benign, soft tissue tumors characterized by large, granular, periodic acid-Schiff (PAS) positive eosinophilic cells that are S-100 and neuron specific enolase positively stained. They are mostly located in the oral cavity and skin. GCTs are not common in the gastrointestinal tract, where the most common location is the esophagus. These tumors are usually seen in the 4th and 6th decade of life and are rare in children. To our knowledge, only one case report exists regarding the endoscopic resection of a cecal GCT in a child. Herein, we report an endoscopic resection of a colonic GCT in a child.

Granular cell tumors (GCTs) are rare, generally benign, soft tissue tumors mostly located in the oral cavity and skin. GCTs are not common in the gastrointestinal tract (1–8% of cases) [1]. The most common location of these tumors in the gastrointestinal tract is the esophagus [2]; the colon is a very rare site for this tumor. A number of surgical techniques, both open and laparoscopic, are suggested for the resection of the tumor. Due to the submucosal origin of these tumors, resection of the tumor with endoscopic method seems adequate for the benign ones but needs close follow-up for synchronous and metachronous tumors or for local recurrence [3,4]. Herein, we present the first endoscopic resection of a colonic GCT in a child.

1. Case

A 13-year-old female was admitted with polyp prolapsus during defecation. She had no rectal bleeding or weight loss. On rectal examination, no mass could be palpated. A colonoscopy was performed, and a large pedunculated polypoid mass was found almost 110th cm from the anus on the ascending colon. A 2.6 × 2.5 × 2-cm polipoid mass with a stalk, covered with normal mucosa, was resected by hot snare polypectomy. 1% epinephrine and 0.8% indigo carmine solution is injected prior to the hot snare resection, submucosally under the tumor. The histopathological examination revealed a granular cell tumor of the colon. Two weeks after the first endoscopy, a second colonoscopy was performed for the exploration of residual tissue. The stump of the first mass, which was 1 × 1 × 1.1 cm in diameter, was resected by hot snare again, and a punch biopsy from the stalk was performed on the border of the tumor.

Under microscopic analysis of both specimens collected from these two different colonoscopies, the tumor was found within the thin fibrovascular stroma with an ulcerated surface. It was composed of diffusely scattered regular cells with small nuclei and clear granular cytoplasm, forming well-organized structures in some parts (Fig. 1). Therefore, the tumor was diagnosed as a GCT.

On immunohistochemical analysis, focal staining was observed with CD 56, S-100, neuron specific enolase (NSE), CD 68, and vimentin (Fig. 2). The punch biopsy from the stalk after the second colonoscopy and polypectomy revealed no infiltration by the tumor, so the tumor was accepted as a tumor free margin. The diagnosis of a GCT of the colon was made based on the findings of both immunohistochemical and morphologic appearance. The absence of mitosis and a proliferation index of less than 2% favored the benign character. A metachroneous mass or metastasis was not determined on computer tomography of the thorax, abdomen and cranium. A control colonoscopy and upper gastrointestinal system

* Corresponding author. Yildirim Oguz Goker Sokak, 5 Gazeteciler Sitesi, C-1 Blok, No. 36, Akatlar, Besiktas, Istanbul, Turkey.
E-mail address: basakerginel@hotmail.com (B. Erginel).

http://dx.doi.org/10.1016/j.epsc.2016.04.020

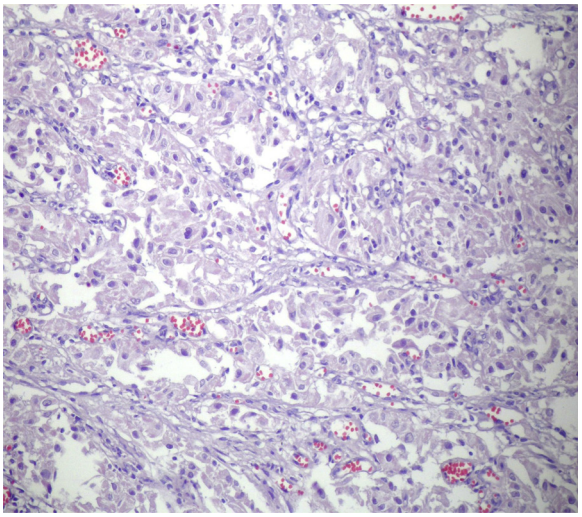

Fig. 1. Diffusely scattered regular cells with a small nucleus and clear granular cytoplasm on hematoxylin eosin stain (×40).

endoscopy was repeated after three months, and no residual mass was identified. The patient was well at her three-year follow-up visit.

2. Discussion

Granular cell tumors are uncommon soft tissue tumors that are thought to have a neuronal origin. They are characterized by large, granular, periodic acid-Schiff (PAS)-positive eosinophilic cells [5]. A positive staining of both the cytoplasm and nuclei of granular cells for S-100 protein and NSE is characteristic for these tumors [5]. In this case, Cytokeratin 7 and 20, chromogranin A and synaptophysin were not stained. Due to the negative staining with cytokeratin 7 and 20, epithelial malignancies were excluded, while due to the negative staining with chromogranin A and synaptophysin, carcinoid neuroendocrine tumors were excluded [6]. Smooth muscle antigen, CD 34, carcioembrionic antigen, epithelial membrane antigen, P53 and human melanoma black 45 were also negatively stained. The histological findings for our patient were thus compatible with those of granular cell tumors (GCTs).

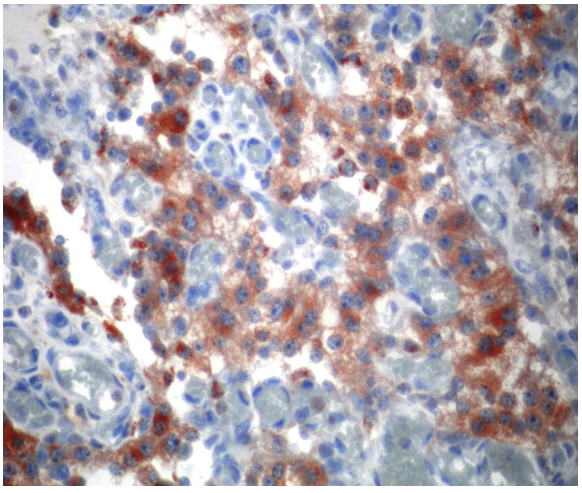

Fig. 2. Large, granular, periodic acid-Schiff (PAS) positive eosinophilic cells that are S-100 positively stained.

GCTs comprise a heterogeneous group of tumors with both congenital and adult types [7]. The origin of the adult type is the Schwann cell, while the origin is unknown in the case of the congenital type. S-100, NSE, alpha 1 antitripsin (A1AT), CD 68 and vimentin are positive for the adult type, an A1AT and CD 68 are positive for the congenital type. Cytokeratin, SMA, demsin, CD57, CD15 and MAC 387 are negative for both types. According to these findings, our patient's diagnosis is adult type GCT and accepted as benign [8–10].

They may occur anywhere in the body, whereas the most common presentation site is tongue and skin [11]. Although most are benign, 1.5–2.7% of GCTs are thought to be malignant. Therefore, resection of these tumors and histopathologic evaluation should be considered for the further treatment. The tumor usually occurs in the 4th and 6th decades of life [12]. Although Sultan et al. reported an endoscopic resection of a cecal GCT in a 10-year-old female patient [13], ours is the second report of an endoscopic resection of a colonic GCT in a child.

Endoscopic resection appears to be safe and effective in adults. Take et al. [14] recently reported a series of 11 successful cases, Khang et al. [15] reported a series of 25 adult patients with 27 benign colorectal GCTs, and Singhi et al. [16] reported 26 cases of colorectal GCTs successfully resected endoscopically in adult patients.

Although there are no guidelines for the treatment of these tumors, conservative management is considered for the ones <2 cm and benign in histopathologic examination for adult patients. Malignancy is correlated to the increasing size of the tumor (>4 cm). Although there is no consensus to follow-up on pediatric patients, we offered an endoscopic follow-up for this patient. Metastasis can be visualized by CT. We did a CT because the mass was almost 4 cm in length and the patient was an adolescent. We do not routinely recommend a CT for small lesions with adult patients due to the low likelihood of malignancy.

With the recent increased use of endoscopy in pediatric patients, these tumors are being more frequently found in children during colonoscopy than before. The possibility of a GCT of the colon should be considered with polypoid tumors of the colon that are coincidentally realized in endoscopy in pediatric patients. Endoscopic resection in these tumors must be considered before proceeding with further aggressive resections. Endoscopic resection seems safe and effective for GCTs in pediatric patients as well as in adults but needs close endoscopic follow-up for metachronous tumors and local recurrence. Gastroscopy should also be considered for synchronous tumors in such children.

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