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Case Report

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MULTIPLE METACHRONOUS GRANULAR CELL TUMORS OF BOTH GASTROINTESTINAL TRACT AND SKIN: AN UNCOMMON CLINICAL ENTITY

Katic V[1], Petrovic A[1], Simonovic M.[1], Grgov S.[2], Rankovic G.[3], Rankovic A.[1], Todorovic J.[1], Mladenovic M[1]

Vuka.

***Corresponding Author: Dr. Katic V.**

Vuka.

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ABSTRACT

Granular cell tumor (GCT) is soft tissue neoplasm occuring in the skin and internal organs. Its clinical behavior is usually benign, although both histological and clinical malignant forms can occur. We report a 52 years man , musician with very hard, sesile, vericous tumor in distal oesophagus, discovered endoscopically. It appeared as a yellow hemispheric protrusion with a thin mucous membrane known as "sweet corn".Patient was presented with dysphagia, gastralgia and substernal pain, 6 years ago. Wide local excision of the verrucous lesion has been done endoscopically. Histologically, oesophagus showed the overlying pseudoepitheliomatous hyperplasia, so extensive that it has been mimick a squamous carcinoma. However, on the base of histologically, histochemically and immunohistochemically examinations , GCT diagnosis has been revealed. In the skin, solitary, brownish dome-shaped nodus has been discovered on periumbilical skin, surrounded by generaluzed lentiginosis without any symptom (" beauty mark", by patient s opinion), existing during last 15 years. The excised lesions did not recur, but newer lesions continued to discover during the last four years: in stomach, duodenum and coecoascendens. On colonoscopy, induced by abdominal colic, the large (to 3 cm) and numerous submucous, nodular yellowish masses with hyalinisation and calcification were found in caecoascendens. GCT diagnosis of colon,on small endoscopical biopsies, was pointed out, followed by right colectomy, 23 cm of length, with good clinical course (24 - month follow-up). Eight months after surgery, patient experienced hemathemesis and melena. Gastroduodenoscopy was performed, revealing numerous white solid and confluent nodules, up to 1 cm in diameter in stomach, while the only one ulcerous change was found in duodenum. By using the biopsies and S- 100 protein, marker, has been also confirmed GCT. Two Years after right colectomy, the new lesions did not revealed in gross appearance. Except he is afraid of dead, the patient is with good health, " living for music and from music", how he likes to say.

KEYWORDS: granular cell tumor, gastrointestinal tract, skin, histopathology, diagnosis, therapy.

INTRODUCTION

Granular cell tumor (GCT) is a benign, mesenchymal and uncommon soft tissue tumor (first described as “granular cell myoblastoma” by Abrikossoff in 1926).[1] Among many hypotheses, the two suggest a possible origin from Schwann or neural cells and from undifferentiated mesenchymal cells.[6, 8, 13] GCT tumors can occur in any part of human body and their common locations are the tongue, breast and subcutaneous tissue.[5,6] Esophagus is the most often affected submucous organ of digestive tract, until GCTs of the colon are very rare.[7] These tumors can manifest as solitary, submucosal nodules or as a multifocal lesions in lower percent.[8, 9] Clinical behavior of GCTs is usually benign, although both histological and clinical malignant forms can also occur, but in only 1-2% in all GCTs patients.[9] GCTs express markers associated with neural differentiation, such as S-100, CD56 and neuron-specific enolase (NSE). The tumor cells are also positive for vimentin.

We are reporting a patient with 28 multiple metachronous granular cell tumors of both gastrointestinal tract and skin, which were removed by both endoscopy and surgical resection.

CASE REPORT

Esophagus

Herein, we report a case of multifocal cutaneous, submucosum oesophageal, stomach, duodenum and colon caecoascendens in a young male, for the rarity. (? of the entity skin). In March 2010, a 45-year-old man, singer by profession, is presented with a 4-month history of epigastralgia, dysphagia and substernal pain. Endoscopic examination of the esophagus demonstrated a sessile protruding polypoid changes of the surface epithelium with a smooth yellow white surface, covered with non-ulcerated mucosa. The change has been removed and sent to patohistological examination. The biopsy specimens also contained a deep submucosal layer of the esophagus. Histological examination of

sections stained with hematoxylin - eosin showed that tumor was located beneath the squamous epithelium, forming nests, strands and sheets of polygonal cells (Fig. 1). Nuclear pleomorphism, prominent nucleoli and mitotic figures were uncommon. The fine eosinophilic, intracytoplasmic PAS positive and diastase resistant granules corresponded to lisozyme (Fig. 2). A characteristic feature of the esophagus GCT was the pseudoepitheliomatous hyperplasia of the mucosa, overlying the tumor, mimicking a well-differentiated squamous cell carcinoma (Fig. 3). Tumor cells were polygonal with abundant granular PAS positive cytoplasm, arranged in cord-like structures, and with small atypical hyperchromatic nuclei, but without mitosis (Fig. 3). In addition, S-100 protein was displayed in granules by the use of ABC technique (Fig 4), suggesting Schwann cells, as the cell of origin of this tumor. From these findings, the tumor was diagnosed as granular cell tumor of the esophagus (benign type).

Skin: In the skin, solitary, brownnish dome-shaped nodus has been discovered on periumbilical skin, surounded by generalized lentiginosis without any symptom („beauty mark "by patients opinion), existing during last 15 years. The last esophagoscopical control was performed after six years of its discover. The lesion has remained unchanged in gross appearance. In this way, the authors have confirmed the suggestion that granular cell "myoblastoma" of the esophagus is more frequent in benign than malign form. Patient received only symptomatic therapy.

Colon

Seven years from then, now a 52 years old man, was presented with unspecific symptoms. He experienced strong pain in abdomen and obstipation. Colonoscopy was performed, and in caeco-ascendant part of the colon numerous polypoid changes were found. During the colonoscopy, numerous samples were collected for pathological examination and PAS positive staining revealed granular cell tumor (Fig. 3 D-treba da bude A). Right hemicolectomy was performed (Fig. 4). Numerous (n=28), nodular and firm yellowish masses with hyalinization and calcification were found, and the largest lesion was 30 mm. Histopahology confirmed the preoperative diagnosis of GCT. Tumors are presented like nonencapsulated masses, composed of large polyhedral cells, containing faintly eosinophilic and uniform coarse granules. Nuclei are small, hyperchromatic and centrally located. Mitoses are uncommon (Fig. 3 A-C – treba da bude B-D).

Gastroduodenum

Eight months after the surgery patient experienced hematemesis and melena. Gastroduodenoscopy was performed. Numerous white, firm and confluent nodules, up to 1cm in diameter, were found in stomach, while the ulcerous change was found in duodenum. Tissue samples were collected, fixed in 10% buffered formalin. Paraffine sections were stained with HE, PAS and ABC immunohistochemical method, by using S-100 protein, specific marker, to confirm the neural differentiation of GCT (Figure 5). Patient received therapy for Helicobacter pylori eradication. Two years after the gastroduodenoscopy, the patients feels well. Three years since right colectomy, no recurrence he has not noted.

DISCUSION

Very first forms of the granular cell tumor, described by Weber in 1854[10] and by Abrikossoff in 1926, were localized in the tongue.[11] The origin of the GCTs long remains unclear for many scientists. By electron microscopic study, as well as by immunohistochemical measuring of S-100 protein, or NSE, has been confirmed that the GCTs originate from Schwann cells (Koichi Miwa, Hattori, granular cell tumor of esphagus 1986).[12,13]

In literature GCT are described as benign (more often) and malignant tumor (1 – 2%), first described by Ravich in 1945.[15] Main classifications of the malignant form of the CGTs are: histologically and clinically malignant type, and histologically benign but clinically malignant type, giving the metastases in lung, bone, and lymph nodes.[16] For the classification of malignancy are rapid growth, ability to frequently recur after resection and the following six histological criteria: nuclear pleomorphism, high nuclear/cytoplasm ratio, increased mitotic rate (>2/10 high power fields), vesicular nuclei, large nucleoli and tumor cell necrosis.[17,18]

Our patient's biggest lesion was 30 mm in diameter and the number of lesions was 28. Despite this fact, malignant potential was not proved and the up mentioned criteria were not full field.

According to the patohistological studies, the predilection places of GCTs are tongue, skin, subcutaneous tissue, head and neck and gastrointestinal tract.[18,19] Incidence for GCTs in gastrointestinal tract has the range from 4-8%.[20] Esophageal GCTs are rare, with only a few cases presented in the published literature. However, implementation of the EUS and advances in endoscopic resection made the diagnosis of these tumors more easily.

Macroscopic presentation of the gastrointestinal GCTs is typically as a solitary, submucosal, yellow-white, small nodule, covered by a normal-appearing mucosa or as multifocal lesions in lower percent of patients.[8,9] Microscopically GCTs are made of large polygonal cells which containing numerous eosinophilic granules and small, uniform nuclei and neural markers, including S-100 protein or NSE.

GCTs could appear at any age. Commonly they are found between the third and fifth decades of life. In contrast to a small female predominance.[21] our patient is male.

In our study, the patient was 45 years old in the time when the CGTs diagnosis was confirmed.

GCTs occurred in a gastrointestinal tract is as a solitary lesions, but sometimes multiple tumors were occasionally reported, as has been confirmed in our case.[8,9]

In patients with multiple GCTs described in published literature, the tumor locations were confined within the same organs. Patients have had double colonic GCT, or double esophageal GCT case, or multiple gastric GCTs, where first tumor was in the antrum, whereas the second GCT was in the same area. Authors of existing studies report that esophageal GCTs predominantly occur in the distal esophagus.[22]

In our case the GCTs was primary detected in esophagus as a sessile protruding verrucous change of the surface epithelium with a smooth yellow white surface, covered with non-ulcerated mucosa, later in caecoascendence and stomach, and solitary subepidermal mass in skin, which is tipicaly in the literature descirbed presentation of the CGTs.

According to the Thumallapally N et al**.** esophageal presented GCTs are often without symptoms and "the majority of lesions are found incidentally during the investigation for other problems".[23] Our patient had epigastralgia, dysphagia and substernal pain in the begining and later, when the other parts of gastrointernal parts were involved, he experianced pain, obstipation and melena.

The most common endoscopic characteristics of esophageal GCT is the presence of a sessile, yellow-white-to-grey elevated lesion with intact mucosa. Rarely, affected patients may have ulceration and necrosis.[8,9] In our case, the ulceration of the lesion was found in duodenum.

THERAPY

Endoscopic polypectomy (endoscopic submucosal dissection) for oesophagal GCTS has become increasing (ENDOSCOPIC THERAPY WAS PERFORMED WITHOUT RELAPSE AFTER 6- YEAR FOLLOW-UP.

CONCLUSION

GCTs are histologically unique and we reported a rare multiple metachronous soft tissue tumor in a rare localization. These tumors can be often manifested with nonspecific clinical symptoms and can pose a diagnostic challenge to medical health care providers. Awareness of GCTs is imperative for the radiologist, surgeon, and pathologist and should be included in the differential diagnosis of gastrointestinal and skin masses. Nevertheless, close follow-up of the once confirmed GCTs should be considered.

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Napomena.

Poseban Conclusion, poslat tebi (1,2,3...) uzmi u obzir a ne ovaj.