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Case Report

Atypical esophageal granular cell tumor: Case report [☆]

Zineb Benchekroun, MD^{a,*}, Amal Akammar, MD^a, Hajar Bennani, MD^a, Meriem Haloua, MD^a, Youssef Alaoui Lamrani, MD^a, Meriem Boubbou, MD^a, Leila Chbani, MD^c, Mustapha Maâroufi, MD^a, Badreddine Alami, MD^{a,b}

^a Department of radiology, CHU HASSAN II FES, Morocco

^b Laboratory of neuroscience of the medicine of Sidi Mohamed Ben Abdellah faculty of Fez, Morocco

^c Anatomic pathology department of CHU HASSAN II of FEZ, Morocco

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ABSTRACT

Esophageal granular cell tumors (GCTs), or also called Abrikossoff's tumor are rare benign tumors originating from Schwann cells most commonly found in the skin, subcutaneous tissue, and tongue. Approximately 5%–8% arise in the gastrointestinal tract, and one-third of these tumors arise in the esophagus [1]. This neoplasm has a benign behavior usually, but there have been described a malignant transformation in 2%–3% of the cases. Herein, we discuss a case of a 70-year-old male patient with no pathological background, admitted for dysphagia evolving in 3 months that was explored with endoscopy and CT, the diagnosis at this level was challenging but the histopathology and Immunohistochemistry confirmed the presence of granular cells thus confirm the diagnosis.

The purpose of our work is to report the uncommon evolution of an Abrikossoff 's tumor located in the esophagus, as a warning of the possible malignant transformation of this tumor mostly benign; also we made a review of the literature.

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Introduction

Granular cell tumors formally known as myeloblastomas or Abrikossoff tumors, most frequently occur in the skin and subcutaneous tissues but can also be found in the gastrointestinal (GI) tract especially in the esophagus (30%–60%) [2,3].

Esophageal GCTs are rarely diagnosed based on its imaging features, therefore this case is being reported in view of the rarity of the lesion and to discuss the various differential diagnosis especially when it's locally aggressive.

GCTs are mostly considered benign tumors according to their clinical, radiological and histological appearances, however few cases of metastasis and local aggressiveness were

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* Corresponding author. Zineb Benchekroun

E-mail address: zineb.benchekroun@usmba.ac.ma (Z. Benchekroun).

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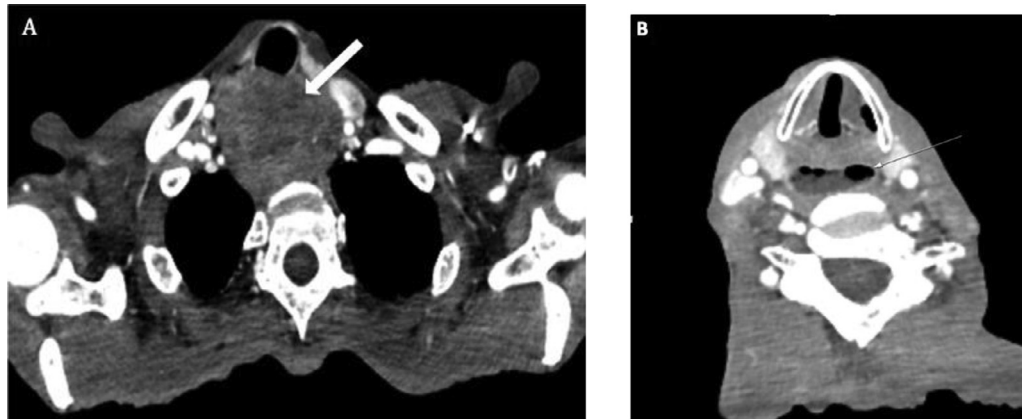


Fig. 1 – Thoracic CT with axial thin slices showing a large homogenous esophageal mass with well-demarcated margins (A) (white arrow) responsible of a narrowing of the esophageal lumen with a dilation of the upstream cervical esophagus (B) (white arrow).

reported in the literature [4,5]. In fact, some studies proposed six histologic criteria for selection of atypical or malignant cases including: increased nuclear-to-cytoplasmic ratio; nuclear pleomorphism; necrosis; spindling of tumor cells; vesicular nuclei with prominent nucleoli; and a mitotic count of more than two in 10 high-power fields (200×field).

Case presentation

A 70-year-old patient with no pathological background, presented to the department of gastro-enterology for progressive dysphagia 3 months prior to evaluation, and weight loss estimated to 10 kg. The physical examination found a dehydrated and undernourished patient, with no further abnormalities. Upper gastrointestinal endoscopy was performed showing an impassable tumor filling the Killian triangle, thus the biopsy couldn't be performed endoscopically. Therefore, the patient underwent a CT scan (Fig. 1) that showed a huge posterior mediastinal mass measuring 60 mm of the greatest diameter, with an esophageal epicenter, presenting well-defined margins with no extension to the adjacent structures. (Fig. 2)

Afterwards, a CT guided biopsy was performed (Fig. 3). The final pathologic diagnosis (Fig. 4) showed a poorly defined tumor composed of sheets of cells or nests separated by thin collagenous stroma (HES*200) that was confirmed by immunohistochemistry (Fig. 4) showing a strong staining for S100. These findings were consistent with a diagnosis of an esophageal Granular cell tumor.

Once the diagnosis was made, the case was discussed in a multidisciplinary consultation meeting that concluded to a feeding jejunostomy as a temporary solution before the subtotal oesophagectomy. Unfortunately, immediately after the jejunostomy the patient discharged against medical advice, and died 1 month after at home.

Discussion

Granular cell tumors (GCTs) are rare and mostly considered benign according to their clinical, radiological and histological appearances. They were first described by Abrikosoff in 1931 and can appear in the tongue, skin, breast, respiratory tract, biliary system, and digestive tract [6]. Several detailed reviews found that approximately 1% – 8 % of all GCTs occur in the gastrointestinal tract [7,8]. Esophageal CGT is considered the most common site of occurrence with a predilection for the distal part (60% of the cases)[7]. In fact, in a study led by Yongsheng Shi and all (Experience with Esophageal Granular Cell Tumors: Clinical and Endoscopic Analysis of 22 Cases) it was reported that approximately 65% of esophageal GCTs are located in the distal part of the esophagus, 20% in the middle part of the esophagus, and 15% in the proximal part of the esophagus [9]. However, another report claimed that the most common location of esophageal GCTs is the middle third of the esophagus [10], and in our case the lesion was detected in the upper esophagus.

CGT are usually solitary tumors, but few cases of multiple lesions in the esophagus or synchronous granular cell tumors in other organs were reported in the literature in 5%–12% of the cases [9,11]. In our report, we have a male patient, which is inconsistent with previous studies that have reported a female predominance with female to male ratio 2.9:1. Esophageal GCTs can occur at any age, but most of these tumors occur in patients between the ages of 40 and 60 years old [12,13].

The clinical manifestations of CGT depend on the size of the lesion. In fact, patients with small lesions less than 20 mm in diameter are usually asymptomatic and the lesions are found incidentally during endoscopy or radiography [14]. However, patients with greater diameter lesions, can present with symptoms such as dysphagia in the first place or with other less common symptoms like the gastro-esophageal reflux disease, dyspepsia, chest pain, cough or nausea.

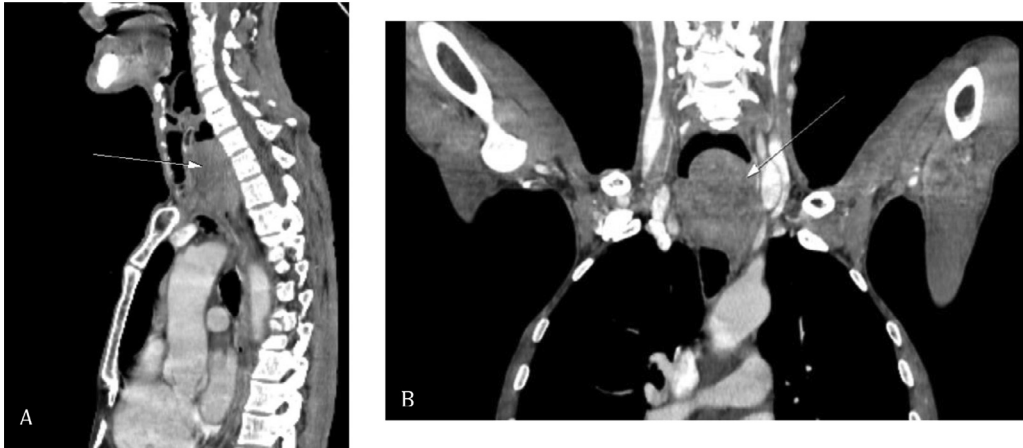


Fig. 2 – Thoracic CT with coronal and sagittal MPRs showing the esophageal tumor with well-demarcated margins with no further extension to the thoracic vertebrae nor to the trachea or the jugulo-carotid vessels.

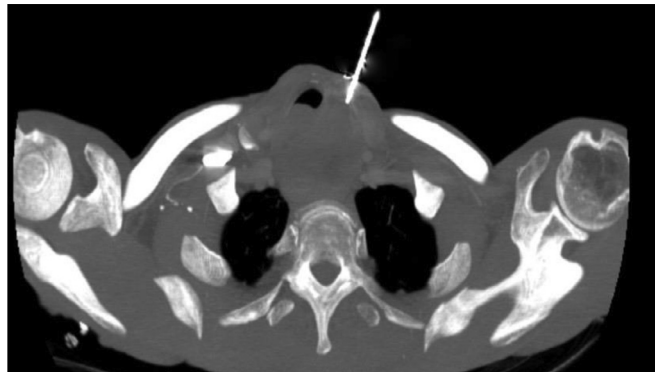


Fig. 3 – Thoracic CT with axial thin cuts showing the biopsy site and the path of the troca.

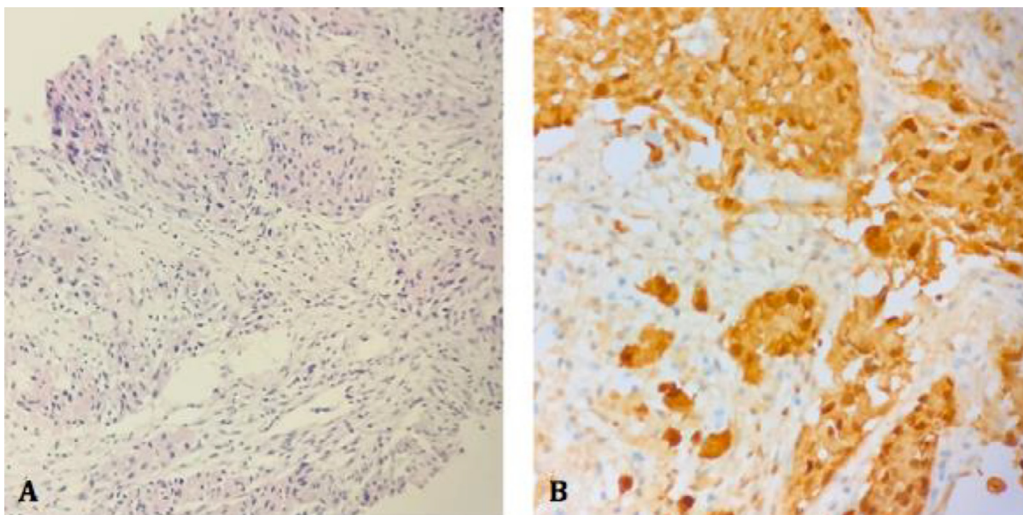


Fig. 4 – (A) Histopathologic examination revealed a poorly defined tumor composed of sheets of cells or nests separated by thin collagenous stroma (HES*200). (B) Immunohistochemistry showing a strong staining for S100.

The diagnosis of Esophageal GCTs is based on the histopathological exam. In fact, there are various methods for tissue diagnosis, including endoscopic biopsy, endoscopic ultrasound or CT-guided fine-needle biopsy. The frequent endoscopic aspect of esophageal GCT presents as a submucosal mass. Whereas the tumor appears as a round, hypo echoic, and homogeneous lesion with clear borders in the endoscopic ultrasound. CT scans on the other hands show mostly a benign tumor appearance described as a well defined tumor with clear borders and no further invasion to the adjacent structures, however few cases of metastasis and local aggressiveness were reported in the literature [4][5] making less than 2% of all lesions [7] and defining malignant GCT. Besides local aggressiveness the size of the tumor is also important. In fact, the malignant potential of GCT increases when it exceeds 50 mm in diameter [15], which is the case in our patient (tumor measuring 60 mm).

As discussed above, the diagnosis of a granular cell tumor can be challenging especially when it comes to the imaging findings. As a matter of fact, the GCTs can have the same radiologic appearance as most of other benign tumors of the esophagus. They all present as smooth in-tramural or intraluminal mass without ulceration or nodularity at barium examination and absence of peritumoral invasion, lymphadenopathy, or distant metastases at cross-sectional imaging. However, the majority of these tumors are small especially when it comes to leiomyomas appearing as smooth or slightly lobulated masses with a whorled appearance, they enhance homogeneously but may contain calcifications, whereas fibrovascular polyps appear as intraluminal masses generally located in the cervical oesophagus, its radiological appearance depends on the proportions of fat and fibrous tissue in it explaining the heterogeneous appearance. [16,17,18]. The radiologic appearance of other benign mucosal based neoplasms of the esophagus including squamous papillomas and adenomas cannot be distinguished from that of a polypoid early esophageal carcinoma [19]. Especially the adenomas that are sessile or pedunculated polyps seen oftenly in Barrett esophagus, necessitating resection because of the risk of malignant transformation

Moreover, Hemangiomas, schwannomas, neurofibromas, and glomus tumors, are some other extremely rare, benign intramural neoplasms of the esophagus, that can be seen.

The typical histological appearance of GCT on one hand is a solid and firm tumor, located in the mucosa or submucosa, non-enveloped with a yellow or yellowish cross section. The tumors are usually composed of sheets or nests of rounder polygonal large cells with abundant eosinophilic granular cytoplasm and small, round, central uniform nuclei [20]. Collagen fiber bundles often separate the tumor cells. The immunohistochemical staining shows GCTs to be positive to S100 protein, neuron-specific enolase, vimentin, and various myelin proteins, and negative for smooth muscle actin and desmin [21].

On the other hand, histological criteria for malignant GCT were proposed by Fanburg-Smith and associates [5], including necrosis, spindling, vesicular nuclei with large nucleoli, a high nuclear-to-cytoplasmic ratio, increased mitotic activity (more than 2 mitoses per 10 high-power fields at 200 magnification), and pleomorphism. Therefore a GCT is considered malignant

if at least three of these criteria are fulfilled. If there are only one or two criteria the tumor is classified as atypical, and there is only focal pleomorphism with none of the other criteria it is classified as benign. In our case, we had no malignant criteria so technically our tumor was considered as benign. However, its radiological size defined it as malignant, so with the gathering of both aspects we concluded to an atypical tumor.

Regarding the treatment, it is determined usually by the endoscopic ultrasonography. As a matter of fact, small lesions less than 1 cm in diameter are preferably followed-up regularly to avoid complication and a conservative approach is preferred [22]. Whereas tumors sized 1 cm or greater or presenting any of the malignancy criteria cited below undergo endoscopic or surgical resection [23]. Endoscopic resection includes endoscopic mucosal resection and sub-mucosal tunnel endoscopic resection. The choice between these techniques depends on the size of the tumor and its attachment to the muscularis propria. The last remaining choice and the most invasive one is surgery, including video-assisted thoracoscopic surgery and conventional open surgery. It is indicated for tumors with high suspicion of malignancy, deep layer invasion, contraindication for endoscopic dissection. In the multidisciplinary consultation meeting held to discuss our case, it was decided for our patient to undergo a surgical resection because of the deep layer invasion of the tumor.

Conclusion

To conclude, GCTs are rather uncommon benign neoplasms but can also show some malignancy criteria defining 3 histopathological types (benign, atypical and malignant tumor). The endoscopical and radiological aspects depend on the degree of malignancy but are mostly round tumors with well-defined margins. The diagnosis is based on the histology which characteristic appearance show large granular cells arranged in groups, with strips of connective tissue separated from the surrounding areas by collagen.

Patient consent

The patient first was discharged against medical advice, then was reported deceased by the family. We tried to reach the family for a consent statement but there were no further answer.

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