Case Report

Imaging of perianal granular cell tumor with lung metastasis: A case report and literature review

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A B S T R A C T

The granular cell tumor or Abrikossof's tumor is a rare tumor, most often benign in evolution. Malignant forms are exceptional. We report, here, a very rare case of granular cell tumor, localized in the perianal region, in a 54-year-old woman with lung metastases. CT and MRI with contrast showed a locally advanced tumor process in the right para-anal region associated with multiple "balloon release" lung lesions. The diagnosis was confirmed by immunostaining after surgical biopsy. Very few cases of malignant granular cell tumors with lung metastasis have been reported in the literature.

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Introduction

Granular cell tumor (GCT), or Abrikossof tumor, was initially described by Abrikossof in 1926 [1]. Predominantly a benign tumor, 1%-2% of the cases are reported to be malignant [2-4]. Women between their 4th and the 6th decades are most commonly affected [5]. Granular cell tumor can occur in any body site and is most commonly seen in the skin and oral cavity (30% occur in the tongue) [6,7]. About 10% of the tumors develop in the gastrointestinal tract, with the esophagus being the most common site and the rectum being the rarest [8]. With only a few cases of perianal localization being reported in the literature, we present an exceptional case of a malignant perianal GCT tumor with pulmonary metastasis.

Patient and observation

A 54 years old woman with no medical comorbidities consults for proctalgia evolution, accentuated during defecation, for 8 months. Digital rectal examination finds no palpable mass; only pain was assessed in the right para-anal region. The CT scan objectified a locally advanced tumor process in the right para-anal region (Fig. 1), associated with multiple pulmonary metastases (Fig 2). MRI showed a low signal in T1 weighted sequences (Fig 3), a high signal in T2 sequences (Fig 4), a high...
Fig. 1 – Axial (A), coronal (B) and sagittal (C) CT scan showing a mass occupying the right para-anal region.
Fig. 2 – CT scan of the lung showing metastatic pulmonary nodules.

Fig. 3 – T1 weighted image showing low signal of the mass.
signal in T1 injected sequences (Fig. 5), and a high signal in diffusion images (Fig. 6). The patient underwent a surgical biopsy which the histology concluded to be a granular cell tumor. Afterwards, the patient The patient was referred to oncology for palliative chemotherapy.

Discussion

Granular cell tumor is a neoplasm of mesenchymal origin and derives its name from the presence of coarse cytoplasmic granularity typically found among its constituent cells [9]. It is more common in females and occurs predominantly in the age group of 40-60 years. It can occur in any part of the body, with the most common sites being the oral cavity and tongue, followed by the skin and subcutaneous tissue. GCTs in the lower digestive tract, especially in the perianal areas, are relatively rare [5]. GCTs are commonly asymptomatic. Although GCTs are commonly unifocal, some may present in multiple locations [10]. Macroscopically, the tumor can present as a nonulcerated nodule or a yellowish-gray sessile polyp with a firm consistency. Because of its asymptomatic presentation, its manuscript (with Author Details) is often found incidentally and frequently mistaken for other submucosal tumors such as carcinoid tumors [11], with both tumors being mucosal or submucosal in location and having similar endoscopic findings, from which it should be differentiated. Histologically, it is thought to have a neurocytological origin [12].

GCT is characterized by the presence of PAS-positive granules, abundant lysosomes, and small and uniform nuclei. Immunohistochemistry shows strong and diffuse positivity for S-100 [10,13]. Imaging features of GCTs are relatively non-specific. On endoscopic ultrasound (EUS), GCT appears as a small (95% <2 cm), hypoechoic, solid, homogenous tumor with invasion of the inner and/or outer layers of the gastrointestinal tract (mucosa and/or submucosa) [14]. Computed tomography can show a well-circumscribed, homogeneous soft tissue mass, typically without calcification. MRI imaging features of GCTs are reported as heterogeneous low signal intensity on T1-weighted images and heterogeneous high signal intensity on T2-weighted images, as well as a heterogeneous or homogenous enhancement on T1WI [15]. GCTs are mostly benign tumors; however, 1%-2% of them can be malignant. A rapid tumor growth or a size greater than 3 cm and ulceration should raise the suspicions of malignancy, such as features like an invasion of the adjacent organs, nuclear and cellular pleomorphism, and mitotic figures [2,3]. With lungs and lymph nodes being the most common sites of metastases [16], definitive diagnosis of GCT is only established by endoscopic biopsy and histopathological studies. Complete excision to negative margins with close clinical follow-up is recommended for granular cell tumors in nearly all locations, whether benign or malignant. Actually, there is no current standard chemotherapy regimen for malignant or metastatic forms. The use of adjuvant radiation therapy has been similarly controversial and ill-defined. In literature, less than 50 cases of granular cell tumors in the perianal
Fig. 5 – T1 injected sequence showing an intense and heterogeneous signal of the mass.

Fig. 6 – Diffusion sequence showing a high signal of the mass.
region have been described, from which 3 reports with malignant characteristics highlight the rareness of the tumor in this localization [17].

Conclusion

We reported a rare case of a malignant perianal GCT tumor with pulmonary metastasis, through which we learnt that the diagnosis is difficult due to non-specific clinical and radiological aspects, and that histology remains the only way to establish a certain diagnosis.

Authors’ contributions

Narjisse Aichouni: conception, literature review, analysis, data collection, writing, review and editing. Yassine Amane: conception, literature review, analysis, data collection, writing, review and editing. Imane Kamaoui: contributor. Siham Nasri: contributor. Imane Skiker: Supervision and data validation. All the authors have read and agreed to the final manuscript.

Patient consent

An informed consent was obtained from the patient.

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