

## Gastric Abrikosoff tumor (granular cell tumor): case report

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**SUMMARY: Gastric Abrikosoff tumor (granular cell tumor): case report.**

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*Granular Cells Tumor (GCT), also called Abrikosoff tumor, is very uncommon lesion of neural derivation. It is characterized by the presence of granular cell; benign and malignant counterparts are known, even if the second ones are rare. It has a slight predominance in female sex and black race; the age range is wide, with peak between fourth and sixth decades of life. Any localisation is possible, although surface lesions (head, neck, trunk, extremities) are far more common than visceral ones (esophagus, stomach, small and large bowel, larynx, bronchi, gallbladder and biliary tract). Surgical en-bloc excision is curative for both benign and malignant forms. Radiotherapy and chemotherapy are not effective.*

*We report the case of a 45 year old man who had a cytologic diagnosis of fusocellular stromal tumor of the gastric fundus during examination for gastritis. He underwent a wedge resection of the gastric wall: at the histological examination neoplastic cells had a granular cytoplasm and immunoassay was positive for S100 protein, PGP 9.5 and NSE.*

*Complete excision guarantees from recurrence and metastases: however a long term endoscopic follow-up is necessary.*

**RIASSUNTO: Tumore di Abrikosoff (tumore a cellule granulari) dello stomaco: case report.**

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*Il tumore a cellule granulari, anche detto tumore di Abrikosoff, è una neoplasia rara di derivazione neurale. È caratterizzato dalla presenza di cellule granulari; sono conosciute forme a comportamento sia benigno che maligno, anche se queste ultime sono poco frequenti. Si presenta con una leggera predominanza nel sesso femminile e nella razza nera; lo spettro dell'età è ampio, con un picco tra la quarta e la sesta decade. Può localizzarsi in tutti gli epiteli, anche se le lesioni cutanee (testa, collo, tronco ed estremità) sono molto più frequenti di quelle viscerali (esofago, stomaco, intestino tenue, colon, laringe, bronchi, colecisti, vie biliari). L'excisi chirurgica "en bloc" è terapeutica sia nelle forme benigne che nelle maligne. La chemioterapia e la radioterapia non sono efficaci.*

*Riportiamo il caso di un uomo di 45 anni al quale, a seguito di una esofagogastroduodenoscopia con biopsie effettuate per pirosi epigastrica, era stata posta diagnosi citopatologica di tumore stromale a cellule fuse del fondo gastrico. È stato sottoposto a resezione a losanga della parete gastrica. All'esame istologico le cellule presentavano granulazioni citoplasmatiche e l'esame immunoenzimatico dimostrava positività per proteina S100, PGP 9.5 e NSE.*

*L'escissione locale completa è sufficiente e scongiura il rischio di recidiva e metastasi: tuttavia è opportuno un follow-up endoscopico a lungo termine.*

KEY WORDS: Abrikosoff - Granular cell tumor - Stomach - Endoscopy - Surgery.  
Abrikosoff - Tumore a cellule granulari - Stomaco - Endoscopia - Chirurgia.

### Background

Granular Cells Tumor (GCT), also called Abrikosoff tumor, is an uncommon lesion derived from Schwann

cells. It was described for the first time by Abrikosoff in 1926 (1). It is characterized by granularity of tumoral cells, due to the accumulation of secondary lysosomes in the cytoplasm.

Any localisation is possible, although surface lesions (head, neck, trunk, extremities) are far more common than visceral lesions. Visceral involvement is encountered as mucosal or submucosal nodules in the esophagus, stomach, small and large intestines, larynx, bronchi, gallbladder, and biliary tract. The gastrointestinal tract harbors approximately 5% of all granular cell tumors.

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Benign and malignant counterparts are known, even if the second ones are rare (fewer than 2%) (2). It is estimated to account for 0.03% of all tumors affecting humans (3). It appears to be more common in black race, and a slight female sex predominance exists (3:2 female to male).

Lesions can be incidental findings, or they may give rise to obstructive or pressure symptoms when they are large enough and in a critical location. At pathology tumor cells have abundant granular eosinophilic cytoplasm with centrally located vesicular or pyknotic nuclei (5). At immunohistochemical examination, the tumor cells stain positively for S-100 protein, neuron-specific enolase (NSE), and NK1-C3 in almost all cases. Positivity with stains for myelin-associated P0 and P2 proteins, myelin basic protein, and Leu-7 is less consistent (6).

## Case report

We describe the case of a 45 year old man, in good clinical conditions, who presented dyspepsia; the upper endoscopy showed a mass of the gastric body, covered by normal mucosa. Erosive duodenitis HP+ was present too. Then, an echoendoscopy with fine needle aspiration biopsy was performed; the lesion measured 2 x 1,3 cm. The cytologic examination revealed this to be a "stromal fusocellular lesion".

Patient was submitted to surgery: at the abdominal cavity exploration the lesion appeared strong in consistence and with regular borders. No other lesions were present. A wedge resection of the whole gastric wall was performed.

The post-operative course was regular, with no complications.

The histologic diagnosis was "granular cell tumor"; immunohistochemical analysis was positive for S100 protein, PGP 9.5, NSE. There wasn't mitotic activity (Ki-67 immunoreactivity was 0%). The lesion was widely included in the margins of section.

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No other therapy was necessary. Endoscopic follow-up has been made every year for early detection of local recurrence.

## Discussion

At present there are no guidelines for the treatment of GCT. Two opposite standpoints are reported: a conservative approach by routine endoscopic follow-up when the tumor measures <10 mm, without evidence of malignancy; surgical excision for large (>20 mm) benign GCTs causing symptoms and when malignancy is suspected (7).

Histologic features of malignant GCTs include necrosis, spindling, vesicular nuclei with prominent nucleoli, high nucleocytoplasmic ratio, cellular pleomorphism, and mitotic figures (Ki-67 immunoreactivity > 10%) (8). However, some malignant lesions are indistinguishable from the benign counterparts; large size (>4 cm), rapid growth, rapid recurrence after previous excision and invasion of the adjacent tissues are reported to be indicative of malignant behavior. Lymph nodae metastases are rare (9).

If the excision is complete, local surgical resection is curative for benign granular cell tumors; wide "en bloc" excision is recommended for malignant lesions. Radiotherapy and chemotherapy are not effective (4).

## Conclusion

Surgical wedge resection represents the first choice treatment of GCT, for large size or symptomatic tumors and when it is impossible to exclude a malignant behavior of the lesion.