Malignant granular cell tumor of the abdominal wall

Tumore a cellule granulari maligno della parete addominale

I. CHELLY, K. BELLIL, A. MEKNI, S. BELLIL, M. BELHADJSALAH, N. KCHIR, S. HAOUET, M.M. ZITOUNA
Departement of Pathology, Hospital “La Rabta”, Tunis, Tunisia

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Summary
The granular cell tumor is an uncommon tumor that usually appears as a solitary small nodular growth and runs a benign course. It occurs widely throughout the body, but is rarely described in the abdominal wall. The authors report a case of malignant granular cell tumor which was arising in anterior abdominal wall of a 67-year-old woman. Malignant variant is rare and the abdominal wall site is extremely uncommon. Regarding this clinical case and the literature the authors purpose to review the criteria of malignancy.

Introduction
Granular cell tumor (GCT) initially described by Abrikossoff as “myoblastic myomata” in 1926, is an uncommon soft tissue tumor, usually benign with uncertain histogenesis. This lesion may develop from any part of the body, predominantly in the head and neck region, and especially on the tongue. Abdominal wall growth is very rare and, to our knowledge, there have only been two previous reports of abdominal GCT.

Case report
A 67 year-old white woman, with surgical history of hepatic hydatic cyst, was admitted in “la Rabta Hospital” in Tunisia (North Africa) for abdominal pain evolving over the previous 2 months. She did not have a fever and the abdominal examination showed tenderness during deep palpation in the right upper quadrant associated with a fixed, firm mass. No evidence of peripheral adenopathy or cutaneous nodules was noted. Computed tomography revealed a hypodense, single mass in the anterior abdominal wall measuring about 10 cm in its greatest diameter. In addition, the tumor was not connected to the liver in any way; no cystic changes or necrosis were observed (Fig. 1). However according to the surgical history, presumptive diagnosis of recurrent hydatic cyst was proposed and surgical excision was carried out. During surgery, a large, irregular, anterior abdominal wall mass measuring up to 10 cm in diameter was discovered. It extensively invaded normal adjacent structures and fat tissue in the absence of any regional adenopathy. The tumor was not completely resected due to its large size and the extensive infiltration of normal structures. The surgically removed specimen consisted of a white, irregular, firm mass measuring 6 x 4 x 3 cm. Histologic examination revealed a solid, soft tissue mass composed of large, round or polygonal tumor cells with abundant, finely granular, eosinophilic cytoplasm containing acidophilic periodic acid Schiff (PAS)-positive, diastase-resistant granules (Fig. 2). The nuclei showed slight variability; most were small, ovoid to round, vesicular and eccentrically located. No atypical cells and no mitotic figures were seen. The cells were arranged in small nests and cords, most ranging from three to 10 cells in size, with indistinct cellular borders (Fig. 3). The tumor was not encapsulated and had invaded adjacent soft tissue. No necrosis and no vascular invasion were identified. Immunohistochemical stains were performed on formalin-fixed, paraffin-embedded tissue sections according to the standard avia-
MALIGNANT GRANULAR CELL TUMOR OF THE ABDOMINAL WALL
din-biotin-peroxydase complex (ABC) method. Antibodies against the following were used: keratin, desmin, S-100 Protein, smooth muscle actin, vimentin, NSE, CD68 and proliferation markers (MIB-1, PCNA). Neoplastic cells showed a strong positive and diffuse cytoplasmic reaction to anti-S100-protein (Fig. 4), NSE, CD68 and anti-vimentin. Reaction for other antibodies was negative. The final diagnosis was of granular cell tumor which was most probably malignant due to the size and invasion. The patient died due to pulmonary embolism three month later.

Discussion

GCTs may arise anywhere in the body but are most frequently found in the head and neck and in the oral cavity (particularly in the tongue), as small, solitary painless nodules. Rarely, it occurs as multiple or solitary lesions in the gastrointestinal tract (10%)2, urogenital tract, central nervous system 3, extra-hepatic biliary ducts, muscle, breast and respiratory tract. Only two cases have been reported in the abdominal wall. They both appeared in the anterior abdominal wall simulating an aponevrotic fibroma. The first, published in 1992, involved a 32 year old Indian woman 4. The second, reported in 1993, occurred in a 67 year-old woman presenting a small aponevrotic granular cell tumour, measuring 2 cm in size, without any biological or histological criteria of malignancy 5. Our case most likely represents the first case of abdominal wall GCT with malignant potential.

GCTs occur most commonly in 40 to 70 year old patients, as often in women as men, with an afro-american predominance. The diagnosis of GCT depends on pathological findings, more especially on the observation of plump histiocyte-like, bland-looking neoplastic cells with abundant granular eosinophilic cytoplasm containing acidophilic, PAS-positive, diastase-resistant granules with neural differentiation. Expressed markers include S-100 protein, CD68 (KP-1), protein gene product 9.5, inhibin-alpha and NSE 2-6. Ultrastructurally,
cytoplasmic granules appear as membrane-bound vacuoles of variable size and shape containing debris, disrupted mitochondria, and myelin structures suggesting schwann cell origin. The neoplastic cells typically express CD68 due to cytoplasmic lysosome content. The significance of inhibin expression with regards to cell differentiation and pathogenesis is unclear and warrants further investigation. GCT should be included in the differential diagnosis of abdominal masses. It can mimic gastrointestinal stromal tumor with epithelioid cells which have CD117 expression. Granular cell reactions bear a close similarity to GCT but can usually be differentiated from it by the fact that the intracytoplasmic granular material is effectively acid fast and autofluorescent as compared with that of GCT. The prognosis in any location is quite good, but a very rare malignant form (1-2%) is described. Fewer than 100 cases have been reported in the literature since their first description in 1945 by Ravich et al. These are epidemiologically similar to their benign counterparts. Furthermore, distinction between benign and malignant GCT is difficult because of histologic similarity and lack of reliable criteria that can predict clinical behaviour. In this way, high cellularity, nuclear pleomorphism, prominent nucleoli, mitotic activity and Ki-67 proliferative index cannot reliably distinguish between benign, atypical or recurrent GCT. However, more than 60% of metastatic GCTs were larger than 4 cm in diameter. Locally infiltrative growth, large size (greater than 5 cm), multifocal occurrence, recurrence at a later date are important but the presence of metastases appears to be the only irreparable criterion of malignancy. Our case most likely represents a malignant granular cell tumor. This diagnosis is suggested by the large tumor size (10 cm) and local invasion. Malignant GCT have a worse prognosis. Approximately 60% died after metastatic invasion, most often within a few years after diagnosis, in regional lymph nodes, lungs, viscera and bone. Only complete surgical resection reduces the risk of recurrence.

References